Surface bone sarcomas are rare malignant bone tumours. Osseous and cartilaginous surface bone sarcomas are the most common, with parosteal and periosteal osteosarcomas, periosteal chondrosarcomas and secondary peripheral chondrosarcomas being the most frequent. Their clinical symptoms are non-specific and include pain for several months, swelling and limited range of motion of the adjacent joints. Prompt diagnosis is important, as biological behaviour, imaging and histopathologic characteristics, treatment and prognosis differ considerably from their conventional intramedullary counterparts. Moreover, their imaging characteristics are not infrequently non-characteristic and may be misinterpreted as juxtacortical benign lesions leading to incorrect diagnosis and treatment, with life-threatening repercussions. Molecular studies and histopathological sampling are essential for accurate diagnosis. There are still numerous issues regarding the biology, pathophysiology and treatment options of these entities due to their rarity.

Keywords: juxtacortical tumours; surface bone tumours

Introduction
Surface bone tumours are neoplasms situated near the bone cortex. They are classified into five different types: osseous, cartilaginous, fibrous, lipomatous and metastatic tumours. The most common types are those producing bone and cartilage. Osseous surface bone tumours include benign tumours such as osteoma, osteoid osteoma and osteoblastoma, and the malignant category of surface osteosarcomas (parosteal, periosteal and high-grade osteosarcoma), while the cartilaginous surface tumour category comprises benign tumours such as bizarre parosteal osteochondromatous proliferation (BPOP), periosteal chondroma, chondromyxoid fibroma, osteochondroma and malignant tumours such as periosteal chondrosarcoma and secondary peripheral chondrosarcoma. There are also exceedingly rare descriptions of periosteal Ewing sarcoma.

Regarding clinical symptoms, these are non-specific and include pain, local swelling and limited range of motion of the adjacent joint. Pain is usually present for several months. Although these lesions have similarities to their intramedullary counterparts, their location modifies their imaging characteristics. Imaging characteristics can be either non-characteristic and may be misinterpreted as juxtacortical benign lesions such as myositis ossificans, stress fracture, subperiosteal haematoma or abscess, osteochondromas, or BPOP, leading to wrong diagnosis with devastating consequences. This article aims to discuss the clinicopathological and imaging features, the current treatment and the prognosis of juxtacortical bone tumours.

Surface osteosarcomas
Surface osteosarcomas are distinct clinicopathological entities of osteogenic tumours rather than a subtype of intramedullary conventional osteosarcoma. Prompt diagnosis is important as their biological behaviour, imaging and histopathologic characteristics, treatment and prognosis differ considerably from those of conventional intramedullary osteosarcoma.

Regarding the terminology, the recent World Health Organization (WHO) classification for bone tumours does
not recommend the term juxtacortical osteosarcomas, while the Union for International Cancer Control (UICC) TNM classification (T=tumor extent, N=lymph nodes, M=distant metastases) of malignant tumours does not consider the TNM staging system for bone tumours suitable for surface osteosarcomas. However, other staging systems, such as the American Joint Committee on Cancer (AJCC) TNM system include these tumours in the bone staging system.

Surface osteosarcomas comprise approximately 4–12% of all osteosarcomas. They comprise three distinct entities, namely the parosteal (PAO), the periosteal (PEO) and the high-grade surface osteosarcoma (HGSO). The parosteal and periosteal subtypes are more common than HGSO. They tend to affect older patients compared to their conventional intramedullary counterparts. They arise on the outer cortical surface; however, the presence of intramedullary (IM) extension does not rule out a surface tumour as a small percentage have IM extension. They may invade or displace neurovascular bundles, tendons or ligaments in close proximity. Okada et al found that PAO invaded and displaced neurovascular bundles in 22% and 62% of cases respectively in cross-sectional imaging.

These three entities have a spectrum of different biological behaviour, ranging from low-grade (PAO), through intermediate grade (PEO) to high-grade (HGSO). Regarding treatment options, PAOs, as low-grade lesions, can be treated successfully with wide excision, while a combination of chemotherapy and wide excision is the standard of care for HGSO. Currently, the cost–benefit balance concerning the use of chemotherapy in the treatment of PEO has not yet been elucidated, and standardized treatment regimens need to be established to determine its effectiveness.

