PICTORIAL ESSAY

Bone Surface Tumours and Tumour-like Conditions

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ABSTRACT
Bone lesions that arise from outside of the medullary canal can be referred to as bone surface lesions. The aetiology of these lesions may be divided into neoplastic and non-neoplastic. Distinguishing the exact origin of the bone surface lesion may be challenging. This article discusses the imaging features of various bone surface lesions, with emphasis on lesion characterisation and clinical features.

Key Words: Adamantinoma; Chondroma; Chondrosarcoma; Myositis ossificans; Osteoma, osteoid; Osteosarcoma

中文摘要
骨表面腫瘤和類腫瘤病症
李昭穎、謝健燊、黎國忠、陳文光、曾慧勤、蕭俊傑、鄭珊珊、鄧國穎
從骨髓管外起源的病變腫瘤可稱為骨表面病變。其病因可分為腫瘤性和非腫瘤性，要區分骨表面病變的確切起源並不容易。本文討論了各種骨表面病變影像學特徵，特別是病變的特徵和臨床表現。

INTRODUCTION
Bone lesions can arise from within or outside of the medullary canal. The latter can be referred to as bone surface lesions. The aetiology of these lesions may be divided into neoplastic or non-neoplastic. Understanding the clinical and radiological features of these tumours and tumour-like conditions is helpful in establishing the diagnosis and guiding further management. This study was approved by the Kowloon Central Cluster / Kowloon East Cluster Research Ethics Committee.

ANATOMY
The bone surface includes three main regions: the cortex, the periosteum, and the fibrous tissues superficial to the periosteum. Lesions that arise from within the cortex can be further subdivided into endosteal and intracortical. An endosteal lesion arises from the inner part of cortex immediately adjacent to the medullary cavity, whereas an intracortical lesion is centred within the cortex. A subperiosteal lesion lies between the cortex and the periosteum. A lesion that arises from the periosteum is termed a periosteal lesion. Those that arise from the fibrous tissue superficial to the periosteum are referred as parosteal or juxtacortical lesions.1 Distinguishing the exact origin of the bone surface lesion on imaging may be difficult. Characterisation of the lesion based on the composition and radiological features can help establish the diagnosis and guide...
COMPOSITIONS
The most common neoplastic bone surface lesions are bone-forming and cartilage-forming tumours. Bone-forming tumours include osteoma, osteochondroma, osteoid osteoma, paracortical osteoblastoma, and surface osteosarcomas. Cartilage-forming lesions include periosteal chondroma, bizarre parosteal osteochondromatous proliferation (Nora’s lesion), and periosteal chondrosarcoma. Rarer bone surface neoplastic lesions composed of epithelial and osteofibrous components include adamantinoma. Tumour-like lesions include myositis ossificans and cortical desmoid.

BONE-FORMING TUMOURS
Surface osteosarcomas can be divided into parosteal, periosteal and high-grade surface subtypes. Different subtypes carry different radiological appearances, treatment options, and prognosis.

Parosteal Osteosarcoma
Parosteal osteosarcoma (Figure 1) is the most common of the subtypes, accounting for 65% of surface osteosarcomas. 95% of cases occur in the age-group of 15-40 years, with a female-to-male ratio of 3:2. It is usually metaphyseal and frequently seen at the posterior distal femoral shaft.

On plain radiograph, it is usually a lobulated, exophytic mass with central dense ossification adjacent to the bone, and may encircle the bone when it is large. Cortical thickening with a relative lack of aggressive periosteal reaction is a feature. A cleavage plane separating the tumour and underlying cortex, corresponding to the interposed periosteum, may be observed on cross-sectional imaging. This feature may be useful in differentiating parosteal osteosarcoma from periosteal or high-grade subtypes of surface osteosarcoma.

Periosteal Osteosarcoma
Periosteal osteosarcoma (Figure 2) is the second commonest subtype of surface osteosarcomas, comprising 25% of all surface osteosarcomas. It commonly affects young patients in the second and third decades of life. Most frequent locations of involvement are the femur and tibia, followed by the ulna and humerus. The diaphysis of the long bones is the most common origin. The 5-year survival rate of 83% is better than that for conventional osteosarcoma, but worse than that for parosteal osteosarcomas. Histologically, it arises from the inner, germinative layer of periosteum.

