Answers and Additional Information for the Orthopaedic Quiz

Case 1
A1. A large lobulated smooth swelling was found on the anterior knee as a result of the thigh. MRI shows an encapsulated subcutaneous swelling with its larger (8x5cm) heterogeneous cyst containing debris connected to a smaller homogenous cyst.

A2. A lesion larger than 8cm requires tissue biopsy to exclude soft tissue sarcoma. An ultra-sound guided biopsy may be done. As a long standing cystic swelling, ganglion cyst is the most likely diagnosis.

A3. Due to the benign nature of the swelling, an extra capsular excision is indicated.

Ganglion Cysts
A ganglion cyst usually presents with a swelling with or without pain and weakness. It represents accumulation of gelatinous fluid within a mucoid cystic degenerative fibrous connective tissue matrix of joint capsule or tendon sheath. At least 10% of the cases are the result of repeated minor trauma or a specific traumatic event. Myxoid degeneration, developmental abnormality, and other associations with injury or arthritis have been postulated as likely causes.

The accuracy rate of diagnosing ganglion cysts by high-resolution ultrasonography has reached 90% or more. Many imaging modalities are now available for studying soft-tissue structures, such as CT-scans or MRI, which are especially useful in detecting the ganglion in unusual locations.

Excision biopsy is the most preferred treatment option for this benign tumour. The key to a successful or adequate excision is to completely excise the stalk and its base at the degenerative portion of the capsule or tendon sheath. Generally, there is a low recurrence rate after adequate excision.

Case 2
A1. Plain radiograph shows sclerotic endosteal with lytic lesions affecting the proximal-third of the femur with intense periosteal reaction or thickening and cortical thickening. The sagittal and axial MRIs show new periosteal bone formation without 'onion peel' like pattern or soft tissue invasion.

A2. The clinical and radiological findings are that of chronic sclerosing osteomyelitis of Garré but Ewing tumour is a close possibility. Intralesional tissue biopsies for histopathological examination and culture are needed to establish the diagnosis. An open biopsy is preferred as needle biopsy is less likely to be successful owing to sclerotic bone. Tissue culture for bacterial isolation needs to be extended for more than 7 days to yield significant growth of slow-growing colony of Propionibacterium acne. Bacterial identification can be enhanced by using PCR method.

A3. Using Cierny-Mader classification of chronic osteomyelitis, this predominantly intraendosteal or endosteal disease is classified as stage-IA disease (anatomical type-1, physiological host type-A).

A4. In the absence of any malignant cells, treat the case as chronic osteomyelitis with trial of three-week course of intravenous antibiotics followed by further three-week of oral antibiotics. The patient may become asymptomatic with pre-emptive suppressive antibiotic therapy against Staphylococcus aureus or Propionibacterium acne. However, based on Cierny-Mader concept of chronic osteomyelitis, medullary reaming followed by 4-6 weeks course of targeted antibiotics is a recommended surgical option for recurrent cases of this special type of chronic osteomyelitis.

Chronic Osteomyelitis Mimicking Tumour
Pain and swelling are the classical presentation of musculo-skeletal tumours particularly osteosarcoma. However infection (chronic osteomyelitis), occult fractures and myositis ossificans also have similar presentation and must be differentiated from tumours and vice versa. Early and accurate diagnosis of infection can be enhanced by Technetium, Gallium or Indium-labeled leukocyte scans. MRI and ultrasound can also detect early presence of fluid, effusion or pus. Figure 5 (IMG 0495) is a radiograph belongs to a 6 year-old child showing pathological supracondylar humeral fracture with extensive new bone formation around distal humerus mimicking osteosarcoma. However, Technetium scans indicated an infective process. This was confirmed later with tissue biopsy.

Figure 6 (IMG 0916) on the other hand is a radiograph belongs to a 9 year-old boy with similar presentation over proximal thigh with fever and leukocytosis which turned out to be Ewing sarcoma.

Case 3
A1. A pathological fracture through a well circumscribed cystic lesion of the humerus. There is an obvious free piece of bone within the cyst (fallen leaf sign). Figure 2 (IMG 3743) shows another example of fallen leaf sign.

A2. ABC is usually more ballooning or expansile than a simple bone cyst. It occurs in older children and adolescents. There are coarse trabeculations within the cyst. An MRI beautifully demonstrates fluid level (cells and serum). It does not resolve by itself.

A3. As the previous fracture united with conservative management, conservative management with cast and sling is indicated. Steroid injection or curettage is necessary if cyst persists.

Unicameral or Simple Bone Cyst
A unicameral bone cyst (UBC), otherwise known as a simple bone cyst, is a fluid-filled cavity in the metaphyseal region of long bones of growing child. It usually occurs in the upper part of the humerus (50-60% of the time) or the upper part of the femur (25-30% of the time). The presence of these cysts in other bones, particularly the calcaneus and pelvis has been described. The cyst is typically lined by thin compressed fibrous tissue. Unicameral bone cysts are considered benign. Some heal spontaneously, while others enlarge. More invasive cysts can grow and may cause a pathological fracture. The risk of a pathological fracture is difficult to predict. However, in a series of predominantly humeral bone UBCs, Mary et al.(2004) found that if the cyst had the following features: width > 30 mm, height > 75 mm, cortical thickness < 2.4 mm and Kaelin index (cyst surface area and shaft diameter ratio) > 5, the risk of fracture increased to 80% or more.

