1. Which of the following is TRUE regarding both osteosarcoma and Ewing sarcoma?

A. Both tumors are associated with specific genetic aberrations  
B. At the time of primary diagnosis, the majority of patients (>50 percent) with both osteosarcoma and Ewing sarcoma have radiologic evidence of metastatic disease  
C. Both osteosarcoma and Ewing sarcoma occur most commonly in the lower extremity  
D. Both osteosarcoma and Ewing sarcoma are most commonly centered in the metadiaphysis

Correct Answer: C. Both osteosarcoma and Ewing sarcoma occur most commonly in the lower extremity

Rationale: C is the answer. Option A is not the answer: Ewing sarcoma is associated with a chromosomal translocation involving chromosomes 11 and 22 in 85-95% of cases. On the other hand, there is no single pathognomonic genetic mutation, translocation of aberration associated with osteosarcoma. Option B is not the answer: Approximately 15% of patients with osteosarcoma have radiologic evidence of metastatic disease at the time of diagnosis. Approximately 25% of patients with Ewing sarcoma have metastatic disease at the time of diagnosis. Option D is not the answer: Ewing sarcoma most commonly occurs in the metadiaphysis and the diaphysis. However, osteosarcoma is most common in the metaphysis.

References:


2. Regarding skip metastasis in primary bone sarcomas, which of the following is TRUE?

A. On MR images, skip metastasis are best detected by T2 weighted sequences  
B. Skip metastasis are distinctly separate from the primary tumor, may occur in proximity to or distal from the primary tumor, within the same bone or across the joint  
C. Skip metastasis occur more frequently in Ewing sarcoma than osteosarcoma  
D. MDP bone scintigraphy has been shown to be more reliable than MRI in the detection of skip metastasis
Correct Answer: B. Skip metastasis are distinctly separate from the primary tumor, may occur in proximity to or distal from the primary tumor, within the same bone or across the joint

Rationale: Option A is not the answer: Using MRI, skip metastasis are best detected by T1 weighted sequences. Option C is not the answer: While skip metastasis are rare overall, the incidence is more frequently reported in the setting of osteosarcoma than Ewing sarcoma. Option D is not the answer: MRI has been shown to be more reliable than MDP bone scintigraphy in the detection of skip metastasis.

References:

Bone Tumors: Benign Lesions That Mimic Malignancy
Micheál Breen MBBChBAO, (Hons) BMedSc, MRCPI, FFRRCSI, DABR

3. Which of the following mimics of malignant bone tumor commonly affects the clavicle?

A. Stress Injury
B. Chronic Noninfectious Osteomyelitis
C. Scurvy
D. Nora Lesion

Correct Answer: B. Chronic Noninfectious Osteomyelitis

References:
Imaging of Chronic Recurrent Multifocal Osteomyelitis Geetika Khanna, Takashi S. P. Sato, Polly Ferguson. Author Affiliations Published Online:Jul 1 2009https://doi.org/10.1148/rg.294085244

4. What is the estimated rate of malignant transformation in children with multiple osteochondromata?

A. 0-5%
B. 5-10%
C. 10-15%
D. 15-20%

Correct Answer: A. 0-5%

References:
Clinical and Imaging Features of Rhabdomyosarcoma
Kieran McHugh, FRCR

5. With rhabdomyosarcoma (RMS) tumors in children, one of the following is recognised as an unfavourable risk factor.
   
   A. Embryonal histology
   B. Tumour size or diameter of 4cm
   C. Parameningeal location
   D. Age less than 10 years

Correct Answer: C. Parameningeal location

Rationale: Embryonal histology typically confers a good prognosis. Tumour size greater than 5cm in any orthogonal plane is generally recognised as a poor prognostic factor as is age greater than 10 years. Other unfavourable risk factors would be alveolar histology, nodal involvement and/or metastatic disease at diagnosis. Favourable sites of disease include the orbit, non-parameningeal head and neck (H&N) sites, and non bladder/prostate genitourinary (GU) sites. Unfavourable sites include the extremities, parameningeal tumours and bladder/prostate primaries.

References:


6. One of the following cancer predisposition syndromes is much more associated with soft tissue sarcomas including rhabdomyosarcomas than Wilms’ tumor.

