Question 1
A 30-year-old man has a painful ankle mass. Radiographs are seen in Figures 1a and 1b, and MRI scans are seen in Figures 1c and 1d. What is the most likely diagnosis?

1. Synovial hemangioma
2. Synovial sarcoma
3. Lipoma arborescens
4. Synovial chondromatosis
5. Pigmented villonodular synovitis

PREFERRED RESPONSE: 4

DISCUSSION: The calcified loose bodies shown on the imaging studies are characteristic of synovial chondromatosis. Pigmented villonodular synovitis and lipoma arborescens do not show calcifications. Synovial sarcoma and synovial hemangioma can mineralize but they tend to be more diffuse, and not associated with loose bodies. Synovial sarcoma is rarely intra-articular.

Question 2

Figures 2a through 2d show the radiographs, MRI scan, and bone scan of a 44-year-old man who twisted his knee and felt a pop. Following the injury he had swelling; however, both the pain and swelling have now resolved. What is the next most appropriate step in the management of the patient’s distal femoral lesion?

1. Radiographic follow-up
2. Biopsy
3. Surgical resection
4. Radiation therapy
5. Chemotherapy

PREFERRED RESPONSE: 1
DISCUSSION: The lesion seen on the radiographs and MRI scan is consistent with an enchondroma. Enchondromas are commonly encountered by orthopaedic surgeons as an incidental finding during evaluation of a patient for pain of other causes. Enchondromas are painless, show no aggressive bone destruction, and commonly exhibit radiotracer uptake on a bone scan. Surgery for enchondromas is not generally necessary, but when clinical and radiographic features suggest a potential chondrosarcoma, action is necessary. Chondrosarcoma almost always occurs in adults. Patients usually present with pain and have characteristic findings on staging studies. Lucencies developing within the calcification, periosteal reaction, and an associated soft-tissue mass all are characteristic features of chondrosarcoma. Biopsies are not typically indicated because of the histologic similarity of benign and malignant cartilage lesions. Radiographic follow-up is appropriate when the clinical suspicion of chondrosarcoma is low. In this patient, the bone lesion was discovered incidentally because of unrelated trauma and the patient is essentially asymptomatic with benign-appearing radiographic features.

Question 3
Figures 3a through 3e show the radiographs and MRI scans of a 16-year-old boy. Which of the following best describes features of the patient’s tumor?

1. Telomere translocations
2. Supernumerary ring chromosomes
3. A translocation involving genes 11 and 22
4. A rearrangement involving the X and 18 chromosomes
5. Genetic alterations in the retinoblastoma gene and p53 tumor suppressor gene

PREFERRED RESPONSE: 5

DISCUSSION: The imaging studies show a classic case of conventional high-grade osteosarcoma. Genetic alterations in the retinoblastoma gene and p53 tumor suppressor gene are common in this tumor. A translocation involving chromosomes 11 and 22 is characteristic of Ewing’s sarcoma. Supernumerary ring chromosomes are seen in parosteal osteosarcoma. Telomere translocations have been described in giant cell tumor of bone. A translocation of chromosomes X and 18 is typical of synovial sarcoma.
Question 4

A 12-year-old girl has a 6-month history of a painful expanding mass in the right lateral ankle. A clinical photograph, radiograph, MRI scans, and histology specimens are seen in Figures 4a through 4f. What is the most appropriate management?

1. Resection alone
2. Resection and chemotherapy
3. Resection and radiation therapy
4. Chemotherapy and radiation therapy
5. Observation

PREFERRED RESPONSE: 1
DISCUSSION: The radiograph shows an expansile radiolucent lesion without matrix production or a periosteal reaction. On close inspection, there are subtle internal septations on the mortise view. The MRI scans show heterogeneous septated areas with prominent fluid-fluid levels. The histopathology on low power shows villous projections and prominent clefts, whereas the high power view reveals prominent spindled stromal cells and interspersed multinucleated giant cells. This is an aneurysmal bone cyst. The treatment of aneurysmal bone cyst is generally intralesional surgery (eg, curettage). However, for patients with large destructive lesions in expendable bones such as the fibula, excision is preferred. The main differential diagnosis in this patient is telangiectatic osteosarcoma. Radiographs of telangiectatic osteosarcoma show geographic bone lysis, a wide zone of transition, and matrix mineralization. Under the microscope, telangiectatic osteosarcoma exhibits pleomorphic cells with at least focal malignant osteoid production. Telangiectatic osteosarcoma is treated with chemotherapy and wide excision.

Question 5
Figures 5a and 5b show the AP radiograph and axial CT scan of an 18-year-old woman who has proximal thigh pain. What is the most appropriate treatment?

1. Observation
2. Curettage and grafting
3. Radiofrequency ablation
4. Wide resection
5. Wide resection and chemotherapy

PREFERRED RESPONSE: 3

DISCUSSION: The imaging studies are classic for osteoid osteoma, a benign lesion. Conventional treatment has involved burring down to find the lesion, followed by curettage. More recent experience with radiofrequency ablation under CT guidance has shown equivalent results with few complications. Curettage in this location would carry significant morbidity and risk for fracture, making radiofrequency ablation the most appropriate treatment. More aggressive resections are not indicated because the lesion has a low chance of recurrence. Observation and/or use of nonsteroidal anti-inflammatory drugs would be reasonable if the patient is willing to live with prolonged pain.

Question 6
What are the five soft-tissue sarcomas that can frequently metastasize to the lymph nodes?

1. Synovial sarcoma, angiosarcoma, osteosarcoma, chondrosarcoma, Ewing’s sarcoma
2. Chondrosarcoma, osteosarcoma, fibrosarcoma, Ewing’s sarcoma, adamantinoma
3. Rhabdomyosarcoma, synovial sarcoma, epithelioid sarcoma, clear cell sarcoma, angiosarcoma
4. Myxoid chondrosarcoma, fibrosarcoma, malignant peripheral nerve sheath tumor, liposarcoma, malignant fibrous histiocytoma
5. Rhabdomyosarcoma, myxoid liposarcoma, acral myxoinflammatory fibroblastic sarcoma, synovial sarcoma, epithelioid sarcoma

PREFERRED RESPONSE: 3

DISCUSSION: When sarcomas spread, they classically metastasize to the lung. Their second most common site to metastasize to is either another bone or soft tissue, depending on whether it is a primary bone or soft-tissue sarcoma. Regional metastases are relatively a rare occurrence - only about 5% of patients with soft-tissue sarcomas. The incidence is slightly higher in patients with rhabdomyosarcoma, synovial sarcoma, epithelioid sarcoma, clear cell sarcoma, or angiosarcoma. In these patients, consideration for sentinel node biopsy should be given.

Question 7
A 36-year-old man has had an enlarging left posterior thigh soft-tissue mass for the past month, and he now reports numbness and tingling in his sciatic nerve distribution. Based on the MRI scan and biopsy specimen shown in Figures 7a and 7b, what is the most likely diagnosis?

1. Myxoid liposarcoma
2. Synovial sarcoma
3. Malignant fibrous histiocytoma
4. Fibrosarcoma
5. Well-differentiated liposarcoma

PREFERRED RESPONSE: 1

DISCUSSION: Most soft-tissue sarcomas have a similar MRI appearance and cannot be differentiated by signal intensity characteristics. Histologic and often immunohistochemical analysis is necessary to subtype soft-tissue sarcomas. Myxoid liposarcomas account for one half of all liposarcomas, with a peak incidence during the fifth decade. Myxoid liposarcomas have a characteristic histologic appearance with myxoid background and an interlacing network of fine vessels. The delicate plexiform capillary vascular network is present throughout these tumors and provides an important clue for distinguishing them from myxomas. Treatment consists of wide surgical resection. Synovial sarcomas have a monophasic or biphasic histologic pattern. Malignant fibrous histiocytoma (undifferentiated pleomorphic sarcoma) is composed of pleomorphic cells in a storiform pattern. Fibrosarcoma has a typical “herringbone” pattern on low power histology. Well-differentiated liposarcoma has bland histology of adipocytes with scattered lipoblasts.

Question 8
An otherwise healthy 15-year-old boy is seen in the emergency department for an injury sustained while playing dodge ball. A radiograph is shown in Figure 8. What is the most appropriate treatment for this patient?

1. Aspiration and injection
2. Resection and reconstruction
3. Sling and pain medication
4. Curettage and bone grafting
5. Intramedullary nail fixation

PREFERRED RESPONSE: 3

DISCUSSION: The patient has a pathologic fracture through a benign-appearing eccentric bone lesion with a soap-bubble geographic appearance, most suggestive of a nonossifying fibroma. The best initial treatment is to provide comfort measures and allow the fracture to heal. Aspiration and steroid injection has been advocated for a unicameral bone cyst, though it should be done after any associated fracture has been allowed to heal. Wide resection is not indicated for this benign tumor. Extended curettage and grafting may be appropriate, but only after fracture healing has occurred unless open reduction and internal fixation is indicated for the fracture itself. Intramedullary nail fixation is unnecessary and generally should not be performed if the histopathologic diagnosis is not known.

A 7-year-old boy has a 3-week history of neck pain. History and physical examination reveal no neurologic symptoms. A radiograph, CT scan, and MRI scans are seen in Figures 9a through 9d. Figure 9e shows a needle biopsy specimen. Based on these findings, what is the most appropriate management?

1. Cervical collar and observation
2. Intravenous antibiotics and cervical collar
3. Chemotherapy and radiation therapy
4. Neoadjuvant chemotherapy alone
5. Neoadjuvant chemotherapy and surgical resection

PREFERRED RESPONSE: 1

DISCUSSION: The findings are consistent with eosinophilic granuloma. Eosinophilic granuloma is a focal destructive lesion of bone commonly seen in children younger than age 10 years. Despite the very aggressive appearance of these lesions, they typically will heal spontaneously or following biopsy. Local corticosteroid injection has been advocated by some. Surgical decompression is indicated in rare cases of cord compression. Chemotherapy is indicated only in disseminated forms of histiocytosis.

Question 10
A 12-year-old boy is diagnosed with osteosarcoma of the left distal femur. The lesion measures 7 cm in its greatest dimension and is associated with a soft-tissue mass that extends into the vastus lateralis. A CT scan of the chest is normal. A bone scan reveals another lesion in the contralateral right proximal femur. Biopsy of the right proximal femur lesion also reveals osteosarcoma. According to the American Joint Committee on Cancer (AJCC), what is the stage of this tumor?

1. II-A
2. II-B
3. III
4. IV-A
5. IV-B

PREFERRED RESPONSE: 5

DISCUSSION: The AJCC staging system for bone sarcomas is based on tumor grade, size, and the presence as well as location of metastases. Stage I tumors are low grade. Stage II tumors are high grade. Stages I and II are subdivided based on size. Stages I-A and II-A are less than or equal to 8 cm in their greatest linear measurement. Stages I-B and II-B are greater than 8 cm in size. Stage III tumors are those that have “skip metastases” which are defined as discontinuous lesions within the same bone. Stage IV-A involves pulmonary metastases, whereas stage IV-B includes patients with non-pulmonary metastases; therefore, the stage of this tumor would be IV-B.

Question 11
A 28-year-old woman has a painless mass on the dorsum of the foot that has been rapidly increasing in size over the last 3 months. It measures 5.7 cm in maximum diameter. A lateral radiograph of the foot is shown in Figure 11a. Sagittal T1-weighted and coronal T2-weighted MRI scans are shown in Figures 11b and 11c. A photomicrograph from the biopsy specimen is shown in Figure 11d. The most appropriate surgical treatment would likely include which of the following?

1. Intralesional excision
2. Marginal excision and split-thickness skin grafting
3. Aspiration and antibiotics
4. Wide resection and flap coverage
5. Below-knee amputation

PREFERRED RESPONSE: 4
DISCUSSION: The clinical presentation, imaging studies, and histology are consistent with a high-grade soft-tissue sarcoma. The MRI scans show a heterogeneous lesion. Histology demonstrates a spindle cell lesion with pleomorphism, atypica, and large bizarre mitoses. Treatment of soft-tissue sarcomas should include at least a wide resection. Intralesional or marginal excision is not advised for high-grade sarcoma. Below-knee amputation is an option for this patient, but most likely limb-sparing procedures for this particular sarcoma would be possible and also more functional while also allowing proper oncologic surgical resection. On this area of the dorsum of the foot, split-thickness skin grafting would likely have a higher failure rate than a fasciocutaneous flap over the tendons. Whereas aspiration of a fluid-containing cystic lesion (ganglion cyst) would be appropriate, it is not appropriate for this solid, heterogeneous lesion.

Question 12
Figures 12a and 12b show the radiographs of a 22-year-old man who reports left knee pain. An MRI scan is seen in Figure 12c, and a photomicrograph is seen in Figure 12d. What stage is this lesion according to the Enneking staging system?

1. I
2. II
3. III
4. 1
5. 2

PREFERRED RESPONSE: 5
DISCUSSION: The radiographs show a purely radiolucent lesion in the distal femur with no cortical destruction. There is no soft-tissue extension noted on the MRI scan. The photomicrograph demonstrates multiple giant cells with a background of mononuclear cells, confirming the diagnosis of giant cell tumor. This lesion is classified as a stage 2 (benign active) tumor according to the Enneking staging system. Using this system, malignant tumors are staged using Roman numerals, and benign tumors are staged using Arabic numerals. Stage 1 tumors are “latent.” Stage 2 tumors are “active,” and stage 3 tumors are “aggressive.” Whereas some giant cell tumors are stage 3, the lesion in this patient does not demonstrate particularly aggressive imaging features, such as cortical breakthrough and soft-tissue extension.


Question 13
A 9-year-old boy with a history of unilateral retinoblastoma has a 2-week history of left arm pain. His mother, who had bilateral retinoblastoma, noted a mass on his left proximal arm 2 days prior to the office visit. A radiograph, MRI scan, and biopsy specimen are seen in Figures 13a through 13c. What is the most appropriate treatment?