**Parosteal osteosarcoma (PAO)**

PAO is the most common surface osteosarcoma (65% of surface osteosarcomas) and represents 4–5% of all osteosarcomas. Peak incidence is at 20–40 years of age with slight female predominance. It is a low-grade malignant bone-forming tumour occurring on the cortical surface of bone, specifically the outer fibrous layer of the periosteum. Most frequent area is the metaphysis of long bones (Fig. 1), but it can also be detected in the diaphysis and metadiaphysis (Fig. 2). The most typical location is...
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the posterior surface of the distal femoral metaphysis followed by the medial surface of the of the femoral diaphysis. The presence of the cleft sign inferiorly (thin arrow) and a densely ossified stuck centrally (arrow) help to differentiate this PAO from an atypical osteochondroma or the rare exophytic fibrous dysplasia and cortical osteoma. Benign periosteal reaction (buttressing) is seen in adjacent cortex (asterisk). (B) Transverse computed tomography (CT) scan image confirms the presence of the broad, of cortical density stuck (arrowhead) and the cleft sign (thin arrows) that separates the ground glass mass (arrows) from the bone cortex. (C) The diaphyseal mass is mildly hypointense to muscles (arrows) on a coronal T1w magnetic resonance (MR) image. (D) On a coronal fat-suppressed T2w MR image the diaphyseal mass is inhomogenously hyperintense. The cortex remains intact with no medullary extension. (E) Intraoperative image of the posterior femur with the parosteal osteosarcoma. (F) The cortex of the posterior femur after tumour resection. (G) The resected specimen. (H) Postoperative radiographs of the femur after resection of the tumour and insertion of an intramedullary femoral nail.

Fig. 2 Parosteal osteosarcoma (PAO) in a 35-year-old female. (A) An X-ray of the right femur shows a smoothly marginated, ground glass density mass adjacent to the medial surface of the of the femoral diaphysis. The presence of the cleft sign inferiorly (thin arrow) and a densely ossified stuck centrally (arrow) help to differentiate this PAO from an atypical osteochondroma or the rare exophytic fibrous dysplasia and cortical osteoma. Benign periosteal reaction (buttressing) is seen in adjacent cortex (asterisk). (B) Transverse computed tomography (CT) scan image confirms the presence of the broad, of cortical density stuck (arrowhead) and the cleft sign (thin arrows) that separates the ground glass mass (arrows) from the bone cortex. (C) The diaphyseal mass is mildly hypointense to muscles (arrows) on a coronal T1w magnetic resonance (MR) image. (D) On a coronal fat-suppressed T2w MR image the diaphyseal mass is inhomogenously hyperintense. The cortex remains intact with no medullary extension. (E) Intraoperative image of the posterior femur with the parosteal osteosarcoma. (F) The cortex of the posterior femur after tumour resection. (G) The resected specimen. (H) Postoperative radiographs of the femur after resection of the tumour and insertion of an intramedullary femoral nail.

Differential diagnosis includes other surface bone sarcomas (Table 1) and benign juxtacortical lesions, the most common of which is myositis ossificans (MO) and rarely diaphyseal parosteal osteoma, parosteal lipoma, bizarre parosteal osteochondromatous proliferation (BPOP) and parosteal exuberant fibrous dysplasia. Regarding the differential diagnosis of BPOP and PAO, BPOP is a rare benign exophytic osteochondromatous lesion that most commonly involves the metatarsals, metacarpals and the phalanges of fingers and toes. Rarely, BPOP can affect large tubular bones, seen as a radiodense well-defined lesion, stuck on the bone cortex simulating a parosteal osteosarcoma. However, unlike PAO, it lacks any cortical erosion, periosteal reaction or intramedullary extension on imaging. On the other hand, to the best of our knowledge, PAO does not affect the small tubulars bones of hands and feet. As regards MO, it is not connected with
Clinical presentation

