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Periosteal Osteosarcoma Arising from the Rib and Scapula: Imaging Features in Two Cases

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Periosteal osteosarcoma is an extremely rare chondroblastic osteosarcoma in the flat bone. There were authors reporting of two cases of periosteal osteosarcoma in the highly unusual sites. One of them arose from the rib, in a 17-year-old male, which appeared as a hypodense juxtacortical mass with periosteal reaction on CT. The other one arose from the scapula, in a 17-year-old female, which showed the intermediate signal intensity (SI) on T1-weighted image (WI), heterogeneous high SI on T2WI, and rim-enhancement on contrast-enhanced T1WI with cortical destruction on MRI.

Index terms: *Bone surface tumor; Osteosarcoma; Rib; Scapula; Chest wall*

INTRODUCTION

Periosteal osteosarcoma (PEROSA) is an intermediate-grade malignancy arising on the surface of bones, frequently in femur and tibia, followed by ulna and humerus in extremities (1, 2). PEROSA of the chest wall, unlike that of the extremities, presents a diagnostic challenge because of the extreme rarity of the lesion, variable clinical symptoms, and suboptimal visualization on radiography of the characteristic features, resulting in misinterpretation of the lesion as a variety of other diseases (3). We present with radiologic findings and review of literature, of the two cases of PEROSA arising from the rib and the scapula.

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CASE REPORTS

Case 1

A 17-year-old male presented with a painless swelling on the right lateral chest wall with duration of 3 months. The past medical history was unremarkable. The patient did not recall any traumatic episodes that preceded his discomfort. On physical examination, a hard, non-movable, and non-tender mass (5 x 6 cm in size) was palpated on the area. The hematologic and biochemical studies were normal.

The chest radiography showed a local soft tissue bulging with 4-cm in size in the vicinity of the right 7th rib. A poorly defined triangular calcific opacity, like a Codman's triangle, was detected on the outer surface of the lateral arc of the right 7th rib. There was no evidence of the rib destruction (Fig. 1A). On the computed tomography (CT) with bone setting, a small juxtacortical hypodense mass was seen on the surface of the rib. The underlying rib showed the periosteal reaction with fine perpendicular spicules extending from the thickened cortex (Fig. 1B). The CT with contrast enhancement showed the fine septal and wall enhancement of the hypodense soft tissue mass with periosteal reaction (Fig. 1C). No definite evidence of cortical destruction or medullary invasion of the rib was seen on either chest radiograph or CT scan. Also, there were

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no abnormalities in the lung parenchyma. The pre-operative differential diagnoses include the primary bone surface tumor (such as periosteal osteosarcoma or chondrosarcoma), soft tissue tumor with secondary periosteal reaction, and tuberculosis.

The patient underwent segmental wide resection of the 7th rib, including the 6th through 8th ribs. The tumor of gross specimen measured 4.5 cm in longest diameter, and it was firmly attached to the outer surface of the 7th rib. Gross specimen in cut-section showed surface tumor of the

