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Extraosseus Ewing sarcoma: An uncommon periclavicular location

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A rapidly enlarging right sternoclavicular mass in a young male was labeled as a nonspecific mass. MRI played a crucial role in characterizing the lesion, helping to define the possible mesenchymal origin and the relative involvement of the surrounding structures. We also discuss the differential diagnosis of an extraosseus Ewing sarcoma (ES), with its imaging findings.

Case report

A 13-year-old male presented to our department with an enlarging, palpable, painless mass in the region of the right sternoclavicular joint. There was no history of trauma, and there was no restriction of shoulder joint movement.

The patient underwent sonography of the right sternoclavicular region, which revealed a hypoechoic, round lesion in the medial periclavicular soft tissues, approximately 18 mm by 9 mm in size, without internal vascular flow (Fig. 1). Subsequent MRI revealed an expansive oval mass. The lesion measured 3 cm by 1.5 cm by 2.5 cm. It had regular edges, low signal on all the pulse sequences, and no variations after fat suppression (Fig. 2). The lesion was in contiguity with the medial clavicle. There was an osteolytic area of 5 mm with high T2 signal within the trabecular bone of the medial clavicle that extended along the proximal shaft for 2 cm. Moreover, this mass displaced the muscle fibers of the platysma without any significant signal abnormality in the muscle. The other muscle and bone structures were normal, as were the neurovascular bundle and the lymph nodes.

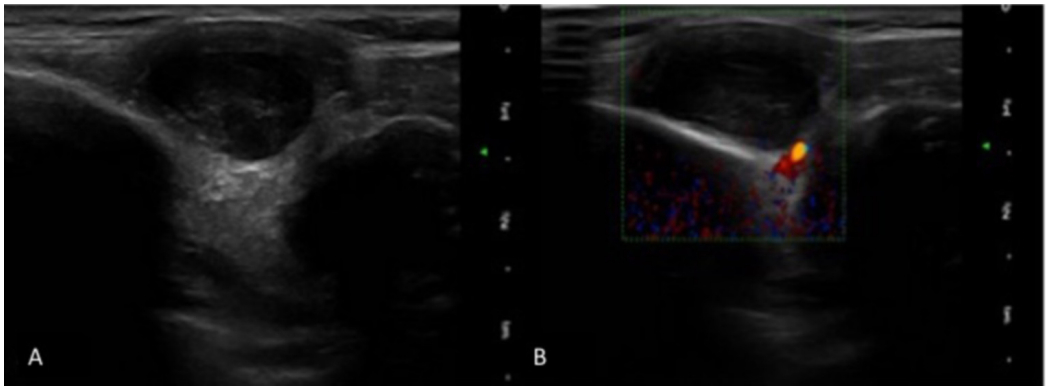

Fig. 1. Ultrasound of the swelling showing a hypoechoic soft-tissue mass surrounding the clavicle (A) without inner vascular signal on color Doppler imaging (B).

A radiograph of the sternoclavicular joint in a right posterior oblique projection confirmed the presence of an osteolytic lesion. We noted the moth-eaten appearance of the metaphyseal region of the clavicle (Fig. 3). After incisional biopsy of the mass, histopathological examination showed