| History            | Demographics | Clinical presentation | Location (most common) | X-rays *First line method | CT *Method of choice for calcifications | MRI *Method of choice for soft tissues | IHC markers Helping molecular findings |
|--------------------|--------------|-----------------------|------------------------|---------------------------|----------------------------------------|---------------------------------------|--------------------------------------|
| Long history (over a year) | Females > males, 3rd decade | Slowly growing bone tumour, occasionally painful | Long bones, metaplasia (distal femur) | Mineralized juxtacortical mass with broad-based stalk 'Cleft sign' frequent | STM Confirm broad-based stuck and central mineralization | Inhomogenous STM with mineralization ± Intramedullary extent, Inhomogenous cartilage cap | SATB2, MDM-2, CDK4 |
| Short duration (weeks–months) | Males = females 2nd–3rd decade | Swelling and/or pain, bone tumour | Diaphysis (femur, tibia) | Broad-based opacity Cortical thickening with sauceration ± perpendicular periosteal reaction within the mass | Same as X-ray, ± STM, additional calcifications | STM, ‡SI on T2w, nodal/septal/palmar enhancement (chondroid type) | MDM-2 and CDK4 overexpression |
| Malignant transformation of a pre-existing osteochondroma (or of multiple osteochondromas) | Prolonged clinical course | Pain, swelling 5–22 cm | Diaphysis or diaphysis-metaplasia of long bones (femur, tibia) | Broad-based opacity Thickened eroded cortex Intramedullary extension | Same as X-ray, ± STM | Inhomogenous mass on T2w images Intermediate myxoid extension | Not relevant |
| | | | | | | | | |

Note: PAO, parosteal osteosarcoma; PEO, periosteal osteosarcoma; HGSO, high-grade surface osteosarcoma; PCS, periosteal chondrosarcoma; SPECS, secondary peripheral chondrosarcoma; CT, computed tomography; MRI, magnetic resonance imaging; IHC: immunohistochemical; STM: soft tissue mass; SI: signal intensity.

Histologically, the tumour is composed of hypocellular areas of spindle cells arranged in fascicles in desmoplastic collagenous stroma with parallel trabeculae of well-formed woven bone (‘streamer pattern’). Spindle cells are characterized by minimal or, less frequently, moderate atypia and low mitotic activity. Areas of cellular matrix, scattered nodules of chondroid cells with atypical morphology and foci of anastomosing and curved bone trabeculae as in fibrous dysplasia can occur. Its low-grade appearance and positivity for MDM-2 and CDK4 are key features in differentiating it from other juxtacortical sarcomas (Fig. 3).

Dedifferentiation of PAO can occur in about 15–43% of cases at the time of first diagnosis. Dedifferentiated PAO is an aggressive high-grade sarcoma with worse prognosis, metastases and high rates of recurrence usually in the form of HGSO, undifferentiated spindle cell or pleomorphic sarcoma with biological behaviour similar to conventional osteosarcoma. Although different, well-differentiated and dedifferentiated areas often coexist.

Characteristic imaging findings include tumour size larger than 11 cm, deeper invasion of the medullary canal and...
large intralesional radiolucencies on X-rays or CT.\textsuperscript{13,17} Moreover, novel evidence suggests that dedifferentiation can be correlated with the amplification and expression status of MDM-2 and CDK4.\textsuperscript{31}

The treatment of choice is wide resection with survival rates at five years of approximately 90\%.\textsuperscript{10} Local recurrence may occur when wide resection is inadequate or in case of dedifferentiation.\textsuperscript{32,33} Zaikova et al, in their review of 63 patients, reported a local recurrence rate of 46\% following intralesional excision as compared to 20\% following marginal and 0\% following wide excision.\textsuperscript{34} Local recurrence is usually seen as a heavily ossified mass.\textsuperscript{33}