1. Observation
2. Wide resection
3. Radiotherapy and wide resection
4. Chemotherapy and wide resection
5. Extended curettage and bone grafting

PREFERRED RESPONSE: 4
DISCUSSION: The radiograph and MRI scan show an aggressive lesion in the left proximal humerus in a child with a history of retinoblastoma. The biopsy specimen shows pleomorphic spindled cells with focal osteoid. There is a well-documented association between retinoblastoma and osteosarcoma because it has been found that most, if not all, such tumors have defects in their RB1 pathway through genetic lesions in the RB1 gene itself or other genes in the pathway. Linkage analysis at the retinoblastoma gene (RB1) locus is required for identification of individuals at risk of developing retinoblastoma and osteosarcoma. Identification of disease-causing mutations is necessary for accurate risk prediction. The treatment for osteosarcoma is chemotherapy and surgery, either wide local resection or amputation. Radiotherapy is not a useful adjunct in the treatment of osteosarcoma. Chemotherapy and wide resection is the preferred treatment.


Question 14
An otherwise healthy teenager has an isolated lower extremity mass. The biopsy of the mass reveals the diagnosis of rhabdomyosarcoma. Management of the patient’s mass should consist of which of the following?

1. Hemipelvectomy amputation
2. Wide resection, chemotherapy, and radiation therapy
3. Wide resection and radiation therapy
4. Marginal excision
5. Observation

PREFERRED RESPONSE: 2

DISCUSSION: Treatment of rhabdomyosarcomas includes wide resection of the primary tumor, chemotherapy, and radiation therapy. Rhabdomyosarcomas are one of the soft-tissue sarcomas where chemotherapy has been shown to have a substantial survival benefit. In all cases without a radical margin, radiation therapy should also be added to the treatment plan. Amputation is rarely needed for local tumor control and is typically selected when an amputation provides better functional results than a limb-sparing procedure. Observation of these tumors is rarely, if ever, indicated. Sentinel node biopsy may be indicated in these patients if there is clinical or radiographic node enlargement.


A 28-year-old man has had left shoulder pain for 1 year. The pain is constant and has steadily worsened over time. History reveals that he underwent curettage for a “noncancerous bone tumor” 5 years ago. A radiograph, bone scan, MRI scan, and biopsy specimens are shown in Figures 15a through 15e. What is the most appropriate treatment for this patient?

1. Wide resection alone
2. Resection and chemotherapy
3. Intralesional curettage and grafting
4. Forequarter amputation
5. Radiation therapy and prophylactic internal fixation

PREFERRED RESPONSE: 1

DISCUSSION: The radiographs and histology are consistent with a clear cell chondrosarcoma. The treatment is wide resection. No chemotherapy is indicated because this is a locally aggressive tumor with minimal risk of metastasis. Intralesional curettage may be appropriate for a benign bone tumor but not for a clear cell chondrosarcoma. Amputation and radiation therapy are not indicated to achieve local disease control. This case illustrates the occasional problems with diagnosis of this tumor.

Figure 16a

Figure 16b

Question 16

A 45-year-old man has a 6-month history of a leg mass and recent ulceration of the skin. The clinical photograph and biopsy specimen are seen in Figures 16a and 16b. What is the most likely diagnosis?

1. Ewing’s sarcoma/peripheral primitive neuroectodermal tumor
2. Extraskeletal myxoid chondrosarcoma
3. Dermatofibrosarcoma
4. Myxoid/round cell liposarcoma
5. Synovial sarcoma

PREFERRED RESPONSE: 3

DISCUSSION: Dermatofibrosarcoma is a rare, monoclonal, cutaneous sarcoma arising in the dermis in the trunk, 47%; lower extremity, 20%; upper extremity, 18%; and head and neck, 14%. Symptoms may be present for a duration of 6 months to 30 years. It may occur at any age, with a peak incidence in the fourth decade; it is more common in men than women (3:2). Most tumors are superficial and less than 5 cm, but 3% are larger than 10 cm. They present early as pink or violet-red plaques surrounded by telangiectatic skin. A nodular growth pattern with ulceration and attachment to deeper structures is observed in advanced and/or recurrent cases. Most cases (85% to 90%) are low grade but (5% to 15%) contain focal, high-grade fibrosarcomatous areas with intermediate-grade tumor (DFSP-FS). MRI is useful in ascertaining tumor extent and depth of invasion. Tumor cells exhibit a storiform growth pattern and infiltrate adjacent adnexal structures and adipose tissue. Fibrosarcomatous transformation can occur over time and is recognized by increased mitotic activity. Because some tumors express platelet derived growth factor, they may be responsive to imatinib. Ewing’s sarcoma, extraskeletal myxoid chondrosarcoma, liposarcoma, and synovial sarcoma are generally deep-seated tumors that can but very rarely cause skin ulceration.

An otherwise healthy 52-year-old man has had a several year history of a slowly enlarging, symptomatic left shoulder, axillary, and chest wall mass. Radiographs are only remarkable for a large soft-tissue mass. Selected sequences of MRI scans are shown in Figure 17a (T<sub>1</sub>), Figure 17b (T<sub>2</sub> fat saturated), and Figure 17c (T<sub>1</sub> fat saturated post-gadolinium). Management of this symptomatic mass should consist of which of the following?

1. Forequarter amputation
2. Wide resection, chemotherapy, and radiation therapy
3. Wide resection and radiation therapy
4. Marginal excision
5. Incisional biopsy

PREFERRED RESPONSE: 4

DISCUSSION: The MRI scans show a large soft-tissue mass that is iso-intense with subcutaneous fat of all sequences. This is diagnostic of a lipocytic (fatty) tumor—either lipoma, atypical lipomatous tumor, or low-grade liposarcoma. As such, no biopsy is necessary because biopsies of these fatty tumors can be fraught with sampling error. The most appropriate treatment of this symptomatic lesion is simple, marginal excision without radiation therapy or chemotherapy. Local recurrence can occur in 25% to 50% of patients at 10 years. Dedifferentiation is rare with subsequent recurrent disease; but when it occurs, it may result in metastases.
Tumoral calcinosis is a hereditary disease that involves which of the following?

1. Calcium metabolism
2. Phosphate metabolism
3. Oxalate metabolism
4. Chloride metabolism
5. Renal tubular insufficiency

PREFERRED RESPONSE: 2

DISCUSSION: Although not completely understood, the phosphate metabolic dysfunction seen in tumoral calcinosis is a hereditary disease. This is characterized by soft-tissue lesions that are calcified, lobulated, well-demarcated lesions usually found over the extensor surfaces of large joints. It is most commonly found around the hip, elbow, shoulder, foot, and wrist joints. Renal tubular insufficiency is associated with acidosis, glucosuria, phosphaturia, aminoaciduria, and mild proteinuria. Oxalate metabolism can contribute to calcium oxalate crystals and kidney stones. Oxalate is found in many food products. Calcium metabolism, which can be abnormal in renal insufficiency, hypervitaminosis D, and milk-alkali syndrome, is normal in tumoral calcinosis. Metabolic alkalosis is often associated with a alteration in chloride metabolism.

A 25-year-old man has a painful mass in a web space of his foot. MRI scans are seen in Figures 19a (T\textsubscript{2} STIR) and 19b (T\textsubscript{1}), a representative gross specimen is seen in Figure 19c, and a H&E stain is seen in Figure 19d. What is the most likely diagnosis?

1. Melanoma
2. Synovial sarcoma
3. Interdigital neuroma
4. Epithelioid sarcoma
5. Giant cell tumor of tendon sheath

PREFERRED RESPONSE: 5
DISCUSSION: Giant cell tumors of tendon sheath are common in the hands and feet. Because of significant hemosiderin deposition, they commonly appear hypointense to skeletal muscle on both T1 and T2 pulse-weighted sequences. The hemosiderin is manifested in the brownish discoloration in the gross specimen. The photomicrograph shows bland spindled stromal cells and abundant multinuclear giant cells. Treatment is marginal excision with relatively low rates of tumor recurrence. Although the foot is not an infrequent site of melanoma and there are some shared radiologic features with giant cell tumor of tendon sheath, histologically melanoma is composed of cells both spindled and epithelioid arranged in nests or clusters. Synovial sarcoma is the most common sarcoma of the foot which radiographically has mineralizations in 30% of cases. It is typically heterogeneous on both MR pulse sequences. Microscopically, monophasic synovial sarcoma contains spindled cells that are arranged in short intersecting fascicles similar to fibrosarcoma. Pseudoglandular areas can be observed in biphasic cases. Epithelioid sarcoma, though common in the hand, is relatively rare in the foot and is histologically distinct from giant cell tumor of tendon sheath. When this tumor secondarily involves bone, it may be confused with osteomyelitis.


Question 20
Osteofibrous dysplasia and adamantinoma are both rare tumors of bone generally occurring in the tibia. Which of the following histologic findings is typically seen only in adamantinoma?

1. Biphasic pattern of glandular epithelial cells surrounded by spindle cells
2. Bone trabeculae with osteoblastic rimming and fibrous stroma
3. Bone trabeculae without osteoblastic rimming within fibrous stroma
4. Multinucleated giant cells with background stromal cells with similar nuclei
5. Herringbone pattern with pleomorphic cells

PREFERRED RESPONSE: 1

DISCUSSION: A biphasic pattern of glandular epithelial cells surrounded by fibrous spindle cells is the distinguishing histologic feature of an adamantinoma. The other choices are the histologic patterns seen with osteofibrous dysplasia, fibrous dysplasia, giant cell tumor, and fibrosarcoma, respectively.

Question 21
A 7-year-old boy has multiple firm, fixed masses about his knees and extremities. Occasionally he has pain when he bumps his knee or around his proximal legs when he is playing soccer. Radiographs are shown in Figures 21a and 21b. A CT scan of the distal femurs is shown in Figure 21c. What is the next most appropriate step in management?

1. Observation
2. Biopsy of the largest lesions
3. Whole body PET scan
4. Removal of all lesions about the knee
5. Bisphosphonate therapy

PREFERRED RESPONSE: 1

DISCUSSION: The patient’s clinical presentation and imaging studies are diagnostic of multiple hereditary exostoses (MHE). Whereas removal of symptomatic osteochondromas is indicated if symptoms are severe enough, biopsy of the lesions or removal of all of the lesions is not indicated. Currently, a PET scan does not have a defined role in the evaluation of MHE patients. Bisphosphonate therapy currently has no defined role in the treatment of MHE patients. These patients are best observed if asymptomatic, and the development of symptoms or masses that grow after skeletal maturity should be evaluated for possible malignant degeneration of these lesions.

A 24-year-old woman has had a mass on her right shoulder for the past 3 months. The mass is intermittently painful, warm, and swollen, particularly after periods of activity. Rest and ice relieve her symptoms. Radiographs are normal. T₁-, T₂-weighted, and contrast-enhanced MRI images are shown in Figures 22a through 22c. What is the most likely diagnosis?

1. Synovial cell sarcoma
2. Carcinoma metastatic to soft tissue
3. Muscle tear with hematoma formation
4. Abscess
5. Intramuscular hemangioma

PREFERRED RESPONSE: 5

DISCUSSION: The lesion is an intramuscular hemangioma. Imaging characteristics can appear quite aggressive, and sarcoma is often in the differential. Fat is a prominent feature in diagnosing these tumors. In addition contrast aids in demonstrating the vascular channels, particularly in cavernous subtype. No necrosis or rim enhancement is demonstrated and no appreciable mass effect is noted, making malignancy or abscess less likely. A muscle tear with hematoma formation could have a similar appearance, but the time course should have allowed for partial or complete hematoma resolution. Observation is generally the treatment of choice, but if symptoms are not relieved by noninvasive means, surgery, sclerotherapy, and sometimes embolization may have a role.

Question 23
What syndrome is associated with the presence of enchondromas and hemangiomas?

1. Maffucci’s syndrome
2. Hunter’s syndrome
3. Multiple hereditary exostoses
4. Ollier’s disease
5. Trevor’s disease

PREFERRED RESPONSE: 1

DISCUSSION: Maffucci’s syndrome is characterized by the presence of enchondromas and hemangiomas. Ollier’s disease is multiple enchondromatosis. Multiple hereditary exostoses is characterized by the presence of multiple exostoses not associated with skin lesions or endocrine abnormalities. Hunter’s syndrome is a mucopolysaccharidosis and lysosomal storage disease that affects all large joints with pain and decreased range of motion. Trevor’s disease is an epiphyseal osteochondroma.

A 4-year-old boy has a 3-month history of limping and pain in the right tibia. According to his parents, he has frequent night pain. There is no history of weight loss, fevers, or night sweats. Examination reveals that the left tibia is moderately tender to palpation. There is no palpable mass or lymphadenopathy. Radiographs, MRI scans, and biopsy specimens are seen in Figures 24a through 24f. What is the fusion protein frequently associated with this disorder?

1. SYT-SSX
2. ASPL-TFE3
3. EWS-ATF1
4. EWS-FLI1
5. TLS-CHOP

PREFERRED RESPONSE: 4

DISCUSSION: The permeative changes and “onion skin” periosteal reaction with no observable matrix production on radiographs are suspicious for Ewing’s sarcoma. The cells are small, round, and monotonous on low power and have large hyperchromatic nuclei and indistinct cytoplasmic borders on high power, supporting the diagnosis of Ewing’s sarcoma. Molecular analysis confirmed the EWS-FLI1 mutation. The radiographic differential diagnosis includes: osteomyelitis, Langerhans cell histiocytosis, and lymphoma. EWS-ATF1 is associated with clear cell sarcoma, while ASPL-TFE3 is associated with alveolar soft part sarcoma. SYT-SSX and TLS-CHOP are associated with synovial sarcoma and liposarcoma, respectively.

Question 25
A 15-year-old girl is referred for evaluation of a tibial lesion noted on radiographs performed after a twisting injury of the left knee. She was asymptomatic prior to the injury that occurred 3 weeks ago. The pain from the injury has now resolved. Radiographs of the left knee are seen in Figures 25a and 25b. What is the next step in management of this patient?