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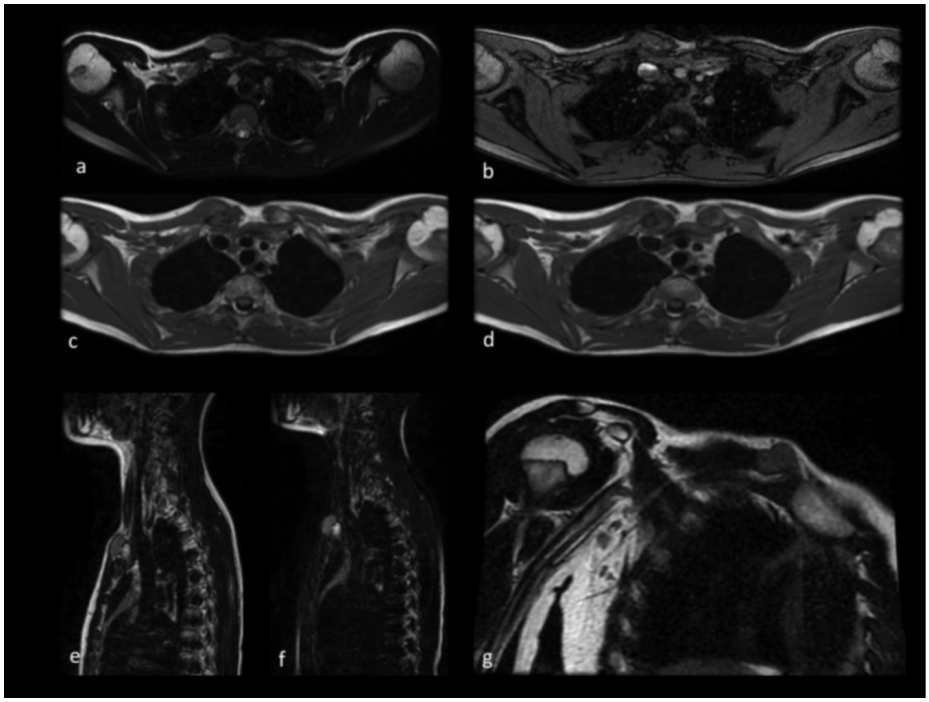

Fig. 2. Axial T2 fat saturated (a) and T1 gradient echo (b) weighted images illustrate the presence of an expansive ovoid mass tightly in contact with the sternal meta-epiphisary side of the right clavicle. Axial T1 (c-d), sagittal T2 and T2 fat-saturated, weighted images (e-f) show the infiltration of the lesion into the ventral side of the clavicle. Note the extension of the tumor with an oblique coronal T1-weighted image (g).

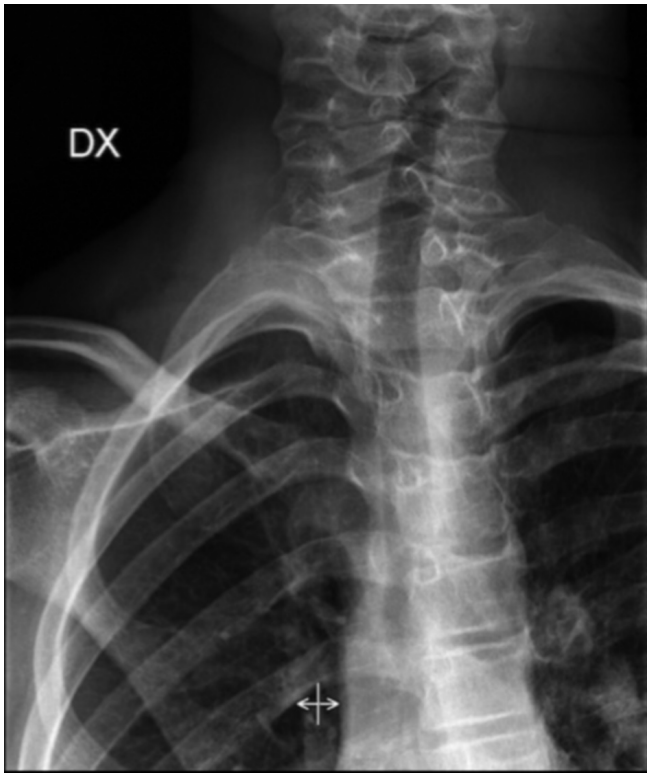

Fig. 3. Right posterior oblique plain film. Note the osteolytic areas at the sternal side of the right clavicle meta-epiphisis.

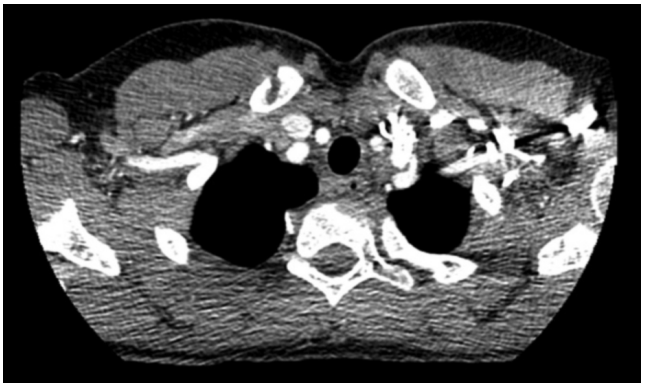

Fig. 4. Axial postcontrast CT slice shows the infiltration of the dysplastic tissue into the bone.

a small-round-cell tumor, undifferentiated, with a vaguely lobular structure woven by fibrous bundles. Periodic acid-Schiff staining revealed prominent cytoplasmic glycogen granulations. Therefore, we made a diagnosis of extraosseus Ewing sarcoma involving the clavicle. A CT scan of the chest was performed for staging; there were no pulmonary nodules, but the bone involvement was confirmed (Fig. 4).

Discussion

Ewing sarcoma is a poorly differentiated neoplasm composed of small-round-blue cells; it was originally described in 1921 by James Ewing (1). Often presenting as a painful, rapidly growing soft-tissue mass 5-10 cm large, it typically arises from the medullary cavity with invasion of the Ha-

versian system and has been described in virtually every bone in the body. It more commonly affects young patients beteen 10 and 30 years of age (2, 3) with a predilection for the male sex (1.5 to 1) (4). Primitive neuroectodermal tumor (PNET) is a related lesion, and Ewing sarcoma and PNET are considered to be ends of a histological spectrum of Ewing's family of tumors (5).

Extraskeletal locations of Ewing sarcoma are less frequent (25%, according to Balamuth et al.) and include the lower extremities (32%), the paravertebral region, (15%), the chest wall (11%), and the retroperitoneum (11%) (6, 7).

To the best of our knowledge, the imaging features of extraskeletal ES have not been extensively described, and it does not always show the characteristic appearances of other types of ES (8-10). Mittal et al reported a case of ES of the clavicle with features of soft-tissue involvement that differ from our findings; they described the typical sunray appearance of the periosteal reaction at ultrasound and a pattern of infiltration at MRI that suggested a secondary localization of the perilesional structures (11).

The most frequent imaging appearance is a permeated, moth-eaten, poorly defined osteolysis that can also be massive. Periosteal reaction is common, especially in younger patients, and the most typical appearance is the onion-skin. Brush-like thickening of the cortex or a sunray appearance of the periosteal reaction can also be seen with radiography or sonography.

Although there are cases of Ewing sarcoma described in the literature of involvement of the trunk, localization in the clavicle or sternum appears to be quite rare. In the more commonly involved long bones, the tumor typically grows in the meta-diaphysis, often involving the epiphysis when the growth plate is closed (4). In our patient, no periosteal reaction was seen, and it was difficult to determine where the primary lesion originated. Our conclusion, considering the acute angles between the mass and the osteolytic area in the meta-diaphysis, was that the tumor arose from the soft tissues around the clavicle. Moreover, even if there were high T2 signal within the bone marrow indicating bone involvement, there was no visible disruption of the cortical bone.

The differential diagnosis raised by this case included other small-round-blue-cell tumors. It is crucial to consider the age of the patient and the anatomic location of the lesion as well as the imaging features. The differential diagnosis may include the following:

1. Other tumors from the Ewing sarcoma family such as primitive neuroectodermal tumor (PNET) and Askin tumor.
2. Hemangiopericytoma, which typically occurs in the lower extremities of older adults but has a rare infantile type (12).
3. Primary intramedullary osteosarcoma, which has a similar demographic incidence but usually has a more aggressive and destructive radiologic appearance (13).
4. Neuroblastoma, which usually occurs in younger patients (under 5 years of age) and generally presents with the manifestations of high catecholamine blood levels (14).
5. Lymphoma, which is more common in older age groups and may cause cortical bone destruction (15).
6. Osteomyelitis with soft-tissue involvement, which may resemble Ewing sarcoma on radiography but usually presents more acutely.
7. Langerhans cell histiocytosis, which may exhibit a less aggressive solid form of periosteal reaction rather than onion-skin layered periosteal reaction (16).