Patients of earlier stages are amenable to hemicortical resection with or without prophylactic fixation. Endoprosthesis, extracorporeal radiation followed by reimplantation or resection arthrodesis could be the alternative methods of surgical reconstruction. Both methods have been found to result in a reasonable functional outcome even in later stages of the disease.\textsuperscript{35} The technique of hemicortical resection for treating PAO was first described by Campanacci et al,\textsuperscript{36} creating a unicortical window with wide margin. The defect is reconstructed using either bone cement, autograft such as fibular autograft, allograft or pasteurized/autoclaved/irradiated host bone.\textsuperscript{37} None of their patients developed local recurrence. Treatment of this aggressive tumour is wide resection. Chemotherapeutic agents offer little or no benefit; however, several authors recommended chemotherapy in cases of dedifferentiation,\textsuperscript{17} medullary involvement\textsuperscript{17,36} or lung metastasis.\textsuperscript{11}

The prognosis of PAO is better compared to other surface bone sarcomas and conventional osteosarcoma, with metastasis and recurrence occurring in rare cases.\textsuperscript{10,38}

**Periosteal osteosarcoma (PEO)**

PEO is a chondroblastic, intermediate to high-grade malignant bone-forming tumour, arising from the inner germinal periosteal layer with periosteal reaction.\textsuperscript{11} The recent WHO classification does not recommend the term juxtacortical chondroblastic osteosarcoma that has been used in the past.\textsuperscript{6}

This surface bone sarcoma is less common than PAO and accounts only for 1.5–2.0\% of all osteosarcomas. Slight male predominance is observed, and the tumour mostly affects the second and third decade of life, with peak incidence in the second decade.\textsuperscript{39,40} The diaphysis of tibia and femur are the most affected sites, whereas other long and flat bones have been sporadically reported.\textsuperscript{40–42} Patients complain of a painful swelling or just pain for a shorter period of time compared to PAO.\textsuperscript{6}

Typical radiologic appearance of PEO is a broad-based soft tissue opacity causing a shallow crater of the outer bone cortex; so-called saucerization. Calcified spiculae running perpendicularly from the bone surface within the mass are common and represent periosteal reaction of ‘sun burst’ type.\textsuperscript{5,16} On cross-sectional imaging, the soft tissue mass is well margined without pseudo-capsule and surrounds approximately 50\% of the bone circumference.
an image-guided biopsy. The diagnosis mainly includes HGSO and periosteal chondro-osseous lesion (Table 1). The diagnosis is usually confirmed by radiological differential characteristics and biological behaviour. It occurs more frequently in males than females, during the second and third decades of life. It commonly arises in the diaphysis or metaphysis of long bones, with the femur, tibia, and humerus being the most affected, and usually measures 5 to 22 cm at initial presentation. Pain and swelling are the most common symptoms. Radiographic descriptions are limited and refer to a broad-based surface bone tumour which occasionally may look similar to a PEO, but in contrast to the latter, it surrounds the bone in more than 50% of its circumference, is usually denser and extends to the intramedullary cavity. The soft tissue component is inhomogenous on T2w MR images, featuring a high-grade sarcoma (Fig. 5A-C). The exact pathogenetic mechanisms of HGSO have not been elucidated yet. Cases developed on PAO display amplification of MDM-2 and CDK4. Histologic features are those of high-grade conventional osteosarcoma, with anaplastic-pleomorphic neoplastic cells that show numerous mitoses, including atypical ones, and foci of bone formation in close proximity to the neoplastic cells. Neoplastic population can have plasmacytoid, epithelioid or fusiform appearance or attain a smaller size when closer to bone matrix. Bone formation can take the form of trabeculae composing large sheets of compact bone or more disorganized trabeculae. HGSO may be osteoblastic, chondroblastic or fibroblastic, and typically contains neoplastic cartilage and/or fibroblastic components. It is usually easily distinguished due to its high-grade features (Fig. 6). The tumour should not contain low-grade elements.