On plain radiograph, it is typically a broad-based soft-tissue mass attaching to the cortex, without an abnormality in the adjacent medullary canal. Periosteal...
reaction is common and can appear as solid non-aggressive cortical thickening or as aggressive Codman triangle, irregular or laminated forms. Periosteal reaction perpendicular to the osseous long axis and extending into the soft-tissue mass may also be seen.12

High-grade Surface Osteosarcoma
High-grade surface osteosarcoma (Figure 3) is rare and the least common subtype, comprising 10% of all surface osteosarcomas.13 It is more typically seen in the second or third decades of life,14 with a male-to-female ratio of 3-4:1.14-16 The lesion commonly affects the diaphysis of bone, especially the mid or distal femur or in the tibial diaphysis. Histologically, a high-grade surface osteosarcoma arises from the surface of bone, with high mitotic activity identical to that of conventional high-grade intramedullary osteosarcoma.8

On plain radiograph, it classically appears as a moderate-to-dense fluffy, immature mass with broad attachment to the cortex. Cortical erosion or thickening may be present. There are several features that may help differentiate high-grade surface osteosarcoma from parosteal or periosteal subtypes of surface osteosarcoma. A lucent cleavage plane between the tumour and underlying cortex is a rare finding in high-grade surface osteosarcoma, in contrast to parosteal osteosarcoma. Moreover, radiating spicules of periosteal reaction are less commonly seen than in periosteal osteosarcoma.14 In addition, high-grade surface osteosarcoma may involve the medulla or sometimes the entire circumference of bone,17 but intramedullary involvement or circumferential tumour involvement are not typical features of periosteal osteosarcoma.

Osteoid Osteoma
Osteoid osteoma (Figure 4) can be classified as cortical, medullary, or subperiosteal subtypes based on
Figure 3. High-grade surface osteosarcoma in a 46-year-old female who presented with thigh swelling for 3 months: (a) radiograph showing cortical thickening with dense sclerotic fluffy periosteal reaction at the mid diaphysis of right femur (arrow). No cleavage plane is seen between the tumour and underlying cortex, which is a differentiating feature from parosteal osteosarcoma. It also involves more than one half of the circumference of the bone, more commonly seen in high-grade surface osteosarcoma. (b) Computed tomography showing an expanded medial cortex of the femoral diaphysis, with spiculated periosteal reaction (arrow). Axial (c) T1-weighted and (d) T2-weighted with fat-saturation magnetic resonance images showing the tumour with deep intracortical involvement (arrows) but no intramedullary involvement.

Figure 4. Osteoid osteoma in a 16-year-old male who presented with left hip pain: (a) the nidus is not clearly shown on radiograph. (b) Computed tomography showing a radiolucent nidus with central mineralisation at the left ilium (arrow), a characteristic finding of osteoid osteoma. (c) Magnetic resonance imaging is less useful in demonstrating the central nidus, but is better at showing bone marrow oedema (arrow). (d) Biopsy of lesion confirmed the diagnosis of osteoid osteoma (arrow). This patient was treated with two sessions of radiofrequency ablation and achieved a good response, with no analgesia required.
radiographic findings. Cortical osteoid osteoma is the most common subtype.\textsuperscript{18} Classically patients complain of nocturnal pain that can usually be relieved by nonsteroidal anti-inflammatory drugs. On radiograph, it typically shows an intracortical nidus that is round or oval and usually measures <2 cm in diameter.\textsuperscript{19} It is usually radiolucent, but may also be calcified.\textsuperscript{1} Cortical thickening and reactive sclerosis in the long bone shafts are typical. On computed tomography (CT), the nidus is usually well-defined and lucent, with central mineralisation. Surrounding cortical thickening and periosteal reaction may be seen. Enhancement of hypervascular nidus may be seen on contrast CT.\textsuperscript{20} On magnetic resonance imaging (MRI), the nidus is of low-to-intermediate signal on T1-weighted images, and of variable signal on T2-weighted images. Adjacent bone marrow oedema may be present. Visualisation of the nidus is better on CT than on MRI. However, MRI is better at demonstrating associated bone marrow and soft tissue oedema, joint effusion, and synovitis.\textsuperscript{21}