1. CT
2. MRI
3. Bone scan
4. Needle biopsy
5. Observation

PREFERRED RESPONSE: 5
DISCUSSION: The radiographs reveal a benign-appearing lesion of the proximal tibia metaphysis with a narrow zone of transition, an eccentric position, and a thin rim of reactive bone. The imaging characteristics are most consistent with nonossifying fibroma. These lesions may occur multiply in 8% of patients. Most lesions are eccentric with a “soap bubble” appearance and may have significant cortical thinning. Pathologic fractures may occur. This lesion was discovered as an incidental finding. Because the patient is asymptomatic, no further work-up or treatment is indicated except for observation. Most lesions resolve spontaneously by adulthood.

Question 26
A 13-year-old girl injured her left shoulder playing volleyball. The shoulder was previously asymptomatic. She was referred for evaluation of a left humeral lesion noted on radiographs obtained after the injury. Currently she has returned to playing volleyball and is asymptomatic again. AP and lateral radiographs of the shoulder are seen in Figures 26a and 26b. What is the next most appropriate step in management of this patient?

1. MRI
2. CT
3. Bone scan
4. Needle biopsy
5. Observation

PREFERRED RESPONSE: 5

DISCUSSION: The radiographs reveal an osteochondroma of the left humerus; therefore, no further work-up is needed. The lesion was found as an incidental finding after an injury and since it is asymptomatic, no further treatment is needed except for observation. An MRI scan or a CT scan could be obtained to evaluate the thickness of the cartilaginous cap if a secondary chondrosarcoma is suspected in an enlarging lesion in an adult. These studies could also be obtained to define anatomic relationships if surgery is planned. A bone scan could be used to identify other lesions but would not likely provide useful information for this patient. Needle biopsy would not provide any useful information.

Question 27
A 55-year-old woman with a history of lung cancer who underwent resection 1 year ago now reports thigh pain. Radiographs are shown in Figures 27a and 27b. A CT scan of the chest, abdomen, and pelvis and a bone scan show no other lesions. What is the most appropriate next step in management?

1. Chemotherapy
2. External fixation
3. Reamed intramedullary nail with reamings sent to pathology
4. Biopsy
5. External beam radiation followed by placement of an intramedullary nail

PREFERRED RESPONSE: 4

DISCUSSION: The radiographs show a lytic lesion that is highly suggestive of metastatic cancer in this clinical setting. However, in this patient with no other bone lesion as demonstrated by bone scan, a primary sarcoma or myeloma cannot be excluded. Therefore, biopsy should be done prior to prophylactic stabilization to avoid inadvertently contaminating the entire femur. Sending reamings during the intramedullary nailing is not an acceptable method of performing a biopsy on an indeterminate lesion. Chemotherapy, radiation, nailing, or external fixation should not be performed prior to establishing a diagnosis.

Question 28
Which of the following is the most common soft-tissue sarcoma of the hand?

1. Chondrosarcoma
2. Rhabdomyosarcoma
3. Epithelioid sarcoma
4. Myxoid liposarcoma
5. Hemangiopericytoma

PREFERRED RESPONSE: 3

DISCUSSION: The hand is a rare location for soft-tissue sarcomas. The most common histologic subtypes seen there are epithelioid sarcoma, malignant fibrous histiocytoma, synovial sarcoma, and clear cell sarcoma.

Question 29
A 70-year-old man has a painful leg mass. The mass has been present for many years and has not changed in size. It is exquisitely tender to palpation. MRI scans are seen in Figures 29a through 29c, and a biopsy specimen is seen in Figure 29d. What is the most likely diagnosis?

1. Lipoma
2. Fibrosarcoma
3. Neurofibroma
4. Synovial sarcoma
5. Giant cell tumor of tendon sheath

PREFERRED RESPONSE: 3

DISCUSSION: Exquisite tenderness is a hallmark of benign nerve sheath tumors. The MRI scans show a well-defined soft-tissue mass within the posterior tibial nerve. The histology is consistent with a neurofibroma and characterized by hypocellularity and “rope-like” or wavy collagen bundles. The MRI scan findings are not consistent with a lipoma. The histology is not consistent with fibrosarcoma (characterized by hypercellularity, atypia, and “herringbone” growth pattern), giant cell tumor of tendon sheath (monomorphic stromal cells, hemosiderin-laden macrophages, and giant cells), synovial sarcoma (hypercellular, monophasic or biphasic epithelioid cells), or lipoma (bland lipocytes).

Question 30
A 10-year-old boy has had increasing left knee pain for the past 4 months. AP and lateral radiographs of the left proximal tibia are seen in Figures 30a and 30b. An MRI scan and biopsy specimen are seen in Figures 30c and 30d. What is the most appropriate treatment for this patient?

1. Surgery and chemotherapy
2. Surgery and radiation therapy
3. Radiation therapy and chemotherapy
4. Surgery alone
5. Radiation therapy alone

PREFERRED RESPONSE: 1
DISCUSSION: The imaging studies demonstrate an aggressive-appearing blastic lesion of the proximal tibial metaphysis. This is highly suggestive of osteosarcoma. The biopsy specimen reveals malignant spindle cells that produce osteoid, thus confirming the diagnosis of osteosarcoma. Treatment of osteosarcoma is multimodal including multi-agent chemotherapy and surgery (wide resection or amputation). Radiation therapy is not used in the initial curative treatment of osteosarcoma.


Question 31
A 72-year-old woman reports pain with overhead activity. Examination reveals a prominent mass near the inferior angle of the scapula. A CT scan is shown in Figure 31a and a biopsy specimen is shown in Figure 31b. What is the most likely diagnosis?

1. Lipoma
2. Lymphoma
3. Hemangioma
4. Elastofibroma
5. Giant cell tumor of tendon sheath

PREFERRED RESPONSE: 4
DISCUSSION: Although elastofibromas can occur anywhere in the body, the most common location is the ventral aspect of the scapula. The CT scan shows a mass anterior to the left scapula and overlying the chest wall with strands of low attenuation representing fat. The biopsy specimen shows a pauci-cellular disease process with dense collagen and elastin fibers. The dark staining areas represent elastin. Lipomas generally are homogeneous with low attenuation on CT. Lymphoma has a fairly uniform small round cell morphology microscopically. Hemangiomas may have mineralizations (phleboliths) on radiographs and CT and microscopically reveal multiple capillary or cavernous vascular spaces with intervening fat. On histologic sections, giant cell tumor of tendon sheath is a spindle cell proliferation with interspersed giant cells and hemosiderin-laden macrophages.

Question 32
A 46-year-old man has a 2-month history of an enlarging medial arm mass. Representative MRI scans and a biopsy specimen are shown in Figures 32a through 32c. What is the genetic abnormality associated with this type of soft-tissue tumor?

1. t(11;22)
2. t(X;18)
3. t(12;16)
4. t(2;13)
5. t(12;22)

PREFERRED RESPONSE: 2

DISCUSSION: The images demonstrate a deep, heterogenous mass with histology showing a typical biphasic pattern seen with synovial sarcoma. Synovial sarcoma is characterized by t(X;18), which leads to the formation of a fusion gene between the SYT on chromosome 18 and one of the adjacent genes on the X chromosome, SSX1 or SSX2. Using RT-PCR, it is possible to detect the SYT-SSX fusion transcript. A characteristic t(11;22) occurs in Ewing’s sarcoma. This is the most studied solid tumor chromosome translocation. This rearrangement leads to juxtaposition of a gene, designated EWSR1, on chromosome 22 with FLI1 gene on chromosome 11. Myxoid liposarcoma is characterized by the presence of t(12;16). Most alveolar rhabdomyosarcoma cells exhibit t(2;13). t(12;22) has been associated with clear cell sarcoma.
Question 33

Figures 33a through 33c show the MRI scans and biopsy specimen of a 9-year-old girl who has had progressive swelling and a mass on her medial elbow for 1 month. The area is increasingly painful to touch and with range of motion. The remainder of her examination is unremarkable. What is the next most appropriate step in management?

1. Chemotherapy
2. Radiation therapy
3. Antibiotic therapy
4. Observation
5. Physical therapy

PREFERRED RESPONSE: 3
DISCUSSION: Cat scratch disease (CSD) is typically a benign and self-limited illness lasting 6 to 12 weeks in the absence of antibiotic therapy. Regional lymphadenopathy (axillary, epitrochlear, inguinal) is the predominant clinical feature of CSD; affected nodes are often tender and occasionally suppurate. Between 25% and 60% of patients report a primary cutaneous inoculation lesion (0.5- to 1-cm papule or pustule) at the site of a cat scratch or bite. The skin lesions typically develop 3 to 10 days after injury and precede the onset of lymphadenopathy by 1 to 2 weeks. Bartonella henselae is now regarded as the etiologic agent of CSD. For many years, CSD has been clinically diagnosed when three of the following four criteria are met in a patient: 1) history of traumatic cat contact; 2) positive skin-test response to CSD skin-test antigen; 3) characteristic lymph node lesions; and 4) negative laboratory investigation for unexplained lymphadenopathy. Treatment consists of azithromycin, ciprofloxacin, doxycycline, or multiple other antibiotics, all of which have been used successfully. Radiation therapy and chemotherapy would be reserved for malignant diseases and would not be appropriate in this setting. Treatment is necessary for this infectious entity; therefore, observation or physical therapy is not indicated.

Question 34

Figures 34a through 34d show the AP and lateral radiographs, MRI scan, and biopsy specimen of a 45-year-old man who has had pain for 3 months. Immunohistochemistry shows CD99 negativity. What is the most likely diagnosis?

1. Lymphoma of bone
2. Ewing’s sarcoma
3. Chondrosarcoma
4. Metastatic adenocarcinoma
5. Osteogenic sarcoma

PREFERRED RESPONSE: 1

DISCUSSION: Lymphoma is a destructive lesion of bone, often with a large soft-tissue mass. Histology shows basophilic cells of varying size with little background stroma. Ewing’s sarcoma histology also shows uniform sheets of small blue round cells, but typically occurs in childhood and shows CD99 positivity on immunohistochemistry. Chondrosarcoma demonstrates malignant cartilage, adenocarcinoma demonstrates a glandular arrangement of epithelial cells, and osteogenic sarcoma is a mixed lytic/blastic lesion demonstrating malignant cells producing immature osteoid.
Question 35
A 45-year-old woman has a slowly enlarging mass over the radial aspect of her middle finger at the level of the proximal interphalangeal joint. It is associated with decreased flexion of the joint and clinically is fixed to the underlying bone. Radiographs reveal erosion of the lateral cortex of the proximal phalanx. Gross observation at the time of surgery reveals that the mass has a yellowish-brown tint and lobulated areas. Histology demonstrates bland fibrous stroma with scattered histiocytes, giant cells, and hemosiderin. What is the most likely diagnosis?

1. Epithelioid sarcoma
2. Giant cell tumor of tendon sheath
3. Gouty tophus
4. Hemangioma
5. Epithelial inclusion cyst

PREFERRED RESPONSE: 2

DISCUSSION: The clinical and pathologic description is typical of a giant cell tumor of tendon sheath. Epithelioid sarcoma is the most common soft-tissue sarcoma in the hand and is composed of a nodular arrangement of tumor cells with epithelioid appearance and eosinophilia with a tendency to undergo central degeneration and ulceration. Gouty tophi have a characteristic white, chalky gross appearance and will demonstrate negatively birefringent crystals on polarized light microscopy. Hemangiomas are composed of a variable amount of fat and vessels. Epithelial inclusion cysts are filled with keratin from desquamation of the hyperkeratotic, stratified squamous epithelial cells that line the cysts.

Figures 36a through 36e show the AP and lateral radiographs, axial CT scan, sagittal MRI scan, and biopsy specimen of an 18-year-old man with knee pain. What is the most likely diagnosis?

1. Fibrous dysplasia
2. Osteoblastoma
3. Osteosarcoma
4. Nonossifying fibroma
5. Osteomyelitis

PREFERRED RESPONSE: 2
DISCUSSION: The most likely diagnosis is osteoblastoma, which has a widely varied radiographic appearance. It can often best be described as a large osteoid osteoma (> 1 to 2 cm). It can be lytic or blastic, but is usually well-margined, indicating its benign nature. Histology shows trabecula with intermixed nonmalignant osteoblasts. Osteoid osteoma is a small (< 1 to 2 cm) lesion with thick sclerotic bone surrounding a small lytic nidus. Histology is very similar to osteoblastoma. Osteosarcoma has malignant cells seen on histology. Nonossifying fibroma is an eccentric lytic lesion occurring in the metaphysis and has a bland fibrous background arranged in a storiform pattern with scattered giant cells under the microscope. Osteomyelitis tends to be partially lytic often with a draining sinus or overlying skin changes. Fibrous dysplasia is characterized by ground glass calcification and a Chinese character pattern on histology.

Which of the following is associated with local recurrence following excision of the lesion shown in Figures 37a through 37c?

1. Age of patient  
2. Effectiveness of chemotherapy  
3. Tumor necrosis  
4. Sensitivity to radiation  
5. Type of graft material

PREFERRED RESPONSE: 1

DISCUSSION: The lesion shown in the images is an aneurysmal bone cyst. These lesions are known to have a local recurrence rate of 5% to 50%. Young age, open physes, stage, and type of surgical removal and resulting margin have all been shown to affect the recurrence rate. Chemotherapy and radiation are not used in the treatment of aneurysmal bone cysts. The percentage of necrosis of the lesion is prognostic in osteosarcoma. The type of graft material does not affect local recurrence.

Question 38
Figures 38a through 38c show the radiographs and CT scan of a 24-year-old man who reports tightness in the left knee and decreased range of motion. A biopsy specimen is shown in Figure 38d. What is the most appropriate treatment plan?

1. Radiation therapy and surgical resection
2. Chemotherapy and surgical resection
3. Chemotherapy, radiation therapy, and surgical resection
4. Surgical resection only
5. Observation

PREFERRED RESPONSE: 4
DISCUSSION: Parosteal osteosarcoma is a well-differentiated malignant tumor arising on the surface of the bone. It comprises less than 5% of all osteogenic sarcomas. It is amenable to surgical resection alone. A number of reports have confirmed that parosteal osteosarcoma has a better prognosis than other osteosarcomas. Histologic features are a bland fibroblastic stroma intermixed with well-developed bony trabeculae. Radiation therapy and chemotherapy have no role in the treatment of low-grade parosteal osteosarcoma. Observation is not appropriate for this lesion.