The treatment of choice is a combination of wide surgical excision and adjuvant chemotherapy. Chemotherapy protocol is similar to conventional osteosarcoma including cases with metastases. The prognosis for HGSO is worse than for the other two types of surface osteosarcoma and is similar to that of conventional osteosarcoma. As a high-grade tumour, it is highly proliferative and may present with satellite lesions and early metastases. Local recurrence is significantly associated with marginal excision.

**High-grade surface osteosarcoma (HGSO)**

HGSO accounts for less than 1% of all osteosarcomas. As a high-grade lesion, it bears similarities to conventional intramedullary osteosarcoma regarding its clinicopathological characteristics and biological behaviour. It occurs more frequently in males than females, during the second and third decades of life. It commonly arises in the diaphysis or metaphysis of long bones, with the femur, tibia, and humerus being the most affected, and usually measures 5 to 22 cm at initial presentation. Pain and swelling are the most common symptoms. Radiographic descriptions are limited and refer to a broad-based surface bone tumour which occasionally may look similar to a PEO, but in contrast to the latter, it surrounds the bone in more than 50% of its circumference, is usually denser and extends to the intramedullary cavity. The soft tissue component is inhomogenous on T2w MR images, featuring a high-grade sarcoma (Fig. 5A-C). Pain and swelling are the most common symptoms.

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Overall prognosis is affected by the grade of the tumour, by the response to neoadjuvant chemotherapy and by the presence of local recurrence. Medullary involvement is not shown as an independent prognostic factor. Overall five-year survival rate is 62%. Extension to the medullary canal is rare and associated with worse prognosis. The Rizzoli Institute study reports five-year overall survival of 82% and disease-free survival of 70%. Adequate response to neoadjuvant chemotherapy and localized disease without metastasis are factors that favour long-term survival.

**Periosteal chondrosarcoma (PCS)**

PCSs are malignant hyaline cartilage tumours arising within periosteum, with fibrous pseudo-capsule formation in touch with it and large size, usually greater than 3 cm.
It is a locally aggressive, though low-grade, malignant tumour. Histological grading is not applicable.6

PCSs represent less than 2–4% of all chondrosarcomas53 and 0.2% of all bone tumours.54 The most commonly involved bones are the distal femur at the metaphyseal or diaphyseal-metaphyseal region or the proximal humerus,5,54–56 followed by the proximal femur, tibia, iliac bones, maxillofacial region, rib, fingers or foot.53 They usually occur in the second to fourth decade of life,54 with a peak in the third decade57 and they are more common in males.54 Information about PCS is scarce, with the largest published series including 36 patients.58 Clinical presentation is non-specific, and symptoms include a painless mass, swelling, deterioration of function and a prolonged clinical course.59

Radiological features of PCS include a large soft tissue mass with a broad-based, non-calcified attachment to bone surface.52,54 The lesion is sharply delineated from the adjacent cortex and contains chondroid-type mineralization with ring and arcs. The bone cortex can appear thickened or thinned and saucerized (Fig. 7A). Chronic periosteal reaction, such as buttressing, is frequent, reflecting the slow development of the tumour, but Codman triangles may be occasionally seen. CT can confirm the chondroid type of internal mineralization and additionally show thin interlobular calcifications and a calcified shell that partially surrounds the tumour. MRI demonstrates a cartilaginous-type tumour, isointense to muscles on T1w MR images (Fig. 7B) and typically hyperintense on T2w images with low signal intensity calcifications, multilobulated contour (Fig. 7C) and peripheral and septonodular enhancement. Intramedullary extension or bone marrow oedema are exceedingly rare, unless the tumour is dedifferentiated.59

A study about the molecular background of the tumour57 depicted loss of canonical Wnt signalling and deregulation of pRb signalling by loss of p16 expression. Mutations in IDH1 and IDH2 genes are documented in a number of PCS.