**CARTILAGE-FORMING TUMOURS**

**Periosteal Chondroma**

Periosteal chondroma (Figures 5 and 6), also known as juxtacortical chondroma, is a benign tumour composed of mature hyaline cartilage, and is the surface variant of enchondroma.\textsuperscript{1} It commonly affects the long bones in children and young adults, with a slight male predominance.\textsuperscript{22,23} The proximal humerus is the most commonly affected site.\textsuperscript{23} On radiograph, it can be a periosteal-based lytic lesion with internal chondroid mineralisation. The adjacent cortex may be saucerised with a varying degree of sclerosis. Marginal periosteal new bone formation may be present.\textsuperscript{24} On MRI, T1-weighted imaging may show a hypointense signal and cortical scalloping or saucerisation. It is hyperintense on T2-weighted images. Perilesional bone marrow oedema may be present. Visualisation of the nidus is better on CT than on MRI. However, MRI is better at demonstrating associated bone marrow and soft tissue oedema, joint effusion, and synovitis.\textsuperscript{21}

As in intramedullary chondroid lesions, the differentiation of a benign from low-grade malignant periosteal chondroid lesion is difficult. Larger lesion size (>3 cm),\textsuperscript{25} intramedullary extension or oedema, and irregular soft tissue margins may suggest malignant chondrosarcoma. Nonetheless, these features may overlap with those of benign lesions.

**Periosteal Chondrosarcoma**

Periosteal chondrosarcoma (Figure 7) is rarer than the benign periosteal chondroma. It occurs in a slightly older age-group, mostly in the second to fourth decades of life,\textsuperscript{26} and commonly involves the femur.\textsuperscript{27} Patients usually present with insidious onset of pain and swelling.

Periosteal chondrosarcoma are usually larger than benign periosteal chondroma. In a case series of 11 periosteal chondrosarcomas, none of the lesions were <3 cm.\textsuperscript{24} On radiographs, it may appear as a juxtacortical soft tissue mass containing popcorn, spotty, peripheral or ring-and-arc calcifications.\textsuperscript{28-31} They tend to be more irregular and permeative on radiographs than chondromas.\textsuperscript{27,28,22} On CT, the lesion is juxtacortical, and can be associated with intact, thickened, or eroded cortex. Peripheral enhancement can be present if contrast is injected. MRI shows a juxtacortical mass with T1-weighted hypointense and T2-weighted hyperintense signals. Intramedullary extension or oedema may occur. Peripheral and septal enhancement following gadolinium injection is seen.\textsuperscript{30}

**Bizarre Parosteal Osteochondromatous Proliferation**

Also known as Nora’s lesion, bizarre parosteal osteochondromatous proliferation (Figure 8) is a rare, benign exostotic osteochondromatous tumour that may occur in the hands, feet, skull, and long bones of the upper and lower limbs.\textsuperscript{33} Recurrence is frequent (about 55%) following resection.\textsuperscript{33} Patients are usually in their third to fourth decade and both genders are affected.\textsuperscript{33,34} It usually presents as a minimally painful pedunculated mass that enlarges slowly over months or years,\textsuperscript{33,35} however a more aggressive growth pattern is also described.\textsuperscript{36} On radiographs, it may appear as a calcified and osseous mass adjacent to the affected bone, which may simulate an osteochondroma or other osteogenic tumour. At an early stage, the lesion can be differentiated from osteochondroma or other osteogenic tumours by the lack of medulla or cortical continuity with underlying bone. Nonetheless when mature, the lesion may appear to adhere to the adjacent bone with a pedunculated or sessile base, with the cortex of parent bone remaining intact.\textsuperscript{1,37} When it appears inseparable from the parent bone, it may mimic other benign or malignant conditions. Histology is required for a definitive diagnosis.

On CT, the calcified and ossified masses show well-defined margins, typically without continuity with the medullary canal of the bones.\textsuperscript{38}

On MRI, the lesion is classically described as cortically based, without intramedullary extension. Tumour
Figure 5. Periosteal chondroma of the proximal humerus in a 1-month-old boy: (a) there is an incidental finding of a cortical-based lucent lesion at the proximal humerus on radiograph (arrow). Adjacent cortical saucerisation and sclerosis are present. (b) Magnetic resonance imaging showing a juxtacortical lesion with saucerisation of the adjacent cortex (arrow). No bone marrow oedema or adjacent soft tissue mass are present.

