

Osteosarcoma of the Rib: A Case Report

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Abstract

Introduction: Osteosarcoma (OS) is the most common primary malignancy of bone in children and adolescents. OS has a predilection for the metaphyseal region of the long bones. The most common site of involvement is the distal femur followed by proximal tibia, proximal humerus, middle and proximal femur, and other bones. Primary OS of chest wall is a very rare entity.

Case Report: We, here, describe a case of a 14-year-old boy who presented to our center with a chest wall swelling and pleural effusion, which was subsequently diagnosed as chest wall OS originating from the rib. We treated him with neoadjuvant chemotherapy (Ifosfamide, Adriamycin, and Carboplatin). He showed an excellent transient response to the chemotherapy, after which he underwent an en bloc resection of the tumor. He was then started on adjuvant chemotherapy, but unfortunately, he relapsed soon after the last cycle and later on succumbed to the disease.

Conclusion: Chest wall OS is an infrequent malignancy. Pathological diagnosis is difficult and requires a high index of suspicion. Data regarding the prognostic factors are scarce, and no concrete guidelines are available for the management of such patients.

Keywords: Chest wall, osteosarcoma, rib.

Introduction

The differential diagnoses of a chest wall tumor in the pediatric population and young adults include primitive neuroectodermal tumors, Askin tumors, rhabdomyosarcoma, other sarcomas, and metastatic lesions in the ribs.

Osteosarcoma (OS) occurs principally in the long bones, while Ewing's sarcoma is frequently seen in flat bones such as the ribs and pelvic bones. OS occurring as a primary tumor in the rib is rare. Chest wall OS is an exceedingly rare entity and there are only case reports in the literature to guide the management. OS of the chest wall comprises 10% of all primary chest wall tumors and 3% of all OSs [1]. In the chest wall, both skeletal and extraskeletal OSs have been reported [2].

The usual skeletal site of origin is rib, scapula, and clavicle [1]. In a case series by Qian et al. [3] on thoracic extraskeletal OSs, the most common presentation was in fact, with a lung mass (40%).

The extreme rarity of this disease makes it very difficult to diagnose it. In most of the case reports, the treating physicians were not able to make a diagnosis upfront. The imaging characteristics are not specific. It poses a diagnostic dilemma for the pathologists as well. Biopsies from the lesion are difficult to interpret which is why most of the patients in the case reports discussed below had undergone upfront surgical resection.

Moreover, the case described here is an OS, which is giant cell-rich histology. Giant cell-

rich OS (GCRO), first described by Bathurst and Sanerkin [4], has been considered an extremely rare variant of OS, and its incidence was initially reported to make up 3% of all OS cases. It is most commonly seen around the knee [5]. It is difficult to differentiate from giant cell tumor of the bone. The presence of metaphyseal or diaphyseal centering, Codman's triangle, and radiographic intralesional fluffs differentiate GCRO from the latter.

Chest wall OS, apart from being a diagnostic challenge, is also a challenge for the oncologists. The correct approach for such tumors, whether to resect upfront or manage with neoadjuvant chemotherapy, the role of radiation therapy, all are questions yet to be answered. There is also scarce literature on

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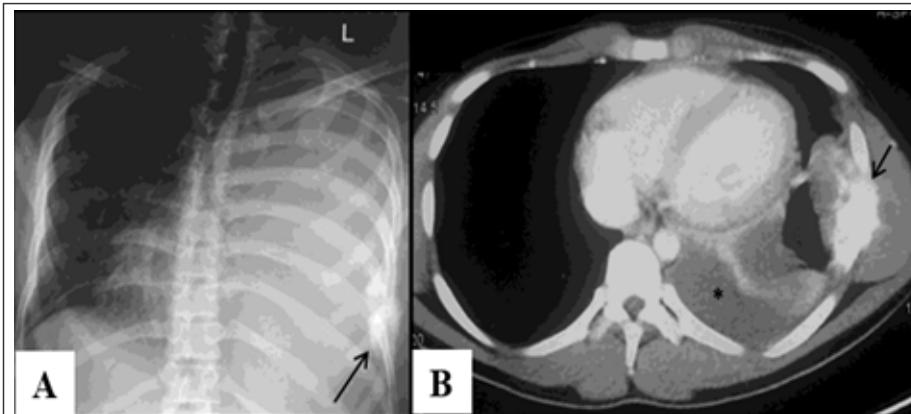


Figure 1: (a) Radiograph shows opacified left hemithorax and lytic destructive lesion of lateral aspect showing osteoid matrix mineralization (arrow). (b) Subsequently performed computed tomography scan confirmed destructive lesion of rib with osteoid matrix mineralization of associated soft tissue (arrow) and left-sided pleural effusion (asterisk).