Histologically, variably sized neoplastic chondroid lobules which invade the cortex, with moderate cellularity and no or mild cellular atypia, are the morphologic hallmark. Foci of myxoid matrix, fibrous bands with small vessels, calcification and endochondral ossification may be recognized, along with metaplastic bone formation in the periphery of

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**Fig. 7** Periosteal chondrosarcoma (PCS) in a 17 year-old female. (A) Anteroposterior radiograph shows a broad-based soft tissue mass (arrows) at the proximal metadiaphysis of the of the left humerus, causing endosteal scalloping. (B&C) T1w coronal magnetic resonance (MR) image shows a mass (arrows) isointense to muscles (B) and hyperintense with a microlobular contour on a coronal fat-suppressed MR image (C). Cortex seems intact. There is a non-marginated area in the adjacent bone which is hypointense on T1w and hyperintense on fat-suppressed T2w image that corresponds to bone marrow oedema, as no malignant infiltration of the medulla was documented on pathology of the specimen (arrowheads). Barely seen punctuate and curvilinear calcifications at the periphery and within the mass (arrows) signifying cartilaginous matrix. (D&E) Intraoperative photographs showing the periosteal mass before (D) and after resection (E).
the neoplasm. Osteoid or bone formation is not noted. In some cases there is invasion of the bone medulla.

Dedifferentiation of a low-grade PCS may occasionally occur. In this case MR imaging reveals a bulky non-mineralized soft tissue component with mixed-signal intensity on T2w images. 60 Differential diagnosis of PCS predominately includes PEO (Table 1) and periosteal chondroma (PC). PCs are more common than their malignant counterparts and usually appear in a younger age group; both PC and PCS share similar imaging appearances. Robinson et al found that size was the most reliable discriminating feature between PCS and PC: reported size of PC ranged between 1.0 and 6.5 cm, whereas that of PCS ranged between 3 and 14 cm. 52 Histologic evaluation of a periosteal cartilaginous tumour has been advocated for a tumour diameter larger than 3 cm. 52,61 PCSs usually present with invasion of the cortex and in some cases of the bone medulla; findings critical in distinction with PC.

Wide surgical resection is the treatment of choice, whatever the grade of the lesion. 56,58 Smaller tumours (less than 3 cm) can be treated with marginal excision with close follow-up (Fig. 7D, Fig. 7E). 52,54,62 Incomplete excision is associated with local recurrence. In case of medullary involvement, treatment guidelines for central chondrosarcoma should be followed. Goedhart et al recommended follow-up with plain radiographs for the next five years. 58 A baseline MRI can be performed after six months and again at two years. 58 The prognosis of PCS is better compared to conventional chondrosarcoma of the same histologic grade. 63 Invasion of the medullary cavity is unusual. Metastases are rare, occur late and have only been reported in grade II and III lesions. 63 The most common site of metastasis is the lungs, and rarely the lymph nodes and skin. In a retrospective review of 24 patients, Papagelopoulos et al have shown that the overall five-year metastasis-free survival was 83%. However, six out of 24 patients died of pulmonary metastases in a mean follow up of 17 years. 62

Secondary peripheral chondrosarcoma (SPECs)

SPECs are malignant cartilage-producing tumours. They develop on the grounds of malignant transformation of a pre-existing osteochondroma, the most common benign cartilaginous lesion of adolescence. 64 They occur as solitary or multiple (hereditary) cartilage-capped bony projections from the metaphysis of endochondral bones adjacent to growth plate. 65 Less than 1% of patients with sporadic osteochondromas may develop SPECs, despite reports of rates as high as 7.3% coming from large referral centers. 67 Of patients with multiple osteochondromas, 1–3% will eventually develop SPECs. 68 Ahmed et al reported that most cases of progression occurred in patients with multiple tumors. 67 Most cases of SPECs are low to intermediate grade, although tumours of higher grade are also possible.

The tumour is far more common after maturity, usually 25 to 45 years of age. 69 The most affected bones are the pelvic bones and the shoulder girdle. Typical presentation is a growing painful mass on an underlying osteochondroma, after skeletal maturation. Sudden onset of pain and increase in the size of the swelling may be hints of malignant transformation. 70 These patients can present with longstanding symptoms lasting between one and two years.