Question 39
A 63-year-old man has had increasing left leg pain over the last several months. History reveals that he has had recurring cyclic pain in the leg for the past several years. Radiographs show an enlarged, sclerotic tibia, with thickened coarse trabeculae and varus bowing. What is the most appropriate management for this patient?

1. Vitamin D
2. Calcium supplement
3. Methotrexate
4. Nonsteroidal antiinflammatory drugs (NSAIDs)
5. Bisphosphonate therapy

PREFERRED RESPONSE: 5

DISCUSSION: Based on the signs and symptoms, Paget’s disease is the most likely diagnosis. In Paget’s disease, an elevated alkaline phosphatase level and high output heart failure may be seen. Hearing loss can be seen when there is involvement of the skull, and malignant degeneration is uncommon but recognized as a risk. Patients are often treated with bisphosphonate medications during the active disease process to help control osteoclastic activity and pain. Vitamin D and calcium are more appropriate for treatment of osteoporosis. Methotrexate is not indicated for the treatment of Paget’s disease. NSAIDs may be helpful to treat pain associated with Paget’s disease but will not alter the clinical course.

Question 40
Radiographs of the right knee of a 21-year-old man are seen in Figures 40a and 40b. What is the inheritance pattern of this disorder?

1. Sporadic
2. Autosomal dominant
3. Autosomal recessive
4. X-linked dominant
5. X-linked recessive

PREFERRED RESPONSE: 2

DISCUSSION: The radiographs show multiple osteochondromas and are thus diagnostic for multiple hereditary exostoses (MHE). MHE is an autosomal-dominant disorder with greater than 95% penetrance. It is associated with mutations of the EXT1 or EXT2 genes. In addition to multiple osteochondromas, affected individuals typically exhibit short stature and angular deformities of the long bones.

Question 41
A 57-year-old woman has a right proximal humerus lesion that was originally discovered when a chest radiograph was obtained. The right shoulder is asymptomatic. An AP radiograph of the right shoulder is seen in Figure 41. What is the next step in management?

1. MRI
2. CT
3. Bone scan
4. Biopsy
5. Observation

PREFERRED RESPONSE: 5

DISCUSSION: The radiograph shows a benign-appearing lesion with stippled calcification. There is no cortical erosion. Equally important is the fact that the patient is asymptomatic. These findings are typical of enchondroma. No further work-up is indicated for this patient except for observation with serial radiographs. If the patient has symptoms, further cross-sectional imaging may be necessary to assess the aggressiveness of the lesion. A bone scan will differentiate monostotic disease from polyostotic disease and the relative uptake may help assess activity. Biopsy is usually not helpful in distinguishing between enchondromas and low-grade chondrosarcomas.

Question 42
Giant cell tumor of the tendon sheath is histologically most closely related to which of the following?

1. Desmoplastic fibroma
2. Dermatofibroma
3. Pigmented villonodular synovitis (PVNS)
4. Myositis ossificans
5. Epithelial inclusion cyst

PREFERRED RESPONSE: 3

DISCUSSION: A giant cell tumor of the tendon sheath consists of multinucleated giant cells, polygonal mononuclear cells, and histiocytes (may contain abundant hemosiderin or lipid). This same cell population is seen in PVNS in addition to hemosiderin. Desmoplastic fibroma is composed of dense and irregularly arranged collagen bundles with infrequent fibroblasts. Dermatofibroma is composed of nodular cellular proliferation consisting of short intersecting fascicles of fibroblastic cells in a loose crisscross or storiform pattern. Myositis ossificans is characterized by zonal proliferation with central fibroblasts and peripheral osteoblast-rimmed bone trabeculae. Epithelial inclusion cysts are filled with keratin from desquamation of the hyperkeratotic, stratified squamous epithelial cells that line the cysts.

Question 43

Figures 43a through 43d show the radiograph, bone scan, CT scan, and biopsy specimen of a 64-year-old woman who reports increasing right hip pain for the past 6 months. Treatment of this lesion consists of which of the following?

1. Surgery alone
2. Chemotherapy alone
3. Radiation therapy alone
4. Surgery and chemotherapy
5. Surgery and radiation therapy

PREFERRED RESPONSE: 1

DISCUSSION: The radiograph and the CT scan show an expansile, destructive lesion of the right acetabulum with stippled calcification. The bone scan shows increased uptake in the area of the lesion. The biopsy specimen reveals hypercellular cartilage, confirming the diagnosis of conventional chondrosarcoma. Treatment consists of surgery alone. In this patient, an internal hemipelvectomy with wide margins would be appropriate. Chondrosarcomas are resistant to both chemotherapy and radiation therapy.


Question 44

Figure 44a shows the lateral radiograph of a 28-year-old-man who has had shin pain for the past 2 years. Sagittal and axial MRI scans are shown in Figures 44b and 44c. A biopsy specimen is shown in Figure 44d. What is the best treatment for this lesion?

1. Observation
2. Curettage and grafting
3. External beam radiation alone
4. Wide resection alone
5. Chemotherapy and wide excision

PREFERRED RESPONSE: 4

DISCUSSION: The imaging showing a bubbly lesion in the anterior cortex of the tibia and the histology showing epithelial nests are consistent with adamantinoma. The best treatment for this lesion is wide resection without adjuvant treatments. Histology demonstrating epithelial cells in a fibrous stroma confirm the diagnosis, and differentiate it from osteofibrous dysplasia, which follows the same anatomic distribution and has an overlapping radiologic appearance. Observation, curettage, and external beam radiation are not indicated for adamantinoma. There is no role for chemotherapy in this low-grade lesion.
Question 45
An 18-year-old woman has had knee pain for the past 6 months. A radiograph and biopsy specimen are shown in Figures 45a and 45b. What is the most appropriate treatment option?

1. Wide resection and reconstruction
2. Radiofrequency ablation
3. Radiation therapy
4. Extended curettage with use of adjuvants
5. Observation

PREFERRED RESPONSE: 4

DISCUSSION: The radiograph and biopsy specimen are characteristic for giant cell tumor of bone. The standard of care involves extended curettage, use of adjuvants to lower the risk of local recurrence, and filling of the void with bone graft and/or polymethylmethacrylate. Multiple adjuvants are acceptable and vary based on surgeon training and preference. These include phenol, liquid nitrogen, sterile water, and argon beam coagulation. Wide resection and skeletal reconstruction should be reserved for extensive bone destruction where salvaging the involved bone is not feasible. Radiation therapy should be used with extreme caution because of the risk of secondary sarcomatous degeneration. In multiply recurrent lesions and more central, hard to access lesions, radiation therapy sometimes can play a role in the management of giant cell tumors.
Question 46
A 50-year-old woman with a recent diagnosis of multiple myeloma is being evaluated for left hip pain and some difficulty with ambulation. She is still receiving her initial systemic management and has extensive skeletal involvement found on her initial skeletal survey and other staging modalities. Once referred for orthopaedic evaluation, an AP radiograph of her pelvis was obtained and is shown in Figure 46. What is the most appropriate surgical option for this patient?

1. Radical resection of the proximal third of the femur and tumor prosthetic reconstruction
2. Cemented hemiarthroplasty or total hip arthroplasty
3. Intramedullary nailing
4. Percutaneous screw fixation
5. Hip screw and side plate fixation

PREFERRED RESPONSE: 2
DISCUSSION: The goal of surgical management of myeloma is the same as it is for bone metastases - to relieve pain and restore function by achieving local tumor control and immediate mechanical stability. Pathologic femoral neck fractures that require surgical intervention should always be treated with removal and hemiarthroplasty or total hip arthroplasty. Bone cement augments structural stability and enables the patient to withstand the stress of immediate motion and function. Internal fixation options are less attractive because they typically do not allow immediate full weight bearing and would require the fracture to heal with the harmful effects of radiation and chemotherapy interfering with the healing process. Radical resection of the proximal femur and the tendinous attachments is not necessary for this patient and would increase the morbidity of the procedures.

Question 47
A 10-year-old boy reports increasing left knee pain. A radiograph, MRI scan, and biopsy specimen are shown in Figures 47a through 47c. What is the most likely diagnosis?

1. Osteoblastoma
2. Chondroblastoma
3. Giant cell tumor
4. Langerhans cell histiocytosis
5. Aneurysmal bone cyst

PREFERRED RESPONSE: 2

DISCUSSION: Chondroblastomas are painful benign tumors that typically occur in patients between the ages of 10 and 25. They almost always occur in an epiphysis or an apophysis. Although they are benign, the MRI scan frequently demonstrates surrounding edema as seen in this patient. The biopsy specimen shows multinucleated giant cells as well as chondroblasts. The chondroblasts are polygonal cells with well-defined cytoplasmic borders and indented nuclei. Osteoblastoma is a bone-forming lesion with interlacing osteoid and fibrovascular stroma similar to osteoid osteoma. Osteoblastoma and osteoid osteoma are differentiated more by size and clinical presentation than histology. Giant cell tumor is composed of multinucleated giant cells with stromal cells sharing similar nuclear morphology. Langerhans cell histiocytosis is a common, usually self-limiting process with a variable radiographic appearance. The lesion is composed of plump histiocytes, eosinophils, and variable inflammatory cells. Aneurysmal bone cysts are not typical in the epiphysis and are composed of lakes of bloods surrounded by variable amounts of fibrous stroma containing giant cells, histiocytes, and hemosiderin.

Figure 48

Question 48
Figure 48 shows the AP and lateral radiographs of a 12-year-old boy with knee pain after a fall. Previous to the fall he denies any history of pain. What is the most likely diagnosis?

1. Unicameral bone cyst
2. Chondroblastoma
3. Fibrous dysplasia
4. Nonossifying fibroma
5. Osteosarcoma

PREFERRED RESPONSE: 4

DISCUSSION: The patient has a nonossifying fibroma. Nonossifying fibroma is classically an eccentric, lytic lesion with a thin sclerotic border in the metaphysis. Unicameral bone cyst is typically central with variable surrounding sclerosis. Chondroblastoma is a lytic lesion in the epiphysis. Fibrous dysplasia is typically a central lytic lesion in the metaphysis or diaphysis with a ground glass appearance. Osteosarcoma is usually a mixed lytic/blastic lesion with bone destruction and formation, without sclerotic margins.

Question 49
The diagnosis of gout can be made either by the presence of tophaceous deposits in the skin or bursae of the extremities or by the presence of which of the following?

1. Elevated urine pH
2. Elevated serum uric acid
3. Calcium pyrophosphate crystals in the synovial fluid
4. Monosodium urate crystals in the synovial fluid
5. Elevated serum phosphate

PREFERRED RESPONSE: 4

DISCUSSION: Gout is an inflammatory arthritis caused by the presence of monosodium urate crystals in the joint. It is characterized acutely by a painful joint that remits after 1 to 2 weeks and recurs periodically. The diagnosis of gout can be made by confirming the presence of monosodium urate crystals in the joint fluid aspirated from the inflamed joint. Patients with gout may also have tophaceous deposits within the skin or bursae of the extremities. Elevated urine pH, serum uric acid, and serum phosphate can all be associated with numerous conditions and are not specific to gout. Calcium pyrophosphate crystals are associated with chondrocalcinosis (pseudogout).

Question 50

A 29-year-old man reports pain in the right little finger. The pain is worse at night. Symptoms were completely relieved with naproxen; however, the patient is no longer able to continue naproxen secondary to gastrointestinal problems. A radiograph is seen in Figure 50a and a CT scan is seen in Figure 50b. What is the best treatment option for this patient?

1. Curettage
2. Observation
3. Acetaminophen
4. Wide resection
5. Radiofrequency ablation

PREFERRED RESPONSE: 1

DISCUSSION: The radiograph and the CT scan demonstrate a sclerotic lesion with a central nidus consistent with osteoid osteoma. The patient’s pain characteristics (worse at night and relieved by anti-inflammatory drugs) are also typical of osteoid osteoma. The best treatment for this patient would be curettage with a power burr which is associated with less than 10% local recurrence. Observation is not a good option for this patient because he is experiencing pain. Studies have demonstrated high levels of cyclooxygenases and prostaglandins in the tumor osteoblasts. This might explain why many patients receive dramatic relief with anti-inflammatory medications. Acetaminophen would not be expected to provide adequate pain relief. Whereas radiofrequency ablation is currently the most common method employed to treat osteoid osteomas, this procedure is contraindicated for lesions in the digits because of the risk of thermal necrosis of the overlying skin as well as the digital neurovascular structures. Wide resection would be overly aggressive for this benign lesion.


**Figure 51**

**Question 51**

A 57-year-old man was treated for an upper extremity high-grade soft-issue sarcoma 9 months ago with resection and postoperative radiation therapy. At his restaging visit, a new solitary lesion is seen on the CT chest scan shown in Figure 51. What is the most effective treatment of this new identified lesion?

1. Wedge resection
2. Pneumonectomy
3. Chemotherapy
4. Radiation therapy
5. Palliative care

PREFERRED RESPONSE: 1
DISCUSSION: For patients with soft-tissue sarcoma, the lungs are the most common site of metastatic disease. Although pulmonary metastases most commonly arise from primary tumors in the extremities, they may arise from almost any primary site or histology. Resection of metastatic disease is the single most important factor that determines outcome in these patients. Long-term survival is possible in selected patients, particularly when recurrent pulmonary disease is resected. Surgical excision of lung metastases from soft-tissue sarcomas is well accepted and should be considered as a first line of treatment if preoperative evaluation indicates that complete resection of the metastases is possible. With an isolated lesion, a wedge resection or lobectomy would be adequate for controlling disease. Pneumonectomy could be used for more extensive disease. Further investigation is needed before chemotherapy can be recommended as additional therapy. Radiation therapy is not typically used for pulmonary metastasis from soft-tissue sarcomas.

Question 52

Figures 52a and 52b show the radiographs of a 30-year-old-man who had Ewing’s sarcoma at the age of 10 treated with radiation therapy for local control. What is the most likely diagnosis?

1- Recurrent Ewing’s sarcoma
2- Osteomyelitis
3- Radiation-induced sarcoma
4- Metastatic adenocarcinoma
5- Bone infarct

PREFERRED RESPONSE: 3

DISCUSSION: The most likely diagnosis is radiation-induced sarcoma, which typically occur years after the radiation is received. The radiographs show a lytic lesion in the area of previously treated Ewing’s sarcoma. Occasionally Ewing’s sarcoma is treated with radiation therapy, typically in locations where limb salvage is not feasible. Recurrent Ewing’s sarcoma would be unusual 20 years after the primary tumor, bone infarct is not purely radiolucent, adenocarcinoma is rare in patients younger than age 40, and osteomyelitis would be an unusual diagnosis in this clinical setting.

Question 53
Based on the radiograph, post-gadolinium enhanced MRI scan, and histologic study shown in Figures 53a through 53c, what is the most likely diagnosis?

1. Undifferentiated pleomorphic sarcoma
2. Synovial sarcoma
3. Liposarcoma
4. Fibromatosis
5. Epithelioid sarcoma

PREFERRED RESPONSE: 4

DISCUSSION: Fibromatosis more frequently occurs in women, with a peak incidence between puberty and age 40 years. On MRI, the tumor is isointense to hypointense with skeletal muscle on both pulse sequences but varies depending on the amount of collagenous stroma; it enhances markedly with gadolinium administration as seen in Figure 53b. Histologically, this tumor is composed of fibroblasts and myofibroblasts in a collagenous matrix with low mitotic activity. Although the tumor recurs frequently (due to its infiltrative growth pattern) despite wide local surgery, it does not metastasize. Chemotherapy using low-dose methotrexate and vinblastine has been reported to be beneficial in treating this tumor. Radiographically, with a few exceptions, it is very difficult to distinguish one soft-tissue tumor from another. Histologically, undifferentiated pleomorphic sarcoma contains spindle malignant cells that have a storiform pattern. Synovial sarcomas contain spindle cells arranged in short intersecting fascicles; they may also have pseudoglandular areas in the biphasic form of disease. Epithelioid sarcoma contains spindled and epithelioid cells arranged in nodules with necrosis and hemorrhage (pseudoangiosarcomatous appearance).

Question 54

The radiographs, bone scan, and MRI scans of a 10-year-old girl are seen in Figures 54a through 54f. A biopsy specimen is seen in Figure 54g. Which of the following represents the treatment option with the best prognosis?

1. Wide resection without adjuvant
2. Methotrexate-based chemotherapy with radiation therapy
3. Methotrexate-based chemotherapy with wide resection
4. Vincristine, doxorubicin, cyclophosphamide, and dactinomycin-based chemotherapy with wide resection, with or without radiation therapy
5. Vincristine, doxorubicin, cyclophosphamide, and dactinomycin-based chemotherapy

PREFERRED RESPONSE: 4

DISCUSSION: The patient has Ewing’s sarcoma. This tumor is best treated with neoadjuvant chemotherapy consisting of a regimen based around vincristine, doxorubicin, cyclophosphamide, and dactinomycin in combination with wide resection or amputation. Radiation therapy may have a role for local disease control with close surgical margins, surgically inaccessible sites, or in the presence of advanced disease. Osteosarcoma is treated with methotrexate-based chemotherapy and wide resection or amputation.

Question 55
A 32-year-old woman has had hip pain for 8 months. Initially, the pain was present with activity but has now progressed to pain also at rest. An AP radiograph of the pelvis, CT scan, and MRI scan are seen in Figures 55a through 55c. Figures 55d and 55e show the biopsy specimens. What is the most appropriate treatment?

1. Observation
2. Curetting and bone grafting
3. Chemotherapy followed by wide excision
4. Radiotherapy
5. Radical resection

PREFERRED RESPONSE: 2
DISCUSSION: Chondromyxoid fibroma is a rare tumor and is most often found in the proximal tibia. Other common sites include the collective bones of the foot and ankle as well as the pelvis. The radiograph demonstrates a radiolucent abnormality in the right ilium that is partially obscured by bowel gas. The CT scan reveals a destructive, low attenuation abnormality with a rim of bone on the periphery and subtle internal matrix. The coronal T2-weighted MRI scan shows a hyperintense signal abnormality with a lobular growth pattern; the transition between the lesion and normal bone is distinct. Low power H&E stained tissue shows a biphasic histologic pattern with hypercellular spindle cells surrounding a relatively hypocellular area. High power reveals very characteristic stellate cells. Treatment for this condition is intralesional (curetting and bone grafting). Local recurrence is reported in 20% to 25% of cases. Chondrosarcoma is in the differential in this case and would require wide local excision. Chondroblastoma is benign, albeit sometimes aggressive and does not respond to adjuvant therapies such as chemotherapy or radiation therapy.


Question 56

A 72-year-old woman is evaluated for sacrococcygeal pain sustained after a twisting injury. Radiographic and MRI evaluation confirms the presence of a nondisplaced fracture at the sacrococcygeal junction. Over a 3-week period, the pain has gotten significantly better. No additional lesions or injuries are noted. Laboratory studies show a serum calcium level of 8.8 mg/dL (normal 8.6-10.3 mg/dL) and a 25-OH Vitamin D level of 14 ng/mL (normal 30-80 ng/mL). What is the most appropriate treatment for this patient?

1. Expectant observation
2. Calcium supplementation
3. High dose vitamin D supplementation
4. Bisphosphonate therapy
5. Surgical fixation of the sacrococcygeal fracture

PREFERRED RESPONSE: 3
DISCUSSION: Chronic Vitamin D deficiency leads to problems with bone health and has been shown to increase the risk of falls in the elderly. Appropriate supplementation of Vitamin D has been shown to decrease this risk. Conversion in the skin decreases with age and may be nearly nonexistent in darkly pigmented individuals. Vitamin D3 is the preferred form for supplementation, but D2 is the form most available by prescription in the US. Hypervitaminosis D is rare and very high doses can be tolerated without significant concern for toxicity. Because the patient has sustained one insufficiency fracture, she is at risk for insufficiency fractures in other skeletal locations, rendering expectant observation insufficient. Her serum calcium is normal, and with a low Vitamin D level, calcium utilization in her system would be inadequate. Bisphosphonate therapy in addition to calcium and vitamin D supplementation may provide a good long-term solution, but should not be instituted until the bone mineral imbalance has been adequately corrected. Surgical fixation of this fracture is not indicated, particularly in lieu of improving symptoms.

Figures 57a through 57c show the radiograph, MRI scan, and photomicrograph of a 13-year-old boy who reports increasing left groin pain. What is the most appropriate method of treatment of this lesion?

1. Surgery alone  
2. Radiation therapy alone  
3. Chemotherapy alone  
4. Chemotherapy and surgery  
5. Surgery and radiation therapy

PREFERRED RESPONSE: 1

DISCUSSION: The radiograph demonstrates an expansile lesion of the left superior pubic ramus. The MRI scan demonstrates multiple fluid-fluid levels consistent with an aneurysmal bone cyst. The photomicrograph demonstrates cavernous hemorrhagic tissue with a benign-appearing spindle cell stroma and giant cells, thus confirming the diagnosis of aneurysmal bone cyst. Surgical management is the recommended treatment for this lesion. Most aneurysmal bone cysts are amenable to curettage and bone grafting. Lesions in expendable bones can be treated with resection. Embolization may decrease intraoperative blood loss when treating large lesions of the pelvis. Radiation therapy has been reported to be effective but is seldom used secondary to the risk of malignant transformation as well as the risk of damage to reproductive organs, physes, and the spinal cord. Chemotherapy has no role in the treatment of this lesion.


Question 58

Figures 58a and 58b show the radiographs of an otherwise healthy 64-year-old man who has had right groin pain for the past 3 months. What is the next most appropriate step in management?

1. Biopsy
2. Observation
3. Radiation therapy
4. Prophylactic fixation of the femur
5. Bone scan, CT scan of the chest/abdomen/pelvis, laboratory studies

PREFERRED RESPONSE: 5

DISCUSSION: The radiographs show a radiolucent lesion in the proximal femur. In a patient older than age 40 years, a new painful bone lesion most likely represents metastatic carcinoma or multiple myeloma even if the patient does not have a known history of cancer. The diagnosis must be firmly established and the patient should be staged prior to initiating treatment. Imaging studies should be completed prior to proceeding with a biopsy. This patient should undergo a bone scan to look for other lesions. A CT scan of the chest, abdomen, and pelvis should be performed to look for a primary tumor as well as other sites of metastases. (Lung or kidney would be the most common primary sites for a patient who presents with metastases of unknown origin.) Serum and urine protein electrophoresis should be obtained to look for multiple myeloma.
A 23-year-old woman who noted 1 day of thigh pain after jogging now reports persistent thigh swelling and can feel a mass. The radiograph, CT scan, and MRI scans are shown in Figures 59a through 59d. What is the most likely diagnosis?

1. Soft-tissue sarcoma
2. Pseudoaneurysm
3. Abscess
4. Osteosarcoma
5. Myositis ossificans

PREFERRED RESPONSE: 5
DISCUSSION: Myositis ossificans (MO) is a reparative lesion that is distinguished by the presence of metaplastic bone formation. The important entity that MO must be distinguished from is extraskeletal osteosarcoma. Extraskeletal osteosarcoma usually occurs in older patients and lacks the zonation phenomenon of MO. Myositis ossificans develops a well-defined ossified rim, maturing peripherally more than centrally, where osteosarcomas do the opposite. Histologically the two may be confusing; therefore, clinical and radiographic evaluation is a critical factor in the diagnosis. Soft-tissue sarcoma, pseudoaneurysm, and abscess do not have zonal ossification patterns.

A 33-year-old man has a painless mass in the anterior thigh. Selected sequences of MRI scans are shown in Figure 60a ($T_1$) and Figure 60b ($T_2$ fat saturated). Biopsy photomicrographs are shown in Figure 60c (low-power) and 60d (high-power). Cytogenetics of the specimen shows a 12:16 translocation. What is the most likely diagnosis?

1. Well-differentiated liposarcoma
2. Myxoid liposarcoma
3. Round cell liposarcoma
4. Fibrolipomatous hamartoma
5. Intramuscular lipoma

PREFERRED RESPONSE: 2

DISCUSSION: The $T_1$-weighted MRI scan shows a heterogeneous fatty tumor within the thigh musculature. The amount of heterogeneity and enhancement are more suggestive of malignancy, but are not diagnostic. The histology shows lipoblasts and primitive mesenchymal cells within a stroma myxomatous tissue with a delicate plexiform capillary network. The cytogenetics demonstrates the characteristic translocation for a myxoid liposarcoma. Treatment is usually consists of wide local excision with or without radiation therapy. Chemotherapy may be considered if there is a significant (25% or greater) round cell component.

Question 61
A 59-year-old man who works as a laborer has had left hip pain for the past 12 months. He reports some worsening with activity and occasional pain that wakes him from sleep. He rates his baseline pain as 3 out of 10. He denies any fevers, malaise, or other systemic symptoms. A radiograph, CT scan, and biopsy photomicrographs are shown in Figures 61a through 61e. Appropriate treatment of this lesion would entail which of the following?

1. Chemotherapy and wide resection
2. Radiation therapy alone
3. Extended curettage and radiation therapy
4. Wide surgical resection alone
5. Observation with repeat scans in 3 months

PREFERRED RESPONSE: 4

DISCUSSION: This is a classic presentation of chondrosarcoma of the pelvis. Wide surgical resection is the treatment of choice. Though chemotherapy may be offered for patients with dedifferentiated chondrosarcoma, the histology of this lesion is consistent with an intermediate to high-grade myxoid chondrosarcoma. Chondrosarcomas are radioresistant, and should be aggressively treated with surgery. Extended intralesional resection of low-grade chondrosarcoma is acceptable in the appendicular skeleton; it is inappropriate in patients with intermediate or high-grade lesions and lesions that occur in the pelvis and other axial sites.

Question 62
Figures 62a through 62c show the radiographs of a 40-year-old man who works as a heavy laborer and has hand pain after sustaining a minor injury. What is the most appropriate treatment for this patient?

1. Biopsy
2. Ray resection
3. Protective splinting followed by observation after fracture healing
4. Protective splinting followed by curettage and bone grafting after fracture healing
5. Acute open reduction and internal fixation combined with curettage and bone grafting

PREFERRED RESPONSE: 4

DISCUSSION: The radiographs are consistent with an enchondroma with pathologic fracture. Allowing the fracture to heal and then proceeding with curettage and bone grafting allows a more limited procedure with satisfactory results compared with acute surgical treatment. Hand enchondromas that have fractured should generally be treated surgically because repeated fractures will typically occur, particularly in a person who works as a laborer. MRI will not add to the diagnostic work-up and the presence of a fracture may confuse the interpretation. Biopsy is not necessary unless aggressive bony changes are seen. Ray resection is generally reserved for bone and soft-tissue sarcomas or other primary malignancies involving the digit.

Question 63
A 47-year-old woman with a history of breast cancer and a recent diagnosis of lung carcinoma with multiple painful biopsy-proven skeletal metastasis now reports progressive weight-bearing pain in her left thigh. Current treatment consists of chemotherapy for the lung mass and radiation therapy. An AP radiograph of the left femur is shown in Figure 63. What is the best option for treatment of her thigh pain?

1. Bisphosphonate therapy
2. Resection and megaprosthetic reconstruction
3. Prophylactic rodding
4. Radiation therapy
5. Chemotherapy

PREFERRED RESPONSE: 3

DISCUSSION: Predicting which patients will sustain a fracture with metastatic bone disease is a difficult clinical decision. Mirel’s rating system is a useful clinical tool but must be used cautiously. Lower extremity peritrochanteric, large, radiolucent, and painful lesions are at highest risk for fracture. The ultimate goal is palliative in nature and designed to limit pain, minimize time in the hospital, and improve the quality of life in these patients with limited survival time. If a high risk lesion is treated with radiation therapy first, the bone will become weaker before starting to regain structural integrity and the fracture risk increases. Timing of prophylactic fixation, radiation therapy, and chemotherapy requires a multidisciplinary team of doctors working collaboratively. Bisphosphonates should be included in the medical treatment for most patients with metastatic bone disease to lower the risk of further skeletal complications. They are not indicated for acute treatment of impending fractures. Resection of a metastatic lesion is reserved for patients in which internal fixation devices will not have adequate bone stock present to allow stabilization and immediate weight bearing or in patients with selected isolated metastases, such as those caused by renal carcinoma. Radiation therapy and chemotherapy have already failed to control progressive bone destruction and pain in this patient.

Question 64
A 45-year-old man has an enlarging, painless soft-tissue mass in his arm. An MRI scan shows a 15 x 8 x 6 cm enhancing, heterogenous deep mass. Biopsy shows a sarcoma. What is the next most appropriate imaging study to obtain?

1. PET scan
2. Bone scan
3. Chest CT
4. Abdomen CT
5. MRI of the chest

PREFERRED RESPONSE: 3

DISCUSSION: Staging takes into consideration the histologic grade, size, depth, and presence of metastasis of a tumor. Locoregional lymph node spread is uncommon for most sarcomas and is evaluated by palpation; suspicious areas require further imaging to assess. The chest is the most common area for soft-tissue sarcomas to metastasize and should be considered for imaging prior to biopsy to assist with the staging of the patient. In some specific subtypes (myxoid liposarcoma), bone scans and abdominal imaging are important to obtain to exclude metastases. The role for PET scans in patients with soft-tissue sarcomas is still being defined.

A 20-year-old collegiate field hockey player has had diminished knee flexion for the past 3 years. Lately she has noted right knee pain that requires her to stop playing. Radiographs are shown in Figures 65a and 65b. What is the next best step in management?

1. Radiation therapy
2. Surgical biopsy
3. Chemotherapy
4. Knee arthroscopy
5. Bisphosphonates

PREFERRED RESPONSE: 2

DISCUSSION: The lesion has the characteristic features of a parosteal osteosarcoma, including its surface location on the posterior distal femur, decreased range of motion, and pain. Biopsy is necessary to confirm a diagnosis before recommending treatment. Chemotherapy and radical resection may be indicated in dedifferentiated disease, but should be considered only after a biopsy and appropriate staging have been completed. There is no role for knee arthroscopy or bisphosphonates.


Question 66
Which of the following genes is involved in the etiology of aneurysmal bone cyst?

1. ASPL
2. ATF1
3. EWS
4. PAX3
5. USP6

PREFERRED RESPONSE: 5

DISCUSSION: Aneurysmal bone cyst was recently shown definitively to be a neoplasm driven by upregulation of the ubiquitin-specific protease USP6 (Tre2) gene on 17p13 when combined by translocation with a promoter pairing. The most commonly described translocation is t(16;17)(q22;p13) which results in juxtaposition of the promoter region CDH11 on 16q22. In the past, all aneurysmal bone cysts were thought to be reactive in nature. Whereas primary aneurysmal bone cysts are now known to be neoplasms, secondary aneurysmal bone cysts are not because no translocation has been identified in them. ASPL is involved in alveolar soft part sarcoma (tX;17)(TFE3-ASPL). ATF1 is involved in clear cell chondrosarcoma. EWS is a gene on chromosome 22 involved in several relevant musculoskeletal oncology conditions, including Ewing's sarcoma (t11;22)(EWS-FLI1), desmoplastic small round cell tumor (t11;22)(EWS-WT1), extraskeletal myxoid chondrosarcoma (t9;22)(EWS-CHN), and clear cell chondrosarcoma (t12;22)(EWS-ATF1). PAX3 is involved in alveolar rhabdomyosarcoma (t2;13)(PAX3-FKHR).


Question 67

A 39-year-old man has multiple bone lesions in his right leg, bowing deformity, and limb-length inequality. Radiographs are shown in Figures 67a through 67c. A biopsy specimen is shown in Figure 67d. When counseling the patient regarding his diagnosis, how would you explain the inheritance pattern for his disease?

1. Autosomal dominant
2. Autosomal dominant with variable penetrance
3. Autosomal recessive
4. Sex-linked recessive
5. No known inheritance pattern

PREFERRED RESPONSE: 5

DISCUSSION: The patient’s radiographs show multiple lytic bone lesions with some femoral deformity. The radiographic differential would include fibrous dysplasia, enchondromatosis (Ollier’s disease), eosinophilic granulomatosis, metastatic disease, multiple myeloma, hyperparathyroidism (brown tumors), and infection. The pathology demonstrates relatively acellular cartilaginous tissue, supporting the diagnosis of Ollier’s disease. Commonly, patients with enchondromatosis have problems with shortened and/or bowing of their affected limbs. Additionally, unlike solitary enchondromas which have a very low risk of malignant transformation (< 1%), patients with enchondromatosis have a significantly higher risk of malignant transformation, up to 25%. Despite the disease and lesions being present from birth, there is no described inheritance pattern associated with the disease.

Question 68

A 10-year-old boy has a 5-month history of pain in the left thigh and knee that has increased in severity such that he is currently unable to walk secondary to pain. He has a large fusiform swelling about the distal femur. A radiograph of both knees is shown in Figure 68a, and MRI scans are shown in Figures 68b through 68d. After complete staging, a biopsy is performed and a specimen is shown in Figure 68e. What is the most likely diagnosis?

1. Osteomyelitis
2. Stress fracture
3. Osteoblastoma
4. Osteosarcoma
5. Ewing’s sarcoma

PREFERRED RESPONSE: 4

DISCUSSION: The patient’s clinical presentation is one of progressive pain. His radiographs show a destructive, bone-producing lesion that has extended outside the bone into the soft tissues. The pathology shows the production of osteoid by malignant spindle stromal cells. This is consistent with osteosarcoma. There is no inflammatory component of the histology to suggest osteomyelitis. Whereas osteoblastoma can be confused with osteosarcoma, the histology of malignant stromal cells as well as the radiographic findings are classic for osteosarcoma. Ewing’s sarcoma can be associated with a significant soft-tissue mass, as seen in the MRI scan; however, histology would demonstrate a small blue round-cell tumor.

Question 69
A 65-year-old woman has a bone mineral density t-score of -2.0. She has no identifiable secondary cause of bone loss. In addition to calcium and vitamin D supplementation, this patient should be considered for

1. calcitonin.
2. hormone replacement therapy.
3. teriparatide.
4. bisphosphonate therapy.
5. observation.

PREFERRED RESPONSE: 4

DISCUSSION: A patient with a bone mineral density t-score of between -1.0 and -2.5 should be considered for osteoporosis prevention therapy. This would consist of calcium (1,200 to 1,500 mg daily), vitamin D (800 to 1000 IU daily), and a bisphosphonate. Calcitonin and teriparatide are used to treat established osteoporosis (t-score of -2.5 or lower). Observation is not recommended with a t-score of less than -1.0. Hormone replacement therapy is associated with an increased risk of cardiovascular disease and breast cancer.

Question 70

Figures 70a and 70b show the radiograph and MRI scan of a 66-year-old man who has fatigue, weight loss, and muscle weakness. Examination reveals marked pain and discomfort in the left mid leg. Biopsy specimens are shown in Figures 70c and 70d. What is the most likely diagnosis?

1. Mastocytosis
2. Multiple myeloma
3. Hyperparathyroidism
4. Metastatic carcinoma
5. Multicentric giant cell tumor

PREFERRED RESPONSE: 3
DISCUSSION: The signs and symptoms of hyperparathyroidism are similar to those in patients with diffuse skeletal metastases. Serum markers are very helpful in making the diagnosis. In this patient, the radiograph shows multiple lesions in the tibia and proximal fibula that have a variable appearance. For example the mid-tibial lesion is radiolucent and slightly expansile whereas the more proximal tibial lesions are radiodense. The proximal fibula lesion is mixed (radiolucent/radiodense). These findings would be very uncommon in patients with myeloma, metastatic disease, or multicentric giant cell tumor. The histopathology shows a bland fibrous stroma with multiple multinucleated giant cells. On higher power, the stromal cells are spindled and the giant cells are relatively small in contrast to giant cell tumor where the giant cells are larger and the stromal cells are more rounded with nuclei that closely resemble those in the giant cells. There is blood extravasation (stromal hemorrhage) and hemosiderin deposition. The constellation of findings is most consistent with brown tumors due to hyperparathyroidism (secondary to a parathyroid adenoma in this patient).


Question 71
A 76-year-old woman has a painful, snapping mass at the inferior angle of the scapula. She has no history of trauma to the area. MRI scans are shown in Figures 71a through 71c. A biopsy specimen is shown in Figure 71d. What is the most appropriate treatment at this time?

1. Observation
2. Excision
3. Radiation therapy alone
4. Chemotherapy followed by wide excision
5. Local steroid injection

PREFERRED RESPONSE: 2

DISCUSSION: Elastofibroma is a benign, likely inflammatory mass that typically occurs deep to the inferior angle of the scapula. It may be bilateral and may be symptomatic with either pain or mechanical symptoms related to its location. The recommended treatment for symptomatic lesions is excision. Small or asymptomatic lesions may also be observed. Biopsy to confirm the diagnosis and to exclude sarcoma seems prudent. Neither radiation therapy or chemotherapy are indicated for this benign tumor. Local steroid injection plays no role in the treatment of benign elastofibroma.

Question 72
Figures 72a and 72b show the radiograph and biopsy specimen of a 92-year-old woman who reports increasing right shoulder pain. What is the most likely diagnosis?

1. Enchondroma
2. Juxtacortical chondroma
3. Conventional chondrosarcoma
4. Dedifferentiated chondrosarcoma
5. Mesenchymal chondrosarcoma

PREFERRED RESPONSE: 4

DISCUSSION: The radiograph shows a lesion of the proximal humerus with stippled calcification suggestive of a cartilaginous neoplasm. The lesion has eroded through the cortex, and therefore, most likely represents a chondrosarcoma. The biopsy specimen reveals a high-grade spindle cell sarcoma adjacent to low-grade cartilage and is thus diagnostic of dedifferentiated chondrosarcoma. An enchondroma is a benign cartilaginous tumor contained entirely within the medullary cavity. A juxtacortical chondroma is a benign cartilage tumor on the surface of a bone. Conventional chondrosarcoma could present a radiographic appearance similar to this case; however, it would not contain a spindle cell component. Mesenchymal chondrosarcoma typically has small round blue cells and vascular proliferation with a hemangiopericytomatous pattern.


Question 73
An otherwise healthy 30-year-old woman with no history of malignancy reports a 1-year history of right hip and low back pain. A radiograph, CT scan, and MRI scan are shown in Figures 73a through 73c. A biopsy specimen is shown in Figure 73d. What is the most likely diagnosis?

1. Aneurysmal bone cyst
2. Hemangioma of bone
3. Sarcoid
4. Paget’s disease of bone
5. Chondrosarcoma

PREFERRED RESPONSE: 2

DISCUSSION: The patient has a lytic lesion in her right ilium with coarsened trabeculae. The radiographic differential for such a lesion would include aneurysmal bone cyst, hemangioma of bone, sarcoid, Paget’s disease of bone, and fibrous dysplasia. The photomicrograph of the biopsy demonstrates small vascular channels lined with a single layer of endothelial cells, supporting the diagnosis of hemangioma of bone. There are no giant cells typically seen in an aneurysmal bone cyst and the early phases of Paget’s disease. There are no granulomas to suggest sarcoid, and the histopathologic features are not consistent with cartilage. Most patients with hemangiomas require no treatment. Lesions causing symptoms are best treated with intralesional excision.

Question 74
A 56-year-old woman has a 5-month history of a rapidly growing mass in the posteromedial aspect of the right leg. A clinical photograph, MRI scan, and biopsy specimen are shown in Figures 74a through 74c. What is the most appropriate treatment for this patient?

1. Observation
2. Wide resection alone
3. Radiation therapy alone
4. Wide resection and radiation therapy
5. Debridement and antibiotics

PREFERRED RESPONSE: 4
DISCUSSION: The clinical photograph shows an aggressive tumor growing through the skin (fungating). The axial T\textsubscript{1}-weighted MRI scan with contrast shows a large mass in the posterior and proximal leg with heterogeneous enhancement and a large nonenhancing center that suggests tumor necrosis, two poor prognostic factors in disease outcome. Finally, the histopathology suggests an undifferentiated pleomorphic spindle cell neoplasm. This is a high-grade sarcoma that requires both radiation therapy and wide excision to local disease control. The histopathology is characteristic of a high-grade sarcoma, not a local infection. The role of chemotherapy for most adult soft-tissue sarcomas remains controversial.


Question 75
An otherwise healthy 12-year-old boy has ankle pain after being kicked while playing soccer. Radiographs are shown in Figures 75a and 75b. Examination reveals tenderness to palpation, but not with weight bearing. He had no pain preceding the incident. What is the next most appropriate step in management?

1. Observation
2. Curettage and grafting
3. Wide surgical resection
4. Whole body bone scan
5. Chemotherapy

PREFERRED RESPONSE: 1
DISCUSSION: The lesion shown is a nonossifying fibroma. With no pain preceding the traumatic episode and painless weight bearing, the lesion does not appear at risk for fracture. Simple observation with repeat radiographs at a time interval (3 to 6 months) to document stability is sufficient. Surgical intervention is unnecessary because the risk of fracture is low and the natural history is one of spontaneous regression during adolescence. The lesion is benign; therefore, chemotherapy is not indicated. While a bone scan may provide some useful information, it is unnecessary for the diagnosis and adds little to management decisions.


Question 76

A 31-year-old patient has had a left medial elbow mass for 1 month. The mass has been increasing in size and has now become very painful and erythematous. MRI scans are shown in Figures 76a and 76b. Laboratory studies show an erythrocyte sedimentation rate of 49 mm/h (normal 0 to 20 mm/h) and C-reactive protein level of 23 mg/L (normal 0 to 0.3 mg/L). Histology showed lymphoid tissue and multiple necrotizing granulomas. What organism is responsible for this clinical picture?

1. Borrelia burgdorferi
2. Trichophyton tonsurans
3. Bartonella henselae
4. Mycobacterium avium
5. Corynebacterium minutissimum

PREFERRED RESPONSE: 3
DISCUSSION: Cat scratch disease (CSD) is an important diagnosis for the orthopaedic surgeon to consider in the differential diagnosis of soft-tissue masses adjacent to epitrochlear or cervical lymph nodes. It is a soft-tissue tumor simulator and a high index of suspicion is necessary in all patients with upper extremity or head and neck adenopathy and a history of cat exposure. Although generally not required for diagnosis, cross-sectional imaging will reveal a mass with surrounding edema in an area of lymphatic drainage. A peripheral blood sample can be tested for Bartonella henselae - the offending organism with this diagnosis. Classically the histology of these lesions when biopsied will show multiple necrotizing granulomas. Mycobacterium avium is the only other organism that would demonstrate a granulomatous reaction and the location is classic for CSD. Borrelia burgdorferi is associated with Lyme disease. Mycobacterium avium may be a source of immunocompromised infections in HIV patients. Trichophyton tonsurans and corynebacterium minutissimum are not associated with orthopaedic diseases.


Question 77
A 45-year-old woman has a painful mass in the dorsum of the right wrist. It is firm and nontender to palpation. She states it has slowly gotten bigger over the past 3 years. You suspect a dorsal wrist ganglion. What is the most definitive way to confirm this diagnosis?

1. Observe it for 1 year to see if it changes dramatically in size.
2. Obtain a gadolinium enhanced MRI scan.
3. Obtain radiographs, looking for scapholunate joint degenerative changes.
4. Perform a needle aspiration and send the aspirate for cytologic examination.
5. Apply direct firm manual pressure over the mass to see if it can be ruptured.

PREFERRED RESPONSE: 4

DISCUSSION: Dorsal wrist ganglions are synovial cysts that arise most frequently from the scapholunate joint. They often extend between the extensor digitorum communis and extensor pollicis longus tendons at the wrist. Aspiration of the cyst is both oncologically safe if done appropriately and also the easiest way to definitively confirm the diagnosis. Clear, yellow viscous fluid/gel is most often aspirated. Cytologic evaluation is mandatory to exclude myxoid neoplasms. Because the lesion has been present for 3 years, further observation is not warranted. The classic presentation, physical examination findings, and location make MRI and radiographs unnecessary. Manual rupture of the mass is not recommended.

Question 78
A 68-year-old woman has had progressive pain in the right thigh for the past several months. She has a history of hypertension, treated with hydrochlorothiazide and osteoporosis treated with alendronate for 10 years. At this point, she is virtually wheelchair bound. Radiographs are shown in Figures 78a and 78b. Additional studies show no signs of systemic disease. What is the most likely etiology of her condition?

1. Prolonged use of bisphosphonates
2. Use of calcium-wasting diuretics
3. Occult metastatic cancer
4. Vitamin D-resistant rickets
5. Disuse osteopenia

PREFERRED RESPONSE: 1

DISCUSSION: The patient has been on alendronate for 10 years and has evidence of a proximal diaphyseal fatigue fracture. These have been associated with long-term use of bisphosphonates. Staging studies have failed to show systemic disease, and while metastasis with an unidentifiable primary does occur, it would be unlikely to present with this radiographic appearance, now recognized to be classic for stress fractures associated with chronic bisphosphonate usage. Hydrochlorothiazide does not cause calcium wasting. Vitamin D-resistant rickets would be a long-standing event and would present much earlier in life, often with pronounced deformities. Whereas the patient’s progression to intolerance of weight bearing likely has led to some degree of disuse osteopenia, the underlying problem is the long-term bisphosphonate exposure.
Question 79

A 32-year-old man reports pain, and examination reveals swelling and tenderness about the knee and distal femur. A radiograph and an MRI scan are shown in Figures 79a and 79b. A bone scan is shown in Figure 79c, and a biopsy specimen is shown in Figure 79d. Cytogenetic analysis of the biopsy specimen failed to show an 11:22 chromosomal translocation. What is the most likely diagnosis?

1. Ewing’s sarcoma
2. Bacterial osteomyelitis
3. Tuberculous osteomyelitis
4. Lymphoma of bone
5. Osteosarcoma

PREFERRED RESPONSE: 4

DISCUSSION: The biopsy specimen reveals sheets of blue cells and therefore falls within the differential of blue cell tumors. The majority of Ewing’s sarcomas will have the 11:22 translocation present. The pathology does not reveal the mixed cell inflammatory process seen in osteomyelitis nor does it show the caseating granulomas of tuberculosis. There are no findings of any osteoid being produced by malignant-appearing spindle cells typical of osteosarcoma. Malignant lymphomas of bone must be considered within the differential for sclerotic lesions of bone with soft-tissue masses as in this patient.

Question 80
An 18-year-old woman has pain and swelling in her lateral hindfoot. On examination, she has significant pain with resisted eversion of the foot. MRI scans are seen in Figures 80a (T1) and Figure 80b (non-fat T2), and a biopsy specimen is seen in Figure 80c. What is the most appropriate treatment for this patient?

1. Marginal excision
2. Transtibial amputation
3. Injection of a radioisotope
4. Chemotherapy and radiation therapy
5. Wide local excision and radiation therapy

PREFERRED RESPONSE: 1
DISCUSSION: The lesion has a stalk that originates in the subtalar joint, fills the sinus tarsi, and effaces the peroneal tendons. Soft-tissue masses that are periarticular should arouse suspicion for synovial sarcoma. Unlike synovial sarcoma however, this lesion was hypointense on both T1 and T2 MRI pulse-weighted sequences because of the large amounts of hemosiderin deposition, characteristic of pigmented villonodular synovitis. Furthermore, intra-articular synovial sarcomas are very rare. Radioisotopes are not very effective for pigmented villonodular synovitis in the foot and ankle because contiguous involvement of multiple joints is not uncommon. Furthermore, skin necrosis can occur with extravasation of radioisotope into the soft tissue. For this reason, complete (marginal) excision is recommended. For recurrent tumors, combined surgery and external beam irradiation has been advocated by some investigators. This is a benign tumor; therefore, aggressive surgical procedures (amputation and wide excision) are not appropriate. Radiation therapy may be a consideration in patients with recurrent and destructive disease.

Question 81

A 26-year-old man has a 1-year history of worsening heel pain. There is no history of penetrating injuries to the heel. A radiograph, bone scan, and MRI scan are shown in Figures 81a through 81c, and biopsy specimens are shown in Figures 81d and 81e. Management should consist of which of the following?

1. Curettage with or without local adjuvants and bone grafting
2. Injection of corticosteroids
3. Irrigation, debridement, and antibiotic treatment
4. Wide resection
5. Chemotherapy and wide resection

PREFERRED RESPONSE: 1

DISCUSSION: With an epiphyseal bone lesion, the radiographic differential would consist of chondroblastoma, Brodie’s abscess, or giant cell tumor of bone. Because of the location in the calcaneus, consideration should also be given for an epidermal inclusion cyst if there is a history of penetrating trauma. The biopsy specimens show a very cellular and vascular lesion with mononuclear chondroblasts with grooved nuclei (“coffee-bean nuclei”). There are some osteoblast-like giant cells. The surrounding chondroid matrix stains pink and has scant areas of fine mineralization outlining the stromal cells (“chicken-wire”). This is characteristic of a chondroblastoma. The preferred treatment for a chondroblastoma in a nonexpendable bone is intralvesional curettage with or without local adjuvants (phenol, liquid nitrogen, argon beam coagulation) and bone grafting (or cement). Some very aggressive chondroblastomas with soft-tissue extension, particularly those in the pelvis, may require en-bloc resection.

Question 82
An 11-year-old boy has a 6-month history of groin pain and a limp. A radiograph is shown in Figure 82a and a biopsy specimen is shown in Figure 82b. What is the etiology of the lesion?

1. Viral infection
2. G(s) alpha mutation
3. t(11;22)
4. t(X;18)
5. Posttraumatic

PREFERRED RESPONSE: 2

DISCUSSION: Fibrous dysplasia is a common benign skeletal lesion that may involve one bone (monostotic) or multiple bones (polyostotic) and occurs throughout the skeleton with a predilection for the long bones, ribs, and craniofacial bones. The etiology of fibrous dysplasia has been linked to an activating mutation in the gene that encodes the alpha subunit of stimulatory G protein (G(s)alpha) located at 20q13.2-13.3. The etiology for Paget’s disease of bone is still unknown but growing evidence shows a possible link to a viral infection. t(11;22) is most commonly seen with Ewing’s sarcoma and t(X;18) with synovial sarcoma.

Question 83
What is the most common complication of total knee arthroplasty in patients with Paget disease?

1. Pain
2. Joint arthrosis
3. Pathologic fracture
4. Malalignment
5. Neurologic compromise

PREFERRED RESPONSE: 4

DISCUSSION: Paget disease affects 3% to 4% of the population in the US older than age 40 years. Urinary hydroxyproline and alkaline phosphatase are elevated in the lytic phase of disease. Insertion mutations in the TNFRSF11A gene encoding RANK are identified. The most common complication of total knee arthroplasty in patients with Paget disease is malalignment. The most frequent complication of hip surgery is hemorrhage. Joint arthrosis, pathologic fracture, and rarely neurologic compromise are sequelae of the disease process itself.

Question 84
A 56-year-old woman has an 8 x 6 x 5 cm intramuscular anterior thigh soft-tissue mass. Biopsy shows a grade 3 undifferentiated pleomorphic sarcoma. A CT of the chest reveals no evidence of other lesions. According to the American Joint Commission on Cancer (AJCC) staging system, what is the stage of this tumor?

1. II-T1a
2. II-T1b
3. II-T2a
4. III
5. IV

PREFERRED RESPONSE: 4

DISCUSSION: The AJCC staging system for soft-tissue sarcomas is based on tumor grade, size, depth, and the presence of metastases. Stage I tumors are low grade. Stage II tumors are high grade. Grade is considered high grade for G3 or G4 tumors on a four-tier grading system and for G2 or G3 on a three-tier grading system. Size is designated by T1 for a size of less than or equal to 5.0 cm or T2 for a size of greater than 5.0 cm in maximal dimension. An “a” or “b” designation immediately follows the size designation to distinguish between superficial (a) and deep (b) tumors. The “deep” designation applies to tumors that involve or are deep to the fascia. Stage III tumors are high grade, deep, and large (eg, T2b). Stage IV tumors include either N1 (nodal) or M1 (distant) metastases regardless of grade. Hence, stage II-T1a would refer to a high grade small superficial sarcoma. Stage II-T1b is a high grade small deep sarcoma. Stage II-T2a is a high grade large superficial tumor. The tumor presented in this case is high grade (G3 or G4), large (>5.0 cm), intramuscular (deep to the fascia), and without metastases. Hence, it is a stage III tumor.

Question 85

Based on the radiographic findings and biopsy specimen shown in Figures 85a and 85b, what is the most likely diagnosis?

1. Ewing’s sarcoma
2. Osteofibrous dysplasia
3. Chondromyxoid fibroma
4. Osteoid osteoma
5. Adamantinoma

PREFERRED RESPONSE: 5

DISCUSSION: Adamantinoma is a low-grade malignant tumor with epithelial differentiation (cytokeratin positive). It can occur in other bones but is most common in the tibia. Cytokeratin is positive in most cases. Tumor metastases occur in up to 20% of patients. There is an association between adamantinoma and osteofibrous dysplasia. Ewing’s sarcoma is a non-matrix producing medullary-based tumor associated with an aggressive periosteal reaction. Histologically it is a small round blue cell tumor. Osteofibrous dysplasia (OFD) is radiographically very similar to adamantinoma because both are cortically based and most common in the tibia. Histologically OFD is composed of bone trabeculae arranged as “chinese letters” with prominent osteoblastic rimming. There are no nests of epithelioid cells. Chondromyxoid fibroma is common in the anterior proximal tibia. It is medullary-based with subtle mineralization and causes thinning of the corticies. Osteoid osteoma is also most commonly periosteally or cortically based but is generally small (< 1cm) and has a sclerotic border.


Question 86
Figures 86a and 86b show the radiograph and biopsy specimen of a 16-year-old boy who reports increasing right foot pain. What is the most likely diagnosis?

1. Enchondroma  
2. Giant cell tumor  
3. Chondrosarcoma  
4. Chondromyxoid fibroma  
5. Nonossifying fibroma

PREFERRED RESPONSE: 4

DISCUSSION: The radiograph shows a benign-appearing bubbly radiolucent lesion with a narrow zone of transition and a thin rim of surrounding reactive bone. The biopsy specimen demonstrates spindle cells with a myxoid cartilaginous matrix. The diagnosis of the lesion is a chondromyxoid fibroma. This rare, benign tumor is usually painful and can be locally aggressive. Enchondroma is typically associated with calcifications and a bland hyaline cartilage on histology. Giant cell tumors are typically radiolucent without the bubbly appearance seen here and composed of multinucleated giant cells with stromal cells having similar nuclear morphology. Chondrosarcoma is uncommon in the hands and feet. Nonossifying fibromas can have a similar radiographic appearance but are histologically distinguished by a storiform pattern of bland fibroblasts with scattered giant cells.

Question 87

Figures 87a through 87c show the AP radiograph and coronal and axial MRI scans of a 50-year-old woman who has had right shoulder pain with overhead activity for the past 6 months. What is the most appropriate treatment of this lesion?