Case 4
A1. MRI shows a subcutaneous well-circumscribed and capsulated calcified mass. The cyst is trabeculated and contains fluid.

A2. Differential diagnoses include calcified sebaceous cyst, calcified haematoma, myositis ossificans, metastatic deposit from prostatic carcinoma etc. The most likely diagnosis is a calcified haematoma.

A3. If biopsy shows an organised haematoma, a complete excision with its capsule is indicated.

Calciﬁed Haematoma
Although there have been a number of reports published on myositis ossificans, soft tissue osteosarcoma, and soft tissue sarcoma, little has been written on calcifying haematoma. These lesions can clinically and radiologically mimic aggressive soft tissue neoplasms. Most of the published cases had history of prior injury to the affected site as long as 20 years earlier. Histological examination showed a cavity containing necrotic debris, occasional fibrin, areas of recent haemorrhage and areas of dystrophic calcification.
Case 5

A1. There is a sclerotic lesion of poor margins projecting from the posterior cortex of the tibia and away from the knee joint. MRI shows a large cartilaginous component displacing calf muscles but not invading them. The mass looks encapsulated.

A2. The most likely diagnosis is malignant sarcomatous transformation of a pre-existing solitary osteochondroma (exostosis). Clinical features indicative of malignant transformation in enlarging tumour of more than 6cm with recent onset of pain occurring after skeletal maturity. Radiographic indicative of malignant changes include increased thickness of cartilage cap of more than 3cm with irregular and indistinct margin, cartilage foci extending beyond the periosteal cartilage cap and variable pattern of mineralization.

A3. Tissue biopsy either by ultrasound-guided technique or open method is a requisite for establishing the diagnosis. A high diagnostic yield is achievable by proper tissue sampling from the base of the tumour. Biopsy site based on bone scans localization of hot spots frequently yielded an inconclusive or negative result as this merely representing mineralization activity of endochondral ossification in the cap. Sarcomas arising from solitary osteochodromas are usually chondrosarcoma, but other sarcomas such as osteosarcomas and fibrosarcomas also occur.

A4. A cautious decision on type of surgical excision margin must be employed. A tumour without soft tissue extension requires marginal extra-capsular excision by including its base and periosteal fibrous band if the risk of local recurrence is to be minimized. Difficulty in anatomic dissection of cap from surrounding soft tissue may indicate a high-grade malignancy and this necessitates a wide surgical excision margin. For this patient, the excised tissue was a low-grade chondrosarcoma and she has been asymptomatic for two years following marginal excision of the tumour.

Low-grade Chondrosarcoma

Chondrosarcoma (CS) is a malignant tumour that produces cartilage matrix. Primary CS is very uncommon, arises centrally in the bone, and is found in children. Secondary CS arises from benign cartilage defects such as osteochondroma or enchondroma. CS is usually classified by anatomic location (intramedullary or classic, periosteal or juxtacortical or surface, and extraskeletal).

The presentation of CS depends on tumour grading. A fast growing high-grade tumour can present with exsanguinating pain. A more indolent low-grade tumour is likely to present in an older patient with regional steady pain and swelling.

A low-grade CS is very close in appearance to enchondromas and osteochondromas by showing occasional binucleated cells and the diagnosis may be difficult as in this case. High-grade chondrosarcomas have increased cellularity, atypia and mitoses. Because most CSs are resistant to chemotherapy and radiotherapy, surgical resection remains the mainstay of treatment. Survival rate has been correlated to histological grade and reduces precipitously with high-grade large axial tumours.

Case 6

A1. Glomus tumour or glomangioma. This benign lesion can pose a diagnostic dilemma to clinicians unfamiliar with the entity. Even with classic presentations, most patients suffered from a delayed in establishing the correct diagnosis for years.

A2. Radiograph depicted a well-defined lucency in the tuft of the distal phalanx. Lateral radiograph is useful as it may show mild scalloping of the distal phalanx at the site of the lesion. Provocative test by application of cool spray or immersion in ice water is helpful to enhance the diagnosis.

A3. Marginal excision brings immediate symptomatic relief and is curative. Recurrence is uncommon and is usually due to inadequate search for additional glomus bodies.

Glomus Tumours or Bodies

The glomus tumours represent specialized arteriovenous anastomoses of preterminal arterioles and efferent veins of the neuromyoarterial glomus bodies that regulate local blood flow and temperature in the fingertips and nail beds. As such, it common or predilected sites include the fingertips and nail beds. However, other unusual sites such as the foot, ankle or the tip of the coccyx have been reported.

The tumours consist of modified perivascular smooth-muscle cells arranged in sheets and nests. They appear as bluish red nodules in the subcutaneous and dermal tissue of the limbs with a predilection for nail beds tissue of the finger. Histologically, they are composed of glomus cells, proliferating vascular spaces and various stromal components encased by a dense fibrous capsule. The sheets of glomus cells characteristically envelop small-diameter vessels.