On radiographs, SPECs exhibit lytic areas of the stalk of the pre-existing osteochondroma, irregular surface and an adjacent radiopaque mass with chondroid-type calcifications. 60,67 Cross-sectional imaging can document the exact size and origin of the mass, the type of calcifications and the anatomic relation of the mass with the adjacent soft tissue structures. It is particularly useful to delineate tumours in complex anatomic areas such as the pelvic and shoulder girdle where X-rays are of limited value because of the superimposition of anatomic structures. 59 The soft tissue component is typically lobulated, hypodense on CT and hyperintense on T2w MRI, whereas calcifications are hypointense on all MR sequences (Fig. 8A–C). A painful

Fig. 8  A 42–year old man with abdominal discomfort and deteriorating left hip pain during the last six months. (A) frontal radiograph of the pelvis shows a bulky soft tissue mass (arrows) with typically cartilaginous rings and arcs calcifications, occupying the left pelvis. A densely mineralized lesion is seen at the left acetabular roof (arrowheads). (B) A computerized tomography (CT) image (bone window) shows an exostosis with lytic areas within it (arrow) of the left innominate bone protruding anteriorly and medially. A space-occupying soft tissue mass with calcified spots (arrows) seems to originate from the osseous protuberance and is displacing the adjacent left wall of the urinary bladder. (C) The mass is typically hyperintense on a fat-suppressed T2w axial image (arrows) and the calcifications are hypointense (arrowheads).
osteoochondroma after skeletal maturation, with enlargement of the cartilaginous cup beyond 2 cm should raise the suspicion of SPECS.60,71 Frequently, a large secondary bursa develops over the top of an osteochondroma, which may share similar imaging features with an abnormally thickened cartilaginous cap;72 however, a bursa has a more saccular than lobulated shape,72 whereas application of specific cartilage sequences differentiates between fluid and chondroid tissue.71

Recently, Tsuda et al stated that preoperative biopsy correctly predicted the histological grade after excision in only 27% of patients with SPECS, so it was difficult to correctly estimate grading of these tumours prior to surgery.69 Both types of SPECS show a propensity to develop in patients with multiple osteochondroma syndrome (5% in comparison to 1% in patients with solitary osteochondroma), who carry germline mutations in EXT1 or EXT2. In atypical chondromatous tumour/chondrosarcoma grade 1 (ACS/CS1), cell population is composed of cells in which EXT1 or EXT2 is biallelically inactivated, which is the characteristic alteration of osteochondroma, and cells that retain at least one functional copy of EXT1 or EXT2, with coexistence of EXT-mutant alleles and EXT-wildtype alleles. In contrast, in high-grade chondrosarcoma the EXT-wildtype cells predominate. Conclusively, data suggest that other factors play their part in tumorigenesis, such as alterations in genes that regulate cell cycle such as CDKN2A in ACS/CS1 or in the p53 and RB1 pathways in high-grade chondrosarcoma, as EXT-wildtype cells are susceptible to progression, given that they are more numerous than the EXT-mutant cells.5 In ACS/CS1, chondroid cells attain a lobular pattern with histologic features such as cystic changes, necrosis, binucleated cells and increased vascularization being common, but not helpful in the differential diagnosis from osteochondroma. Tumour cell nodules might be seen in the soft tissue, without connection to the main tumour, and calcifications are easily seen. Morphologic signs of the pre-existing osteochondroma are often easily detected. Invasion of the stalk is rare and indicative of progression. Differential diagnosis of osteochondroma, ACS/CS1 and progression of osteochondroma to ACS/CS1 on histologic grounds alone is not feasible and clinical as well as imaging correlation is crucial for correct diagnosis.

Peripheral chondrosarcoma (PECS) grade 2/3 is characterized by lobular configuration and increased cellularity, with evident mitoses, nuclear size variation and prominent nucleoli (Fig. 9). Nuclear condensation and small nuclear size or binucleation may be present. The cartilaginous matrix might show myxoid changes and at the periphery the neoplastic cells can attain spindle morphology. Endochondral ossification may be noted, which is feature of the pre-existing osteochondroma.