1. Observation with serial radiographs
2. Biopsy
3. Curettage and grafting
4. Wide resection
5. Chemotherapy followed by wide resection

PREFERRED RESPONSE: 1

DISCUSSION: The images show a mineralized lesion in the metaphysis of the proximal humerus, with no cortical disruption or endosteal scalloping. The mineralization pattern is suggestive of a hyaline cartilage neoplasm. There is no soft-tissue mass on the MRI scans. This is consistent with a benign enchondroma. Observation is the treatment of choice for the lesion. Biopsy is not indicated and would be difficult to interpret. The other treatments listed are for low- or high-grade chondrosarcomas. Other explanations for the patient’s shoulder pain, such as rotator cuff pathology or impingement, should be sought and treated.

Question 88
A 51-year-old woman has shoulder pain after a minor fall. A radiograph, MRI scan, and bone scan are seen in Figures 88a through 88c. Biopsy specimens are seen in Figures 88d and 88e. What is the most likely diagnosis?

1. Osteosarcoma
2. Enchondroma
3. Fibrous dysplasia
4. Chondrosarcoma
5. Chondromyxoid fibroma

PREFERRED RESPONSE: 4
DISCUSSION: The radiograph shows an expansile, calcified tumor of the proximal humerus. The MRI scan shows an associated soft-tissue mass and the bone scan demonstrates increased uptake, although these findings could be associated with a pathologic fracture. Pathology is consistent with chondrosarcoma. Enchondroma could present with a similar pattern but should generally be smaller, less expansile, and have no soft-tissue extension; it is also less likely to be associated with a pathologic fracture. Additionally, the pathology would show more benign-appearing cartilage. Chondromyxoid fibroma is more typically eccentric and is associated with radiologic and pathologic findings consisting of benign chondroid, fibrous, and myxoid elements. The radiograph and pathology are not consistent with osteosarcoma or fibrous dysplasia.


Question 89
Based on the lesion seen in Figure 89a and the biopsy specimen seen in Figure 89b, what is the most likely diagnosis?

1. Schwannoma
2. Nodular fasciitis
3. Lipoma
4. Hemangioma
5. Synovial sarcoma

PREFERRED RESPONSE: 1
DISCUSSION: The most reliable sign in diagnosing schwannoma is the percussion sign (radiating pain in the affected nerve distribution on percussion of the lesion). Although sensory deficits are reported, motor weakness is rare. The sagittal MRI scan demonstrates both the string (attenuation of the nerve above and below the tumor) and target signs (circular low signal area within the mass). The biopsy specimen shows the palisading nuclei (Verocay bodies) typically seen in this disease. Nodular fasciitis is a benign tumor that arises in the deep fascia. It is fairly homogeneous and hyperintense on T2 pulse-weighted images and isointense to hypointense on corresponding T1 images. Lipoma is homogeneous bright on both T1 and T2 pulse-weighted images. Hemangioma is usually intramuscular and heterogeneous because of vessel thrombosis and mineralization and adjacent interspersed fat. Synovial sarcoma is more often periarticular and located within muscle. It too can be very heterogeneous because of its cellularity, intralesional necrosis, collagenization, and mineralization.


Question 90
What syndrome is characterized by polyostotic fibrous dysplasia, cafe-au-lait spots with serrated borders, endocrine abnormalities, and unilaterality?

1. Jaffe-Campanacci syndrome
2. Hunter’s syndrome
3. Maffucci’s syndrome
4. Multiple hereditary exostoses
5. McCune-Albright syndrome

PREFERRED RESPONSE: 5

DISCUSSION: This is a description for McCune-Albright syndrome. Jaffe-Campanacci syndrome is characterized by multiple nonossifying fibromas, cafe-au-lait spots, and the absence of neurofibromas. Hunter’s syndrome is a mucopolysaccharidosis and lysosomal storage disease that affects all large joints with pain and decreased range of motion. Maffucci’s syndrome is associated with multiple hemangiomas and enchondromas. Multiple hereditary exostosis is characterized by the presence of multiple exostoses and is not associated with skin lesions or endocrine abnormalities.

Lichenstein L, Jaffe HL. Fibrous dysplasia of bone: A condition affecting one, several or many bones, the graver cases of which may present abnormal pigmentation of skin, premature sexual development, hyperthyroidism or still other extraskeletal abnormalities. Arch Pathol 1942;33:777.
Question 91
A 25-year-old man has had low back pain for the past 4 months. An AP pelvis radiograph, CT scan, MRI scan, and biopsy specimen are shown in Figures 91a through 91d. What is the most likely diagnosis?

1. Giant cell tumor
2. Chordoma
3. Osteosarcoma
4. Hemangioma
5. Rectal adenocarcinoma

PREFERRED RESPONSE: 1

DISCUSSION: Imaging shows a lesion in the sacrum, with extension out of the bone. Histology shows multinucleated giant cells with deposits of hemosiderin. This is consistent with giant cell tumor. Chordoma commonly occurs in the sacrum as a midline lesion of notocord remnants, but the histology would show physaliferous cells. Osteosarcoma occasionally occurs in the sacrum, but histology would show malignant cells and osteoid. Hemangiomas in the spine are typically small, intraosseus lesions with vertical striations. Rectal adenocarcinoma would originate anterior to the sacrum and show a glandular pattern on biopsy.

Question 92
An 11-year-old boy reports pain after throwing a ball in gym class. He denies prior pain in the arm. Radiographs of the humerus are shown in Figures 92a and 92b. What is the next most appropriate step in management?

1. Staging studies (MRI, bone scan, and chest CT)
2. Open biopsy
3. Sling and closed fracture care
4. Curettage and bone grafting
5. Open reduction and internal fixation

PREFERRED RESPONSE: 3

DISCUSSION: The radiographs reveal a fracture through a lesion of the proximal humerus. The central lucency without any obvious matrix and thinning of the cortical bone are typical findings of an unicameral bone cyst. The proximal humerus is the most common site for this lesion. Typically these fractures should be allowed to heal prior to treatment, which may include, classically, aspiration and injection of corticosteroid. Recently other treatment options, including the injection of bone marrow aspirate or bone graft substitutes, have been reported, with open curettage and bone grafting reserved only for recurrent or nonresponding cysts.

Question 93

Figures 93a through 93d show the MRI scan, CT scans, and a biopsy specimen of a 23-year-old woman who reports left-sided back pain that travels around the lateral aspect of the thigh and down the lateral aspect of the left leg. The pain is relieved with anti-inflammatory medications. It is not associated with weight bearing. What is the most likely diagnosis?

1. Osteosarcoma
2. Osteoblastoma
3. Giant cell tumor
4. Aneurysmal bone cyst
5. Stress fracture

PREFERRED RESPONSE: 2
DISCUSSION: Osteoblastoma is a bone-forming lesion characterized by a mixed lytic-blastic lesion of variable size greater than 2 centimeters. Histology shows osteoid formation with benign rimming osteoblasts and an intervening highly vascular stroma. Osteosarcoma can look similar radiographically but histology typically shows malignant spindle cells interspersed in osteoid. Giant cell tumor histology shows multiple multinucleated giant cells in a background stroma of stromal cells with similar nuclei. Aneurysmal bone cyst typically has lakes of blood surrounded by a thin benign fibrous membrane. Stress fracture does not typically have a large lucent central lesion on radiographs.


Question 94
What is the primary problem in rickets osteomalacia?

1. Defect in the zone of proliferation within the physis
2. Defect in type I collagen
3. Defect in the ext-1 gene
4. Low level of calcium
5. Production of dysplastic fibrous bone

PREFERRED RESPONSE: 4

DISCUSSION: Rickets is a disorder of bones in children that results from decreased calcium available in the blood resulting in poor mineralization of bone that can lead to fractures and deformity. The most common cause of rickets is from vitamin D deficiency but it can also be caused by poor nutrition or gastrointestinal disease that results in poor calcium absorption such as celiac disease or severe diarrhea from other causes. Rickets is not primarily a physeal disorder. Osteogenesis imperfecta is caused by a defect in type I collagen. A defect in the ext-1 gene is often seen in patients with multiple hereditary exostoses. Fibrous dysplasia also can result in bone deformity and fractures due to production of dysplastic fibrous bone but is not caused by calcium or vitamin D deficiency.

A 42-year-old man has pain and swelling about his knee that has been worsening over the last 6 months. Examination shows an effusion, bogginess in the suprapatellar pouch, and a decrease in range of motion when compared to the contralateral knee. Radiographs are shown in Figures 95a and 95b and a sagittal MRI scan is shown in Figure 95c. A biopsy specimen is shown in Figure 95d. What is the most appropriate treatment for this lesion to minimize the risk of local recurrence?

1. Observation
2. Intra-articular injection of methylprednisolone
3. Limited arthroscopic synovectomy
4. Open anterior and posterior synovectomy
5. Intra-articular injection of radioisotope

PREFERRED RESPONSE: 4
DISCUSSION: The clinical presentation, radiographic studies, and biopsy specimen are all consistent with pigmented villonodular tenosynovitis (PVNS). Most agree that the lowest chance of recurrence for diffuse PVNS of the knee is with open anterior and posterior synovectomy. However, this can be associated with significant morbidity. Arthroscopic synovectomy, while associated with a higher recurrence rate, is preferred by some due to lower morbidity. Regardless of the aggressiveness of synovectomy, the recurrence rate of diffuse PVNS remains significant. The cause of PVNS is controversial, but most believe it is a non-neoplastic inflammatory process that results in intra-articular synovial thickening, bloody effusion, pain, and even intraosseous erosion. Intra-articular radioisotopes have been removed from the market because of the risk of hematomyelopoetic diseases. Observation alone will likely lead to disease progression and joint destruction. Intra-articular steroids will not halt the progression of PVNS and, therefore have no role in its treatment.


Question 96
Acral bone metastases (to the hands and feet) are most likely the result of a primary solid organ tumor in which of the followings structures?

1. Liver
2. Lung
3. Breast
4. Thyroid
5. Prostate

PREFERRED RESPONSE: 2

DISCUSSION: Lung cancer is the most likely solid organ cancer to metastasize distal to the elbow and the knees. The most common sites for metastatic disease are the vertebral bodies, pelvis, and long bones. Genitourinary tumors may also metastasize to distal sites.

Question 97

A 13-year-old girl has had a painless thigh mass for the past 3 months. A biopsy is performed and chromosome analysis reveals a t(X;18)(p11:q11) translocation (involving the genes SYT-SSX). What is the most likely diagnosis?

1. Liposarcoma
2. Synovial sarcoma
3. Myxoid chondrosarcoma
4. Gastrointestinal stromal tumor
5. Malignant fibrous histiocytoma

PREFERRED RESPONSE: 2

DISCUSSION: Although synovial sarcoma accounts for 6% of all soft-tissue sarcomas, it represents 16% of soft-tissue sarcomas in adolescents and young adults, making it and rhabdomyosarcoma (which accounts for 15%) the most common soft-tissue sarcomas in this age group. The (X;18)(p11;q11) translocation is characteristic of synovial sarcoma. Myxoid chondrosarcomas often demonstrate a t(9;22) translocation. Well-differentiated liposarcomas and malignant fibrous histiocytomas may demonstrate a ring form of chromosome 12. Gastrointestinal stromal tumors may exhibit monosomies of chromosomes 14 and 22.

Question 98
Figures 98a through 98c show the radiograph, MRI scan, and biopsy specimen of a 13-year-old girl who reports increasing right ankle pain for the past 2 months. Work-up reveals no other lesions. What is the most appropriate treatment for this patient?

1. Surgery alone
2. Radiation therapy alone
3. Chemotherapy alone
4. Surgery and radiation therapy
5. Surgery and chemotherapy, with or without radiation therapy

PREFERRED RESPONSE: 5

DISCUSSION: The imaging studies show a permeative destructive lesion of the distal fibula with periosteal reaction. The biopsy specimen demonstrates a small blue cell tumor consistent with Ewing’s sarcoma. The most appropriate treatment for this patient would be a combination of surgery and chemotherapy. Local control of Ewing’s sarcoma is usually achieved by surgery with wide margins. In some cases, radiation therapy can be used for local control if wide resection would be associated with unacceptable morbidity. Chemotherapy, however, is required in all cases to treat systemic disease even if no metastases are revealed on the initial work-up.

Question 99
When compared with postoperative external beam radiation therapy for the treatment of soft-tissue sarcomas, preoperative radiation therapy is associated with which of the following?

1. Higher wound complications
2. Higher doses of radiation
3. Larger volumes of tissue irradiated
4. Worse overall survival
5. Worse functional results

PREFERRED RESPONSE: 1

DISCUSSION: Preoperative radiation therapy requires a lower dose of radiation (5,000 cGy versus 6,600 cGy) and lower volume of tissue, with no difference in survival and a trend toward better functional outcome, compared with postoperative radiation. Preoperative radiation is associated with a significantly higher wound complication rate (35% versus 17%).

Question 100

A 43-year-old woman has a pathologic right acetabular fracture seen in Figure 100. Laboratory studies are unremarkable but a bone scan shows multiple skeletal areas with increased activity, and a CT scan of the chest/abdomen/pelvis shows some visceral involvement and also a right breast mass, suspicious for a primary lesion. What is the next most appropriate step in management?

1. PET scan
2. Biopsy
3. Referral to medical oncologist
4. Total hip arthroplasty with fixation of the acetabular fracture
5. Hospice referral

PREFERRED RESPONSE: 2

DISCUSSION: The next most appropriate step in management is to proceed with a biopsy of the most accessible site. This can be done by the surgeon or by an interventional radiologist trained in core biopsy techniques. It is imperative to make a pathologic diagnosis prior to proceeding with any further medical, surgical, or radiation treatments. A histologic diagnosis at this point is the only way a medical oncologist can have a meaningful discussion with the patient about their disease, its natural history, and ultimately discuss treatment options and prognosis. Further imaging at this point only delays the time to histologic evaluation. Consideration of surgical stabilization can be delayed until a diagnosis is established and a multidisciplinary approach is initiated. This fracture can be treated at least temporarily with nonsurgical protected weight bearing with a walker or crutches. Hospice may soon serve a useful role but a diagnosis must first be rendered and a limited life expectancy anticipated.