Figure 6. Periosteal chondroma of the femur in a 22-year-old female who presented with a 2-month history of knee swelling: (a) radiograph showing a cortical-based lytic lesion with adjacent sclerosis and periosteal new bone formation (arrow). Internal calcification is evident. (b) Computed tomography showing a lucent lesion with sclerotic rim that is continuous with the elevated periosteum (arrow). Internal faint calcifications are seen. Magnetic resonance images showing an oval cortical-based mass with (c) isointense T1-weighted signal (arrow) and (d) hyperintense T2-weighted signal, with adjacent cortical erosion (arrow). (e) Peripheral enhancement (arrow) is seen on a post-gadolinium image. Excisional biopsy confirmed periosteal chondroma.
involvement of the medullary canal can be seen in more aggressive forms.\textsuperscript{36} It is of low signal intensity on T1-weighted images. On T2-weighted image, a hyperintense cartilage cap is sometimes seen.

**EPITHELIAL AND OSTEOFIBROUS TUMOURS**

**Adamantinoma**

Adamantinoma is a rare malignant tumour composed of epithelial and osteofibrous components. It can be classified as classic or differentiated. Classic type adamantinoma is typically seen in patients aged 20 to 50 years.\textsuperscript{39,40} Differentiated adamantinoma (Figure 9), also known as osteofibrous-like or juvenile adamantinoma, is usually found in those aged 20 years or younger.\textsuperscript{41} Patients commonly present with lower leg pain, swelling, or deformity.

On radiograph, both types of adamantinoma share similar features. They are usually central or eccentric osteolytic tumours in the diaphysis or metaphysis of the tibia, slightly expansile, with sharply or poorly delineated margins. They frequently involve the anterior tibial cortex, with septations and peripheral sclerosis.\textsuperscript{42} They are commonly intra-cortical lesions, but cortical destruction and extra-cortical soft tissues

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*Figure 7. Periosteal chondrosarcoma in a 37-year-old female who presented with a left shoulder mass: (a) radiograph and (b) computed tomography showing a juxtacortical mass at the proximal humerus metadiaphysis, with ring and arc calcifications. Cortical thickening and erosion are seen (arrows). (c) Whole-body bone scintigraphy showing increased tracer uptake over the left proximal humerus (arrow). Biopsy confirmed a periosteal chondrosarcoma.*
Figure 8. Bizarre parosteal osteochondromatous proliferation in a 40-year-old female who presented with a bony mass in the little finger: (a, b) radiographs showing an exophytic bony exostosis that appears contiguous with the underlying bony cortex, without cortical destruction (arrows). Excision was performed, with recurrence noted 3 months after surgery. (c, d) Radiographs showing recurrence of the exophytic bony mass (arrows). (e) Computed tomography showing the exostosis without bone destruction, medullary continuity or associated soft tissue mass (arrow). (f) T2-weighted magnetic resonance image showing bony outgrowth with a cartilaginous cap that is hyperintense (arrow). No intramedullary extension is seen. Re-excision of the mass was performed, and pathology results confirmed the diagnosis.

Figure 9. A 17-year-old male who presented with left knee pain: (a) frontal and (b) lateral radiographs showing a large well-defined geographic lytic lesion at the proximal tibia at a subarticular eccentric location (asterisks), subsequently biopsied and proven to be a giant cell tumour. There is another smaller lytic lesion at the anterior cortical region of the tibial proximal diaphysis (arrows), demonstrating well-demarcated margins and peripheral sclerosis. No cortical destruction, periosteal reaction, or extra-cortical soft tissue is seen. On (c) sagittal and (d) axial computed tomographic images, the biopsy-proven giant cell tumour is again shown (asterisk). The smaller lytic lesion at the anterior tibial cortical region (arrows) is more conspicuous than on radiographs. It demonstrates slight expansion, thinning of the anterior cortex, well-defined margins and peripheral sclerosis. No periosteal reaction or extra-cortical soft tissue is evident. Histopathology of this lesion showed differentiated adamantinoma. Wide excision with bone grafting was offered for this patient.
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may be seen in 15% of cases. Periosteal reaction is not frequently seen. Synchronous lesions in the ipsilateral fibula need to be looked out for, as these occur in 10% to 50% of cases. As classic adamantinoma is a low-grade malignancy with potential for metastasis, surgical intervention is the management of choice. The treatment for differentiated adamantinoma is less well-established. Some authors recommend observation of small differentiated adamantinoma in children, and wide resection for progressing lesions.