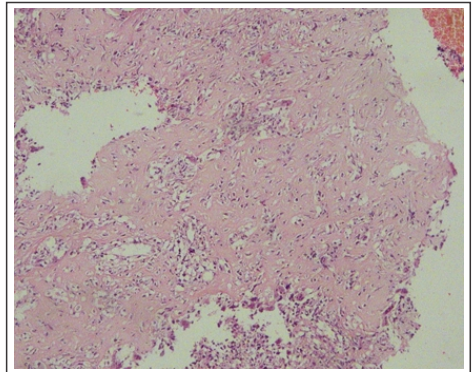


Figure 2: Low power images of osteosarcoma showing abundant deposition of malignant osteoid around the tumor cells. The osteoid is lacunar and eosinophilic in nature (H and E $\times 100$).

what are the common histologies of these OSs, how they respond to chemotherapy, and their prognostic factors.

Case Report

A 18-year-old male presented to our center with complaints of pain and a gradually increasing swelling over the left side of chest for 1 month. On examination, he had a 3 cm \times 4 cm tender, hard swelling over the left lateral aspect of the chest. His chest X-ray is described above (Fig. 1a). The swelling was fixed to the underlying bone. There was no regional lymphadenopathy.

His contrast-enhanced computed tomography (CT) chest (Fig. 1b) was suggestive of an expansile bony mass lesion with sclerotic changes containing flocculent calcification with adjacent soft-tissue component in the shaft of left 7th rib with minimum left pleural effusion.

A biopsy was taken from the lesion and sent for histopathological examination (HPE). Over the course of the next few days, his pleural effusion increased, and he developed shortness of breath, for which he required

intercostal tube drainage. Left-sided pleurocentesis was done; however, the cytological examination was normal. HPE (Fig. 2-4) revealed a tumor with morphological features suggestive of GCROs with osteoid formation making a diagnosis of OS.

He was started on Ifosfamide, Adriamycin, and Carboplatin neo-adjuvant chemotherapy regimen (Daw et al. – OS99) [6]. Clinically, his swelling and pain decreased markedly after the first cycle of chemotherapy. Post three cycles, the evaluation revealed $>30\%$ decrease in the soft-tissue component of the mass with complete resolution of left pleural effusion suggestive of partial response (Fig. 5a and b). Two additional cycles were given (total 5), following which he was taken up for surgery. He underwent en bloc (6th, 7th rib, and soft tissue) excision of the tumor with wedge excision of the left lower lobe nodule and reconstruction with steel wire, latissimus dorsi flap, and mesh. HPE of the resected specimen confirmed a high-grade conventional OS with mitosis rate of 3–4/high power field. The involved bony and

soft-tissue margins were free of tumor; however, the closest margin was 0.8 cm. The treatment effect was seen in the form of necrosis, sclerosis, and inflammation was 40% (grade II A: $<50\%$ response). After surgery (Fig. 6a and b), he was started on adjuvant therapy, which he completed.

The post-treatment scans, however, showed recurrence of the disease. The patient's performance status had worsened after the surgery. He and the family and attendants were counseled regarding the aggressive nature of the disease. He was managed with palliative care but later on succumbed to the illness.

Discussion

Only 1–2% of OSs occur as primary chest wall OSs [7]. There are only few case reports published and one case series which throw some light on this topic. These are difficult, hence, to diagnose and no consensus guidelines are available to guide the management of such patients. Much time is consumed for diagnosing such patients and initiating the treatment. In our case as well, many discussions with the pathologist were held. To complicate the matter, even more, ours was a giant cell-rich histology which further delayed the process. The patient eventually started worsening and developed a massive pleural effusion which needed intercostal drainage.

Burt et al. [8] published a case series in 1992, of primary bony and cartilaginous sarcomas of chest wall in their 40-year experience at MSKCC (Memorial Sloan-Kettering Cancer Center). Out of the total patients, 38 were OSs. The median age of patients was 42 years in this series. Male-to-female ratio was 1.5:1. Most of the patients presented with a painful chest mass. In this series, 30% of the patients

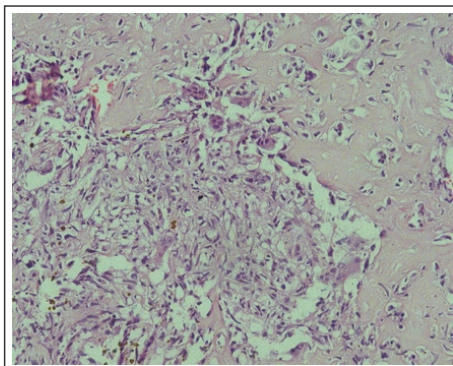


Figure 3: High power pictures showing malignant osteoblasts with moderately pleomorphic nuclei and moderate to abundant cytoplasm (H and E $\times 200$).