   A. Li-Fraumeni syndrome
   B. Denys-Drash syndrome
   C. Perlman syndrome
   D. Simpson-Golabi-Behmel syndrome

Correct Answer: A. Li-Fraumeni syndrome

Rationale: Cancer predisposition syndromes can be associated with a variety of malignancies but Li-Fraumeni syndrome is in particular associated with soft tissue sarcomas including rhabdomyosarcomas. Answers B-D are typically associated with Wilms’ tumor predisposition. Other syndromes associated with a risk of developing rhabdomyosarcomas include neurofibromatosis type 1 (NF1) and DICER 1 syndrome (pleuropulmonary blastoma, cystic nephroma, embryonal or fusion negative RMS).
References:


Overview of the Clinical and Imaging Features of the Most Common Non-rhabdomyosarcomatous Soft Tissue Tumors
Simon C. Kao, MD, FACR

7. Regarding pediatric nonrhabdomyosarcoma soft tissue tumor (NRSTS):

A. NRSTS makes up 85% of the soft tissue sarcomas (STS) in children
B. The most common site is in the head and neck region
C. The clinical evidence of genetic predisposition is rare
D. Low-grade tumors have a 25% risk of metastasis

Correct Answer: C. The clinical evidence of genetic predisposition is rare

Rationales: Most NRSTS are sporadic. However there are rare genetic predisposing conditions, such as malignant peripheral nerve sheath tumor in neurofibromatosis type 1, soft tissue and bone sarcoma in Li-Fraumeni syndrome, and soft tissue sarcomas in patients with Werner’s syndrome. Option A is not correct. The NRSTS makes up 50-60% of all pediatric STS, the rest being rhabdomyosarcoma. Option B is not correct. The most common site of involvement is in the lower extremity. Option D is not correct. The low-grade tumors have a 2-10% risk of metastasis.

References:


8. In pediatric synovial sarcoma:

A. Synovial sarcoma is the most common STS in infants
B. The presence of ASPSCR1-TFE3 (ASPL-TFE3) fusion transcript is a sensitive and specific test for this tumor
C. Calcifications are seen on radiography in 5% of cases
D. MRI findings may mimic a venous malformation near a joint

Correct Answer: D. MRI findings may mimic a venous malformation near a joint
Rationales: The use of susceptibility-weighted sequences may be helpful in distinguishing a synovial sarcoma mass from a venous malformation by showing blood products in the latter. Option A is not correct. Infantile or congenital fibrosarcoma is the most common STS in infancy, representing approximately 10% of pediatric STSs. Synovial sarcomas account for approximately 6-10% of all STSs. Option B is not correct. The presence of SYT/SSX fusion transcript is a sensitive and specific test for synovial sarcoma. ASPSCR1-TFE3 (ASPL-TFE3) fusion protein is seen in alveolar soft-part sarcoma. Option C is not correct. Calcifications are seen on radiography in 30% of cases. They appear as areas of low signal intensity on T1- and T2-weighted images.

References:


9. In malignant peripheral nerve sheath tumors (MPNSTs):

A. The reported incidence is 2-5% in patients with neurofibromatosis type 1
B. Most MPNSTs exhibit hyperintensity on T1W and T2W images, and central and uniform contrast enhancement
C. PET-CT is not useful differentiating benign from malignant peripheral nerve sheath tumors
D. This is associated with t(12;15)(p13;q25) translocation

Correct Answer: A. The reported incidence is 2-5% in patients with neurofibromatosis type 1

Rationales: The reported incidence is 2-5% in patients with neurofibromatosis type 1. Also, two-third of patients with MPNST has neurofibromatosis type 1, while the rest is sporadic. Option B is not correct. Most MPNSTs exhibit hypointensity on T1W images, hyperintensity on T2W images, and peripheral and irregular contrast enhancement. Mixed high signal intensity can be seen on T1WI due to hemorrhage. Option C is not correct. PET-CT is useful differentiating benign from malignant peripheral nerve sheath tumors Important MRI findings raising suspicion of malignancy include diameter >5cm, ill-defined margin, intratumoral lobulation, central hemorrhage or necrosis, edema, calcification, loss of “target” sign, and inhomogeneous contrast enhancement. A cut off SUV max value of 4 has been used to identify lesions concerning for malignant transformation (mostly from plexiform neurofibromas) with high sensitivity and specificity. Option D is not correct. This translocation is seen in infantile fibrosarcoma. NF-1 is associated with changes in the NF-1 gene, a tumor suppressor gene located in the proximal long arm of chromosome 17 encoding the neurofibromin protein. There is no translocation or fusion protein in MPNSTs.

References:


