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An unusual presentation of low-grade clavicle osteosarcoma: a case report and literature review

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DISCLOSURES

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Osteosarcoma (OS) is a rare disease with approximately 800-900 newly diagnosed cases each year in the United States. Of those, the majority occur about the knee. The distal femur is the most common site, followed by the proximal tibia, with the proximal humerus being a distant third. OS of the clavicle has been reported, with the earliest case report dating from 1975.¹ Since then, additional case reports of high-grade OS of the clavicle have been published.^{2,3} We describe the case of a 16-year-old female who presented with a mass on her right medial clavicle, which was confirmed to be a low-grade central OS.

CASE PRESENTATION

The patient is a 16-year-old female who presented to the Emergency Department (ED) for evaluation of a mass on her right clavicle, after being evaluated by her primary care physician (PCP). She noted an enlarging mass over the previous 2 months but stated that it had been asymptomatic until 4 days prior to presentation to her PCP, at which time she had developed tenderness to palpation and pain with range of motion of the right arm. X-rays were obtained at the PCP's office and she was referred to the ED for further evaluation. She denied constitutional symptoms.

At the ED visit, she was noted to have an area of erythema and tenderness over the medial aspect of the right clavicle with increased bony prominence. A chest x-ray demonstrated medial clavicle enlargement with periosteal reaction and sclerosis (FIGURE 1).

MRI demonstrated a 6-cm x 3.8-cm x 4.1-cm mass arising from the right medial clavicle with cortical destruction and concomitant displacement of the right subclavian and brachiocephalic veins (FIGURE 2). A CT-guided biopsy was performed 1 week later and demonstrated low-grade OS. The pathologist was concerned about the possibility of sampling error and the presence of a higher-grade component, as low-grade OS of the clavicle had not been reported.

The patient was evaluated by a pediatric hematologist/oncologist 2 weeks later after having obtained the biopsy and a PET/CT scan. At that time, the PET/CT showed an FDG-avid mass at the clavicle without evidence of pulmonary metastatic disease (FIGURE 3). She was subsequently evaluated by orthopedic oncology, at which time a discussion was had regarding further treatment. There was essentially no literature to guide the surgical and medical teams, as low-grade clavicular OS is unknown. Based on the evidence of localized, low-grade disease, the determination was made to proceed

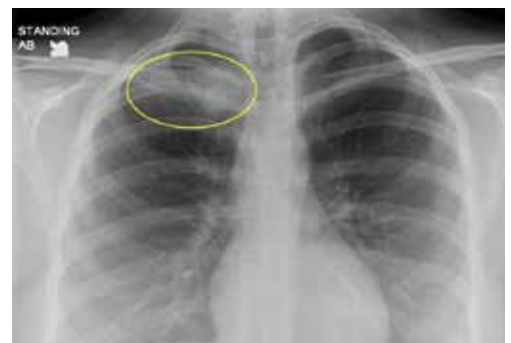


Figure 1. Chest x-ray obtained at the PCP visit.

with surgical resection. In the event that high-grade disease was identified at the time of final pathological evaluation, the pediatric hematology/oncology team felt that administering all of the patient's chemotherapy postoperatively would be acceptable and not affect her long-term prognosis. CT and CT angiogram were obtained for further operative planning (FIGURE 4).

Given the intimacy of the mass to the subclavian vessels, she was also seen preoperatively by pediatric general and cardiothoracic surgeons. The plan was formulated to have them in the operating room for mobilization of the subclavian vessels and in the event that a sternotomy was required for proximal control of the vessels. Following this visit, the case was discussed at the multidisciplinary pediatric tumor board and the consensus was to proceed with surgical resection.

Surgical Technique

General endotracheal anesthesia was administered without complication. The patient was positioned supine with a soft bump under her shoulders to place her neck in slight extension and thus facilitate access to the clavicle and great vessels. A 14-cm oblique incision was made over the subcutaneous clavicle extending to the contralateral sternoclavicular joint. Dissection was carried down to the fascia and the biopsy site was excised with the skin paddle. Dissection was carried through the sternocleidomastoid superiorly and the pectoralis major inferiorly, to 8 cm lateral from the right sternoclavicular joint. The clavicle was osteotomized well lateral of the palpable tumor and a marrow margin was sent for frozen section, which was found to be negative.