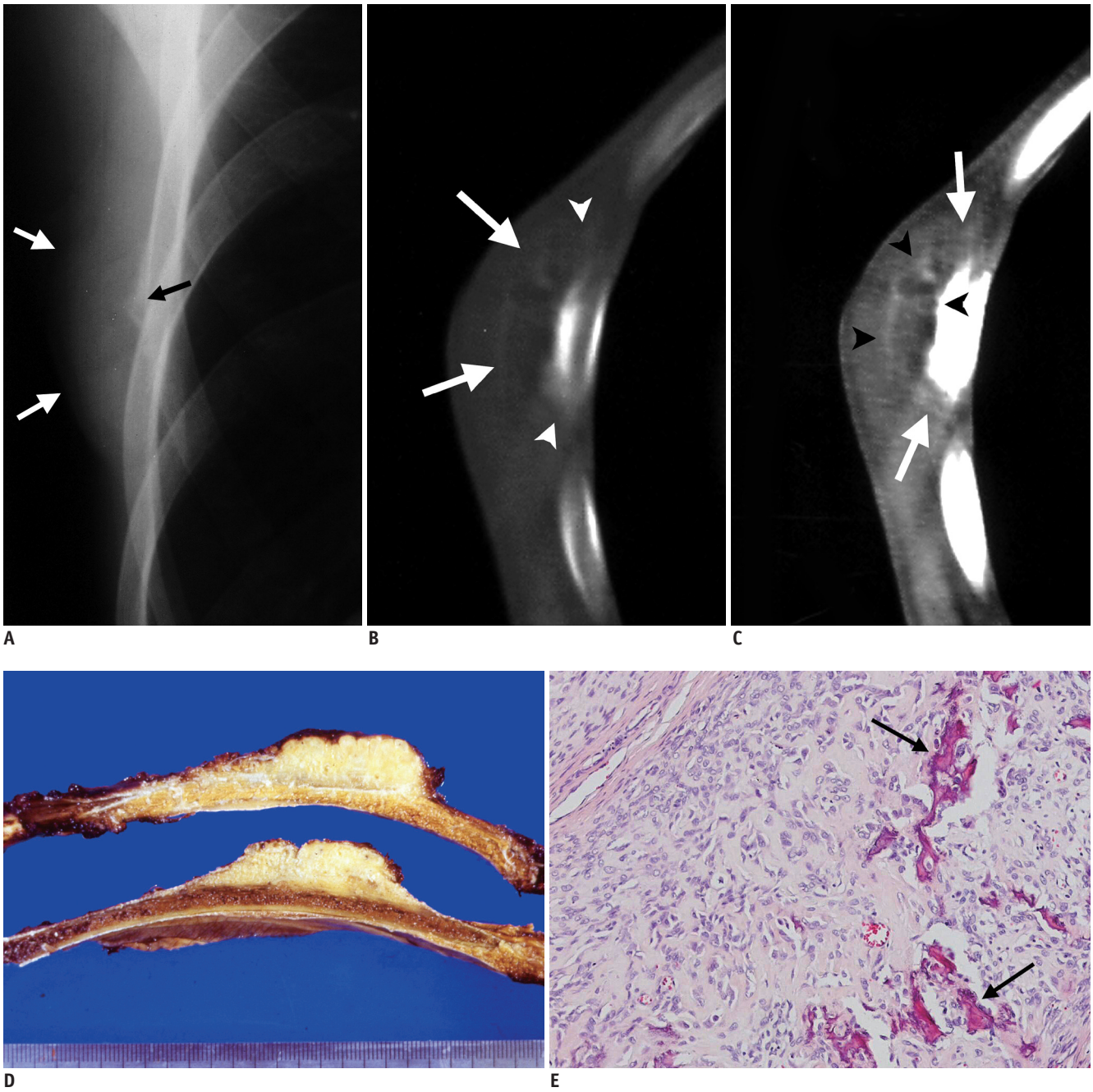

Fig. 1. 17-year-old male with periosteal osteosarcoma in right 7th rib.
A. Simple chest radiograph shows poorly defined triangular calcific opacity (Codman's triangle, black arrow), with periosteal reactions (white arrows) on outer surface of lateral arc of right 7th rib, without evidence of rib destruction. **B.** Non-enhanced CT scan with bone setting shows thin-walled, juxtacortical, hypodense mass (arrows) and periosteal reaction with fine, perpendicular spicules extending from thickened, adjacent cortex of rib (arrowheads). **C.** CT with contrast enhancement demonstrated fine septal and wall enhancement of hypodense mass (black arrowheads) and periosteal reaction with cortical thickening (white arrows). **D.** Gross specimen in cut-section shows surface tumor of rib with medullary infiltration. **E.** Microscopic examination shows definite areas of calcified tumor osteoid (arrows) within chondroid matrix (hematoxylin and eosin stain, x 100).

rib with medullary infiltration (Fig. 1D). The CT examination of the specimen demonstrated a juxtacostal mass with an elevated periosteum, a Codman's triangle, and suspicious medullary involvement. Finally, the tumor was proved to be PEROSA microscopically with medullary extension, predominantly consisting of the chondrosarcomatous components and definite areas of the calcified tumor osteoid (Fig. 1E). Over the 5 years of follow-ups, the patient remained free of recurrent or metastatic disease with no adjuvant therapy.

Case 2

A 17-year-old female presented with a painful mass in the right chest wall with the duration of a month. The patient

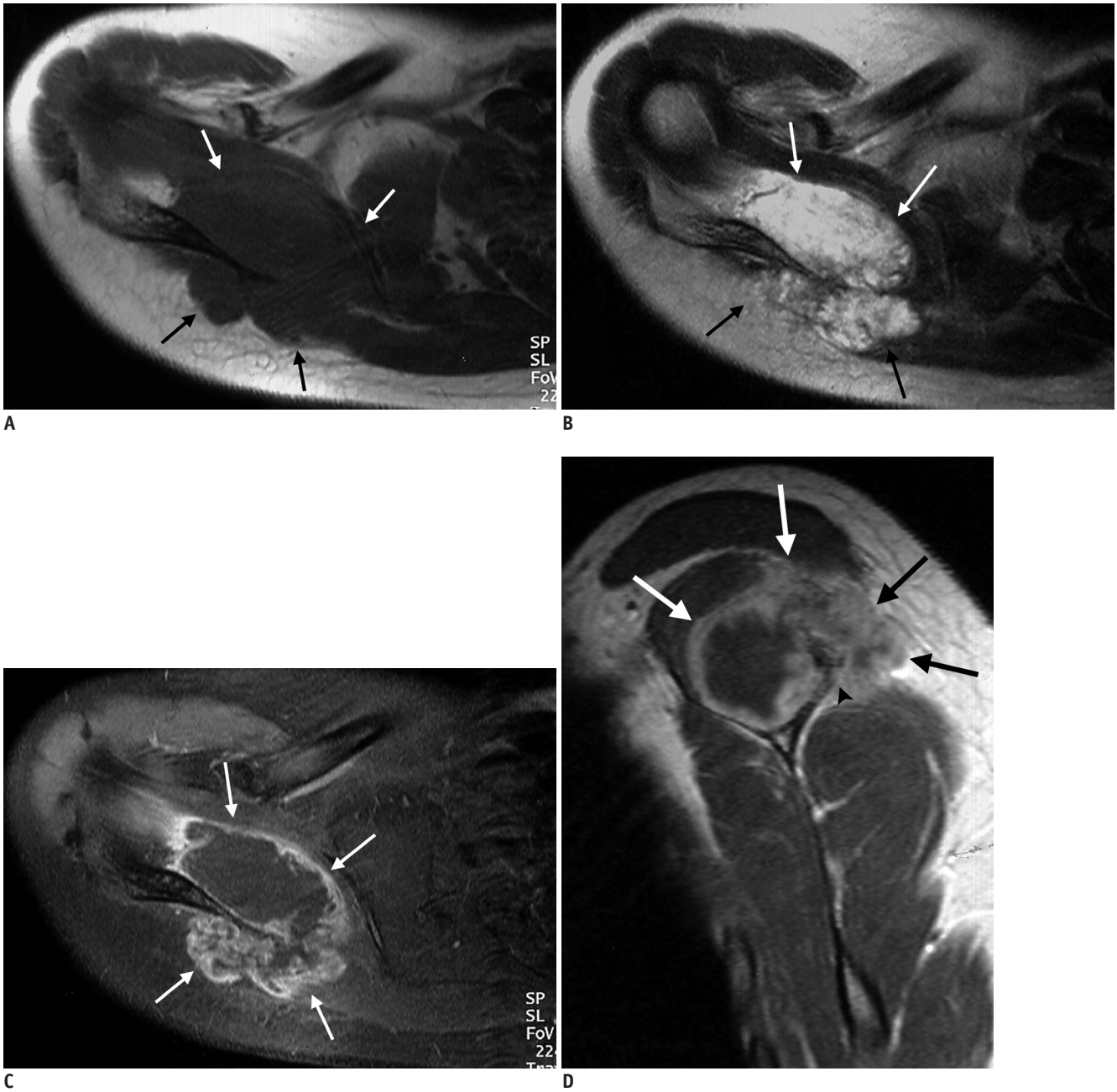

Fig. 2. 17-year-old girl with periosteal osteosarcoma in right scapula.
A, B. MR imaging shows intermediate signal intensity in mass on T1-weighted image (**A**, arrows) and heterogeneous high signal intensity on T2-weighted image (**B**, arrows). **C.** Axial scan of fat-saturated and contrast-enhanced T1-weighted image shows rim enhancement of mass (arrows). **D.** On sagittal scan of contrast-enhanced T1-weighted image, mass is mainly seen located in supraspinatus muscle in suprascapular fossa (white arrows), with cortical destruction of underlying scapular spine (black arrowhead), and partially extending posteriorly to subcutaneous fat layer (black arrows).

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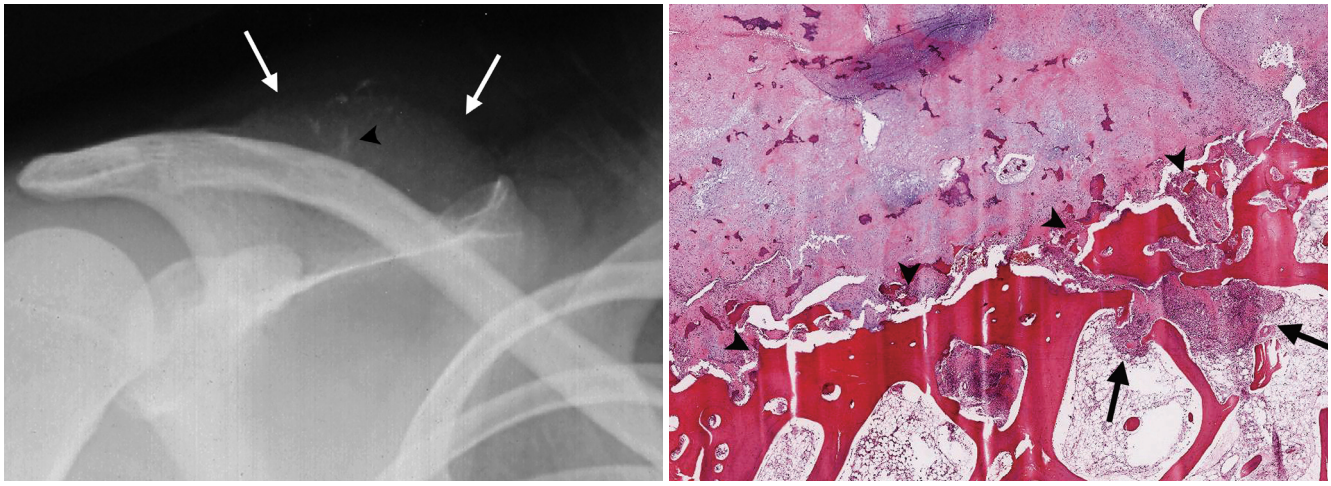

E F

Fig. 2. 17-year-old girl with periosteal osteosarcoma in right scapula.
E. Post-MR fluoroscopy on craniocaudal projection, with patient in supine position, well-depicts soft tissue opacity (white arrows) with inner calcified matrix (black arrowhead) in suprascapular fossa. **F.** Microscopically, cortical bony erosion (arrowheads) and focal intramedullary involvement (arrows) are present (hematoxylin and eosin stain, x 10).

did not have any previous trauma, and she accidentally found the mass while taking a shower. The past medical history and laboratory findings were unremarkable. The physical examination revealed a firm, tender mass (2 x 2 cm in size) on the posterosuperior aspect of the right shoulder. There was no limitation in the range of motion or sensory change of the right shoulder.