In conclusion, we present an uncommon case of extraskeletal Ewing sarcoma with nonspecific sonographic and radiographic features. The MRI helped to characterize the lesion by defining the soft-tissue components of the tumor and the involvement of the adjacent bones.

References

1. Ewing, J, The classic: diffuse endothelioma of bone. Proceedings of the New York Pathological Society. 1921;12:17. *Clin Orthop Relat Res*, 2006. 450: p. 25-7. [PubMed]
2. Enzinger, FM, *Primitive neuroectodermal tumours and related lesions, in soft tissue tumours*, S.W. Weiss, F. M. Enzinger, Editors. Maryland Hts, MO: Mosby; 1995:929-64.
3. Weiss, SW, Sobin LH, Enzinger FM, *Histological typing of soft tissue tumours.* 2nd ed. International histological classification of tumours. Berlin, New York: Springer-Verlag; 1994:49.
4. Campanacci, M, *Bone and soft tissue tumors: Clinical features, imaging, pathology and treatment.* Springer Verlag Wien ed. 1999:1175-1179.
5. Kransdorf, MJ, *Primitive neuroectodermal tumor (PNET)/ extraskeletal Ewing sarcoma, in imaging of soft tissue tumors,* M.J. Kransdorf, Editor. Lippincott Williams & Wilkins. 2006: 1076.
6. Grier, HE, The Ewing family of tumors—Ewing's sarcoma and primitive neuroectodermal tumors. *Pediatric Clinics of North America*, 1997. 44(4): p. 991. [PubMed]
7. Balamuth, NJ, Womer RB, Ewing's sarcoma. *The Lancet Oncology*, 2010. 11(2): p. 184-192. [PubMed]
8. Allam, K, Sze G, MR of primary extraosseous Ewing sarcoma. *AJNR Am J Neuroradiol*, 1994. 15(2): p. 305-7. [PubMed]
9. Okeeffe, F, Lorigan JG, Wallace S, Radiological features of extraskeletal Ewing sarcoma. *British Journal of Radiology*, 1990. 63(750): p. 456-460. [PubMed]
10. Rose, JS, et al., Extraskeletal Ewing sarcoma with computed tomography correlation. *Skeletal Radiol*, 1983. 9(4): p. 234-7. [PubMed]
11. Mittal, A, et al., Sunray appearance on sonography in Ewing sarcoma of the clavicle. *Journal of Ultrasound in Medicine*, 2010. 29(3): p. 493-495. [PubMed]
12. Enzinger, FM, Smith BH, Hemangiopericytoma. An analysis of 106 cases. *Hum Pathol*, 1976. 7(1): p. 61-82. [PubMed]

13. Shirkhoda, A, Peuchot M, Extraosseous Ewing's sarcoma. Computed tomography evaluation before and after chemotherapy. *Clin Imaging*, 1989. 13(2): p. 142-6. [PubMed]
14. Funahashi, Y, Yoshino Y, Hattori R, Ewing's sarcoma: primitive neuroectodermal tumor of the prostate. *Int J Urol*, 2009. 16(9): p. 769. [PubMed]
15. Barnardt, P, Roux F, The role of imaging in the evaluation of extraskeletal Ewing's sarcoma: case series. *SA Journal of Radiology*, 2013. 17(1): p. 30-33. DOI:10.7196
16. Stoller, JK, Diagnostic imaging: Orthopaedics. *Radiology*, 2005. 234(3): p. 886-886. DOI: http://dx.doi.org/10.1148/radiol.2342042574