Dissection was continued circumferentially. Assistance from pediatric general and cardiothoracic surgery was required

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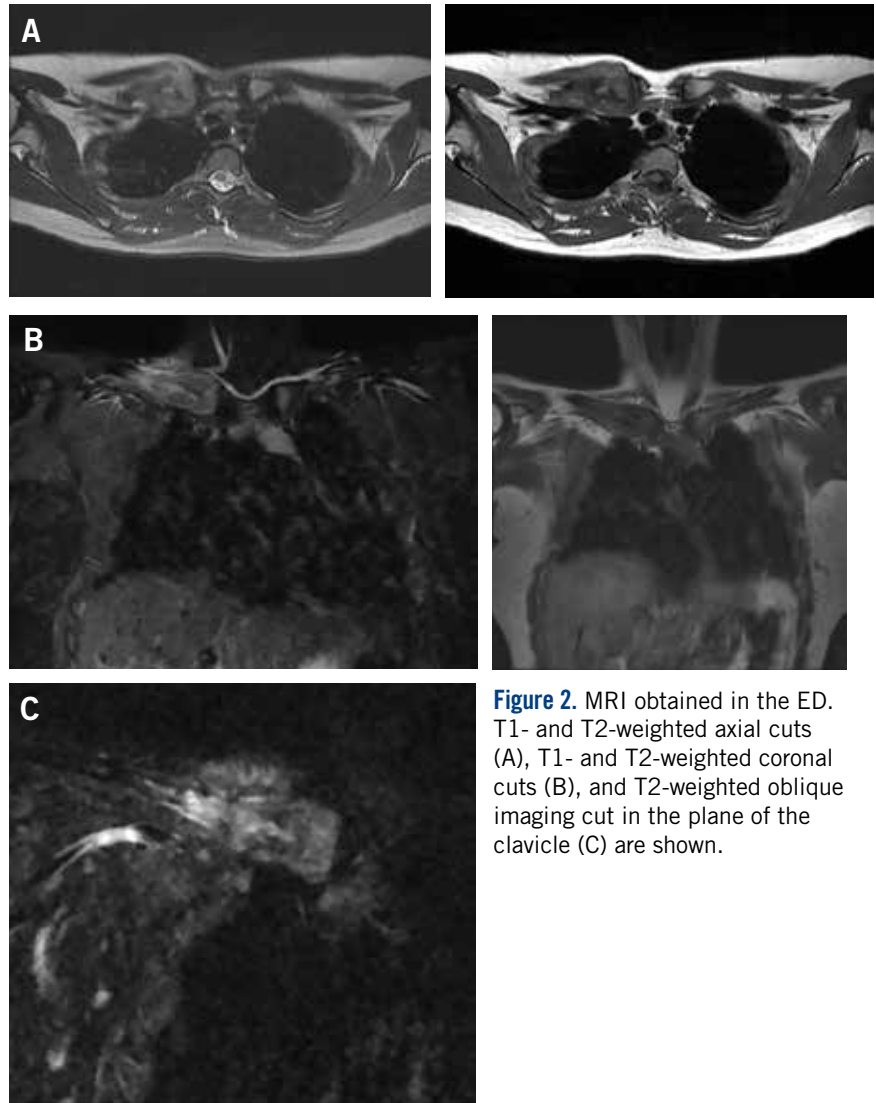


Figure 2. MRI obtained in the ED. T1- and T2-weighted axial cuts (A), T1- and T2-weighted coronal cuts (B), and T2-weighted oblique imaging cut in the plane of the clavicle (C) are shown.

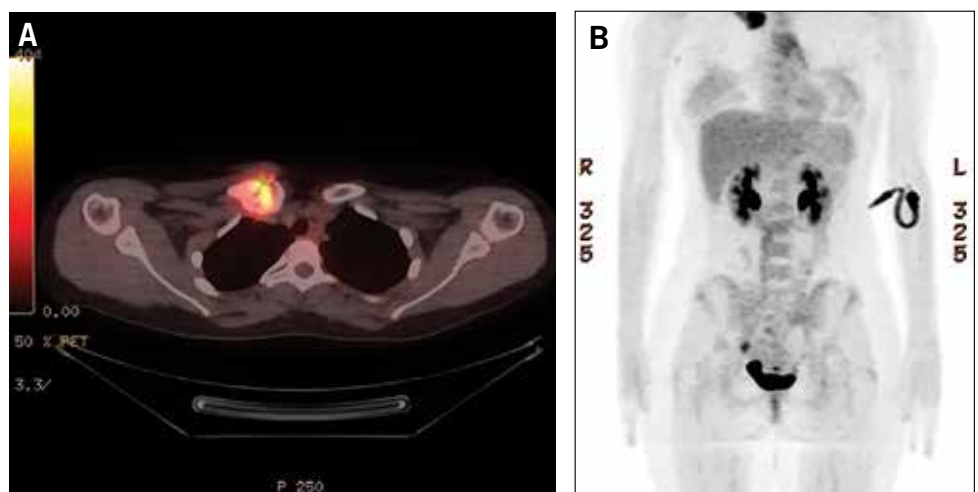


Figure 3. PET/CT obtained prior to medical oncology appointment. (A) FDG-avid mass at the medial aspect of the right clavicle. (B) Whole body topogram with no evidence of metastatic disease.

at the inferior aspect of the mass to assist with exposure and control of the subclavian vein (FIGURE 5A). A large branch of the subclavian vein near its junction with the internal jugular vein was found to be involved with the tumor and thus required suture ligation. The subclavian vein was noted to be intimate with the mass and somewhat friable. With the vein mobilized, a cuff of normal tissue was obtained inferiorly and superiorly to the mass. Medially, the sternoclavicular joint was disarticulated (FIGURE 5B). At this point, the specimen was delivered from the operative field and tagged in the usual fashion (FIGURE 5C). A medial soft tissue margin from the sternal side of the sternoclavicular joint was also sent and found

to be negative for tumor. The wound was closed in layered fashion over a ¼” Penrose drain. A soft dressing was placed, and the patient was successfully extubated and transferred to the post-anesthesia care unit in stable condition.

Postoperative Course

The patient was found to be neurologically and vascularly intact on postoperative exam and was discharged on postoperative day 1.

She was seen 14 days postoperatively and was doing well at that time, with full range of motion of the shoulder, elbow, wrist, and hand. Final pathology confirmed a low-grade OS with extraosseous extension. All margins were negative ex-

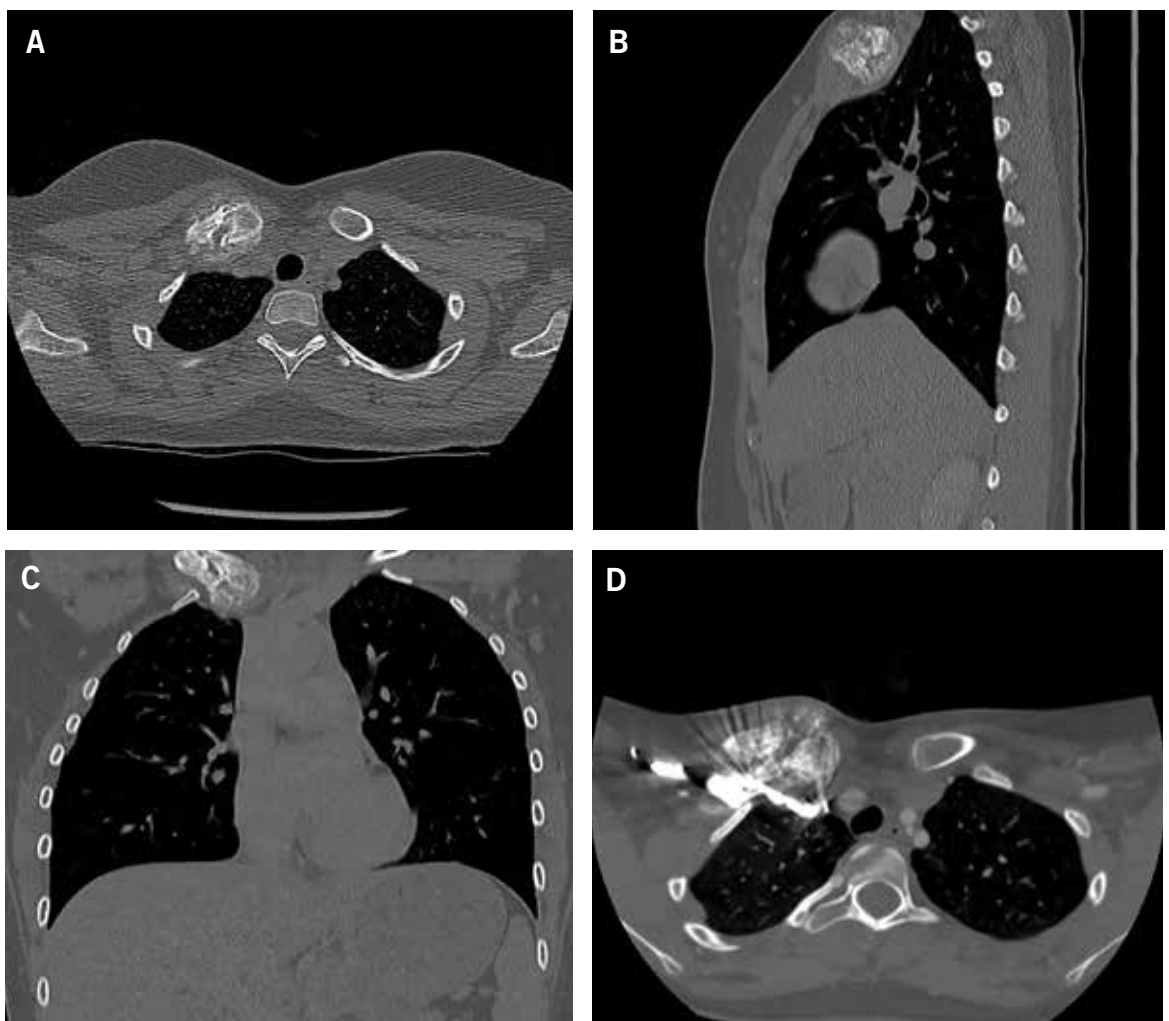


Figure 4. Chest CT and CT angiogram obtained prior to surgery. Representative axial (A), sagittal (B), and coronal (C) cuts are shown. Proximity of the mass to the subclavian vein is demonstrated on CT angiogram (D).

cept the medial (sternoclavicular joint) margin and the inferior margin adjacent to the subclavian vein. The intraoperative frozen section from the medial margin was negative for tumor.

The pediatric hematology/oncology team determined that, as no high-grade areas were identified, chemotherapy should be deferred. The positive margins were also discussed with the patient and her family specifically regarding further possible treatments. The findings from the pathology were discussed in a multidisciplinary tumor board and it was felt that, given the low-grade nature of the lesion as well as the high morbidity and risk of mortality with further surgery, additional surgery would be potentially more harmful than helpful. Additionally, low-grade OS is extremely resistant to radiotherapy. The plan remains to monitor her for local recurrence as well as metastases with serial imaging.

DISCUSSION

The clavicle is one of the first bones in the body to ossify but one of the last to have final physeal closure. Its unique characteristics have led to various descriptions, such as a “short tubular bone” versus a “flat bone.”^{4,5} Of note are its paucity of a true intramedullary space and scanty red marrow, which make it an unlikely site for a primarily intramedullary-based neoplasm to arise.⁴ However, it has also been noted that malignant lesions are more common in the clavicle than benign lesions, and special attention should be paid to aggressive-appearing lesions in the clavicle.

Radiographs can be misleading as well. Prior studies have demonstrated that low-grade central OS can be readily misdiagnosed as fibrous dysplasia, desmoplastic fibroma, nonossifying fibroma, osteoblastoma, and aneurysmal bone cyst.⁶ Findings found in low-grade OS can include evidence of cortical interruption, local soft tissue mass development, intramedullary involvement, cortical destruction, and poor margination; however, low-grade OS is typically sclerotic and highly trabeculated. Cross-sectional imaging can help differentiate between

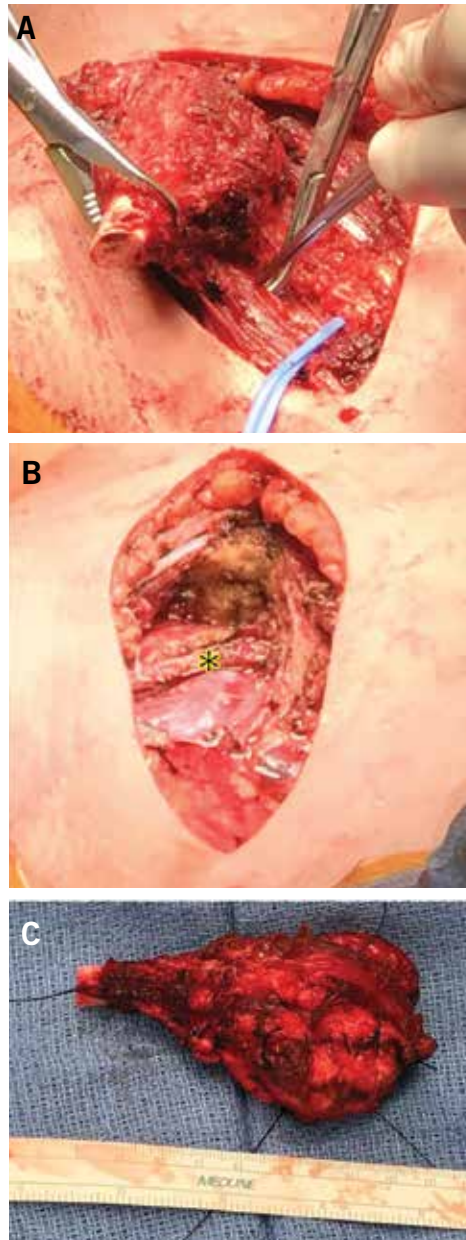


Figure 5. Intraoperative photographs. (A) The lateral osteotomy of the clavicle has been performed and the clavicle is being retracted away from the vessels. There is a vessel loop around the subclavian vein and a large branch leading into the tumor is being dissected free. (B) The tumor bed once the specimen has been resected, subclavian vein at the base of the wound bed (marked with an asterisk). (C) The specimen once resected. Final measurements were 9.2 cm x 4.5 cm x 3.1 cm.

OS and other more benign pathologies and should be considered in the clavicle where biopsy may be perilous.⁵

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> FINAL PATHOLOGY
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› THIS REPORT DEMONSTRATES THE NEED FOR MULTIDISCIPLINARY SARCOMA CARE, PARTICULARLY IN INSTANCES OF UNUSUAL DIAGNOSES.

The difficulty of clavicular biopsy has been reported. Not only does clavicular anatomy make biopsy hazardous, but also the potential for sampling error does exist. In a case report of one patient with a high-grade lesion, fine needle aspiration biopsy was initially diagnosed as an aneurysmal bone cyst but was ultimately found to be osteosarcoma.² Histology of low-grade lesions usually demonstrates minimal cytological atypia, rare mitotic activity, and variable osteoid production.⁵ Lower mitotic indices typically make wide resection curative for these patients, without the need for chemotherapy.

In this case, wide resection was carried out with the subclavian vein as the posterior-inferior margin and the sternoclavicular joint as the medial margin. Though the intra-operative medial margin was clear of disease, final pathology demonstrated focal (microscopic) involvement of the posterior and medial margins. A study of soft tissue sarcoma evaluated positive margins and concluded that the imperative of preservation of vital structures supersedes the need for negative margins.^{7,8} The rate of metastasis and overall survival was similar to surgical resections with positive margins. In the case of our patient, further resection would have carried significant morbidity and possibly mortality, including sacrifice of the major vessels to the arm below and entering into the sternum and thoracic cavity. The likely disability as well as the hazards of surgery were deemed to be too great to justify further excision. Frequent cross-sectional imaging will be necessary to evaluate the presence of

recurrent or metastatic disease. To our knowledge, this is the first documented case of low-grade clavicle OS. This report demonstrates the need for multidisciplinary sarcoma care at a center of excellence, particularly in instances of unusual diagnoses. **TSJ**

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