A posteroanterior view on a radiograph of the shoulder showed ill-defined and increased soft tissue density in the superior aspect to the right scapula. On MR imaging, the mass showed intermediate signal intensity on T1-weighted image (WI), compared to that of the chest wall muscle (Fig. 2A), and heterogeneous high signal intensity on T2WI (Fig. 2B). The rim enhancement of the mass was seen on the axial, fat-saturated, contrast-enhanced T1WI (Fig. 2C). On sagittal scan of the contrast-enhanced T1WI, the mass was seen to be located in the supraspinatus muscle of the suprascapular fossa, and it looked to be attached to the superior margin of the underlying scapular spine with some cortical destruction. The mass was also observed to partially extend posteriorly to the subcutaneous fat layer (Fig. 2D). Fluoroscopy in craniocaudal projection, with the patient in the supine position, well-depicted the soft tissue opacity containing calcified matrix (Fig. 2E). The pre-operative differential diagnoses included the primary surface bone tumors, such as periosteal osteosarcoma or conventional chondroblastic osteosarcoma, and the soft tissue tumors with secondary marrow invasion.

Incisional biopsy was performed for the mass, and the biopsy specimen demonstrated a malignant tumor with chondrosarcomatous features. One week later, a wide excision of the tumor with partial scapulectomy was performed. At the operation, the mass was mainly found in the supraspinatus muscle, attached to the scapular spine, with some portion extending posteriorly to the subcutaneous layer, which correlated to the preoperative imaging findings. Gross specimen showed a 6 x 4 cm-sized solid mass, with gray to yellowish appearance, and cartilage formation in the surface of the cut. The sagittal CT scan of the specimen also revealed a soft tissue mass with calcifications. The tumor was pathologically confirmed as PEROSA, which is predominantly composed of moderately differentiated chondrosarcomatous components and tumor osteoid. The focal intramedullary involvement was noted (Fig. 2F). The patient received adjuvant chemotherapy for 6 months, and there were no evidence of recurrence or metastasis for the subsequent 5 years.

DISCUSSION

To our knowledge, PEROSA is extremely rare in flat bones (1). In the study of Burt et al. (4) with 38 cases of chest wall osteosarcoma, 13 cases (34%) including the post-radiation osteosarcoma arose in the rib and 12 cases (32%) arose in the scapula, but no PEROSA was found. We searched through medical literature published in English before August 2013, in PubMed website with keywords such as 'periosteal osteosarcoma', 'rib', 'clavicle', 'scapula', and 'sternum'. There was only four reported cases of PEROSA involving the chest wall: two in the clavicle (5); one in the

rib (2); and one in the scapula (6).

Classical radiologic features of PEROSA of the long bones have been well-known. The tumor appears as a soft tissue mass that is broadly attached to the bony cortex, which shows cortical thickening, scalloping, and perpendicular spiculation of the mineralization. It elevates the periosteum to produce Codman's triangles and fusiform surface enlargement (7). However, with simple radiograph, the characteristic features of the PEROSA arising from the rib or scapula may be visualized suboptimally, and may be misinterpreted as a lung cancer, pleural lesion, and other variety of disease (2, 3, 6, 7). The radiologically differential diagnoses of PEROSA include fracture with callus, periostitis, heterotopic ossification, calcific tendinopathy, other primary bone surface tumors (parosteal osteosarcoma, periosteal chondrosarcoma, high-grade surface osteosarcoma), metastatic carcinoma, and tuberculous infection of the chest wall (2, 3). The post-fracture callus tends to have a more conspicuous and orderly deposition of the osteoid and immature bone (2). The parosteal osteosarcoma shows a well-defined, lobulated osseous mass attached to the bone by a narrow stalk and the characteristic radiolucent line interposed between the mass and subjacent cortex (7). Periosteal chondrosarcoma tends to be a round mass with extensive granular opacities and the underlying cortex showing saucer-like depression with thickening and sclerosis (8). The high-grade surface osteosarcoma usually arises in the diaphysis and they show perpendicular periosteal reaction, which makes the differentiation with PEROSA difficult. However, unlike PEROSA, the high-grade surface osteosarcoma tends to surround the bony circumference more extensively (7). The tuberculosis of the chest wall has a wide spectrum of radiologic findings of commonly encountered juxtacortical lesion, with or without the bone destruction, which makes it difficult to distinguish from PEROSA, especially in endemic areas. The rarity of the incidences of PEROSA on the flat bone also leads to differential diagnosis.