Fig. 9 (A) Hematoxylin-eosin stain (X40) and (B) hematoxylin-eosin stain (X100): the neoplastic population is composed of chondroblastic cells of moderate atypia widely invading bone cortex. (C) Hematoxylin-eosin stain (X100): the neoplastic cells extend to the adjacent soft tissue, invading skeletal muscle. (D) Hematoxylin-eosin stain (X200): on high power view, the chondroblastic neoplastic population is characterized by moderate cellularity and moderate atypia. Reactive bone formation on the grounds of periosteal reaction is noted on the left. Given that the tumour mass is located on bone surface, the morphological findings are consistent with PECS grade 2.

Note. PECS, peripheral chondrosarcoma.
Differential diagnosis from osteochondroma and ACS/CS1 is based on morphologic characteristics of the lesions, as nuclear pleomorphism and mitoses are encountered only in PECS. No osteoid or bone formation is noted, and the lesion is located at the surface of the bone, tumour characteristics that are useful in distinction from PEO and central chondrosarcoma, respectively.6

Surgical excision with wide margins is the treatment of choice. Tsuda et al recommend that a secondary chondrosarcoma arising from osteochondroma of the pelvis must be resected with wide/radical resection margins. Resection with wide/radical surgical margins is important to minimize the risk of local recurrence, especially for patients with high-grade tumours and hereditary multiple exostoses. However, the morbidity of surgery and the risk of local recurrence can be balanced in limb secondary chondrosarcomas, which show low risk of death and metastasis. Wide/radical margin was associated with improved local-recurrence-free survival (p = 0.032) and local recurrence was associated with worse disease-specific survival (p = 0.005).69 Distant metastasis is uncommon, and prognosis is favourable for most patients. Overall survival at five years is approximately 90%.71 Local recurrence remains a significant problem for approximately 10–20% of patients. In the study by Tsuda et al, 29% of patients developed local recurrences.69 However, other authors have reported higher rates of local recurrence (16–52%).67,68 Patients with SPECS of the pelvis are especially at risk for local recurrence.73 In the study by Tsuda et al, a total of 51 patients with SPECS occurring from osteochondromas were reviewed. The ten-year disease-specific survival for all patients was 89.4%. Local recurrence occurred in 15 patients (29%), more commonly in pelvic (37%) compared with limb tumours (19%). Four patients with pelvic tumours died from progression of local recurrence. No patient with limb tumour died of disease.69

**Periosteal Ewing sarcoma**

Periosteal Ewing sarcoma (PES) is a very rare surface malignant bone tumour, comprising 3% of all Ewing sarcomas. The male-to-female ratio is 2.2:1 with a peak incidence in the second decade of life. The femur is the most common site of PES. Pain and a palpable mass are the most frequent symptoms. Symptoms and signs similar to those of infection such as fever, leucocytosis, malaise, local reddening, heat, and dilated blood vessels over the palpable mass may be noted. In terms of localization and imaging, it is similar to PEO involving the diaphysis or metadiaphysis of long bones, inciting extrinsic cortical erosion and lacking intramedullary involvement on imaging. Solid periosteal reaction such as Codman triangle type may be seen. However, PES lacks matrix mineralization, which is typical for periosteal osteosarcoma and is moderately hyperintense on water-sensitive MR images, unlike the chondroid-type hyperintensity of PEO.5

The histopathologic features of PES are the same as in medullary or extra-skeletal forms of Ewing sarcoma, which are characterized by small round cells with round and centrally located nuclei. CD99 membranous positivity is essential for Ewing sarcoma diagnosis. The tumour is also characterized by presence of FET-ETS fusions.6 Treatment of PES involves chemotherapy, radiation therapy, and surgical excision with wide excision, although segmental diaphysis removal has been reported.74

**Conclusions**

Surface bone sarcomas are rare. A multidisciplinary approach is essential, as the combination of clinical information, radiological features, molecular studies and histopathological findings leads to correct diagnosis and patient handling. There are still numerous issues regarding the biology, pathophysiology and treatment options. The treatment implications of an accurate and early diagnosis are of paramount importance.

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