MIMICS OF BONE SURFACE TUMOURS

Cortical Desmoid

Cortical desmoid (Figure 10) is a benign, self-limiting fibrous or fibro-osseous lesion that is believed to be related to chronic stress applied by the adductor magnus aponeurosis or medial head of gastrocnemius. It is usually seen at the posteromedial surface of the distal femoral metaphysis at the attachment of the adductor aponeurosis to the medial supracondylar ridge. It commonly occurs in young males of 10 to 15 years old. These patients may be asymptomatic or complain of pain and soft tissue swelling. In some cases, a history of preceding trauma may be reported.

On radiograph, it typically shows a saucer-shaped irregular radiolucent cortical defect with adjacent sclerosis and periostitis in the characteristic location of the posteromedial cortex of the distal femoral condyle. Diagnosis can usually be made on radiograph, but CT may be used in equivocal findings. CT may demonstrate focal cortical thickening and fragmentation at the insertion of the medial head of the gastrocnemius tendon. Symmetrical findings may be seen on the contralateral side.

This lesion may be mistaken for other surface aggressive lesions such as osteosarcoma, chondrosarcoma, fibrosarcoma, or osteomyelitis. The classic clue is its location, and the absence of other aggressive features such as soft tissue mass or medullary involvement, sometimes further supported by the history of trauma or strenuous exercise. It is important to distinguish this benign entity from other aggressive lesions, as this is a ‘don’t-touch’ lesion, not requiring biopsy for diagnosis. It frequently heals spontaneously and may disappear by 20 years of age. If biopsy is performed, the lesion can be mistaken for malignancy, resulting in unnecessary amputation.

Figure 10. A lytic lesion in an 8-year-old female following right knee contusion: (a) lateral radiograph of the right knee showing a lucent lesion with sclerotic base at the posteromedial aspect of the distal femoral metaphysis (arrow). (b, c) Computed tomography showing a lytic area with sclerotic base at the posteromedial metaphysis of the distal femur, and at the insertion site of the medial head of the gastrocnemius muscle (arrows). No associated soft tissue mass is seen. Findings are typical of cortical desmoid, a ‘don’t-touch’ lesion, and biopsy should be avoided.
Myositis Ossificans

Myositis ossificans (Figure 11) is a pseudotumour that arises from muscles. It is a benign and self-limited entity but can mimic an aggressive surface bone lesion. It is most commonly seen in young adults, affecting the large muscles of the extremities. Approximately 60% to 70% of cases are associated with previous trauma. It usually presents as pain and soft tissue mass, or as an incidental finding. Contrary to malignant bone tumours, pain is most severe in the early course of the lesion. On radiograph it is usually shown as faint calcification within 2-6 weeks of symptom onset. A more sharply circumscribed bone mass may appear after 6-8 weeks. Differentiation from malignancy can be challenging, but there are features that can help in suggesting the diagnosis. A centrifugal maturation pattern may be observed in myositis ossificans, with more mature denser bone seen in the periphery of the mass. Secondly, a complete lucent zone between the lesion and underlying bone on at least one of the projections may be seen in myositis ossificans, whereas incomplete separation may be seen in parosteal osteosarcomas due to the presence of a pedicle extending from the tumour from the parent bone. Intact bone cortex despite the presence of periosteal new bone formation is another feature that points to benignity.

On CT, a rim of peripheral calcification with central area of low attenuation may be seen next to the bone in the early phase at 4-6 weeks. As the lesion matures, complete ossification of the mass may be demonstrated. As in plain radiographs, recognising the imaging features that favour benignity is important, as pathological distinction between myositis ossificans from osteosarcomas using biopsies may be difficult. Serial radiographic follow-up and conservative treatment with non-steroidal anti-inflammatory drugs are appropriate for most patients.

CONCLUSIONS

Bone surface lesions are not frequently encountered, and making a correct diagnosis based on imaging alone is often challenging. Biopsy is often required to establish a pathological diagnosis to aid in further management. Recognising the clinical and radiological features of tumour-mimics can help minimise unnecessary invasive examination or intervention for benign lesions.

REFERENCES


Figure 11. A 12-year-old female who presented with left forearm swelling for a month, without any history of trauma: (a) a rapidly appearing ossified mass over the proximal ulna was found on radiograph (arrow). (b) On computed tomography, a completely ossified mass is showing (arrow). In this case, the ossified mass was inseparable from the underlying bone. However, no other suspicious features such as cortical destruction or aggressive periosteal reaction are evident. (c) Serial follow-up radiographs showing remodelling of the ossified mass that gradually resolved 2 years after presentation (arrow).
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