CT may provide valuable information such as assessment of tumor origin, tumor matrix, calcification within soft tissue mass, and localization of the masses. This indicates of the differentiation from more common lesions that may be confused on radiographs (3). Although our case on the rib did not show characteristics of calcification, the CT is useful for differentiation of chondroid tumor, in which the hypodense chondroid matrix and punctate calcifications are some of the specific findings.

The MR imagings may reveal more precisely the margin and extent of the mass, and also may provide information for differential diagnosis. PEROSA shows similar signal intensity to the muscle on T1WI and heterogeneous high signal intensity on T2WI, which reflects the predominancy of the chondroid components within the mass. The peripheral or septal patterns of contrast enhancement also suggests the cartilaginous lesion. The adjacent bone may show focal areas of the signal change due to reactive marrow edema. Less commonly, intramedullary invasion of the tumor may be seen as continuity from the soft tissue mass (7). These MR findings are helpful for differentiating PEROSA from the high-grade surface osteosarcoma, which does not show high signal intensity on T2WI or peripheral enhancement, due to lack of high-water content and cartilaginous component (7). However, MR imaging is less sensitive than CT for detection of mineralization in the mass.

Medullary involvement is rarely found in the cases of PEROSA, showing direct continuity between the overlying surface tumor and the area of bone marrow infiltration. It is distinguished from the areas of simple marrow signal change, which shows intervening cortex without definite continuity with the overlying surface tumor. Also, cortical bone adjacent to PEROSA usually appears to be thickened and scalloped, in which the bone permeation or destruction is not a common finding of PEROSA (7). In the study of Hall et al. (1) with 61 cases of PEROSA, the gross medullary extension was observed in 2 cases, microscopic medullary extension in 1 case, and cortical bone permeation in another case. Murphey et al. (7) also reported 1 case of medullary invasion, accounting for 2% of their series. Moreover, Sonobe et al. (9) and Suehara et al. (10) each reported 1 case of PEROSA with medullary invasion. Therefore, with the unusual and distinctive radiologic and histologic findings of PEROSA, the medullary involvement or cortical bone permeation should not be precluded as the diagnosis (1, 7). In our case of the rib, the medullary cavity did not seemed to be involved according to the pre-operative imaging studies including the CT, but the intramedullary involvement was found on the specimen CT and was pathologically proved. In our case with scapula involvement, the intramedullary involvement was suggested on sagittal scan of fat-saturated and contrast-enhanced T1WI, and it was confirmed by the pathologic examination.

Periosteal osteosarcoma is locally aggressive malignant tumor, so that inadequate surgical resection can result in recurrence. Therefore, currently recommended treatment

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for PEROSA consists of wide segmental resection, with or without neoadjuvant chemotherapy (5). The metastatic potential of PEROSA is much lower than that of conventional osteosarcoma, with a rate of distant metastasis of 10–20%, and mostly common to the lung (1). The prognosis of PEROSA is better than that of the conventional intramedullary osteosarcoma and high-grade surface osteosarcoma, but it is worse than that of parosteal osteosarcoma (5).

In summary, we report two rare cases of PEROSA arising from the flat bones of chest wall, the rib, and the scapula. Although diagnosis of PEROSA arising from the rib and scapula is challenging, due to their rarity in location and suboptimal visualization on radiographs, the cross-sectional imaging modalities such as CT and MR are helpful for detection, diagnosis, and differentiation of PEROSA from other chest wall diseases.

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