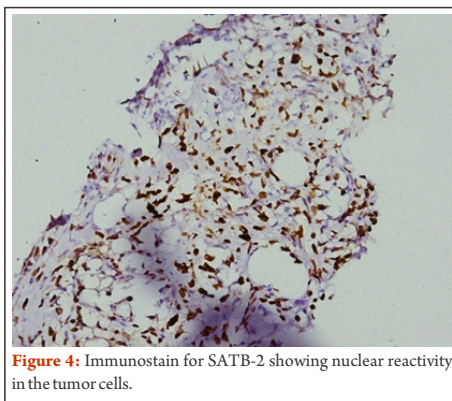


Figure 4: Immunostain for SATB-2 showing nuclear reactivity in the tumor cells.

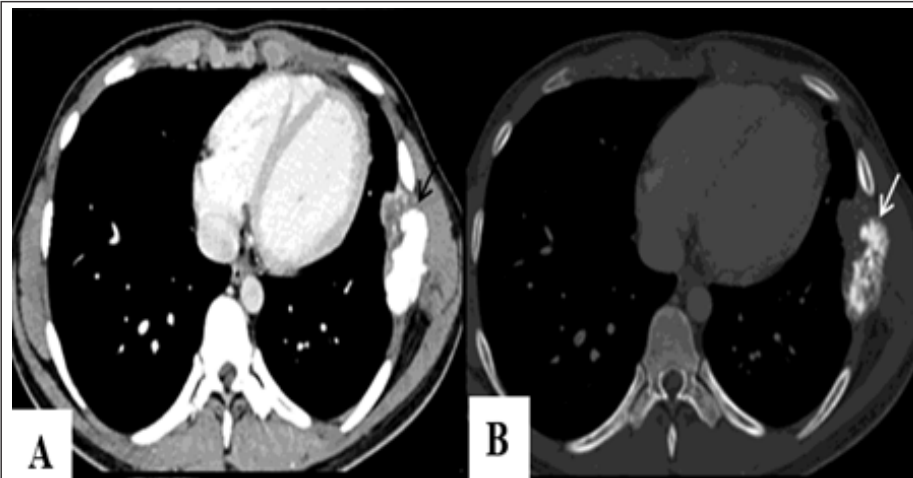


Figure 5: (a and b) Follow-up computed tomography after drainage of pleural effusion and five cycles of chemotherapy showed resolution of pleural effusion and reduction in size of soft-tissue component of the mass (arrow in A) and persistence of osseous lesion (arrow in B).

had received previous radiation to the chest wall, most commonly for Hodgkin's lymphoma. Of the 38 OSs of the chest wall, 13 (34%) arose in rib, 12 (32%) in scapula, 10 (26%) in sternum, and 3 (8%) in clavicle. Thirteen patients (34%) initially had synchronous metastases (10 to lung alone, 1 to lung and scalp, 1 to cervical lymph node, and 1 to diaphragm). Of the 25 patients who had localized disease upfront, 52%, later on, developed metastasis.

Our case was in concordance with the clinical presentation as reported in the literature, but in addition to that, our patient also had a rapidly progressive symptomatic pleural effusion, which had to be addressed even before starting with the definitive therapy. There has been no mention of any characteristic finding on imaging in the case of rib OSs, and the classic finding of sunburst appearance may not be seen [9, 10]. Our patient had flocculent calcification on

imaging which can be a clue for the diagnosis of OS.

Due to the rarity of this tumor, it poses a difficulty in the diagnosis and is usually not the first differential the treating oncologist or the pathologist keeps in mind when dealing with a case of a chest wall mass. In a case report by Qian et al. [3], a biopsy from an anterior mediastinal mass of a middle-aged lady was non-conclusive. The imaging was also difficult to interpret regarding the origin of the tumor. She eventually underwent a complete surgical resection of the tumor, histology, and immunohistochemistry of which was suggestive of OS. One of the reasons for undergoing with the surgery upfront may be a low index of suspicion for osteosarcoma and to resolve the diagnostic dilemma.

Initial diagnosis of this tumor in a small biopsy can be extremely challenging, and multiple biopsies and consultations with a

sarcoma specialist pathologist are required in order to establish a correct diagnosis before a definitive excisional surgery is performed. In the report by Rad et al. [11], a 57-year-old man presented with a sternal dense sclerotic swelling. A CT guided biopsy was inconclusive. The diagnosis of OS was subsequently made with an incisional biopsy from the lesion. In the case report by Sabatier et al. [2], the diagnosis was delayed by 4 years in a 30-year-old male with the left chest wall swelling. His MRI initially was suggestive of features of myositis ossificans. Percutaneous biopsy showed areas of osteogenesis with mature bony trabeculae in muscle. The intertrabecular space revealed benign-appearing fibroblastic proliferation. The retained diagnosis was myositis ossificans. The patient was subsequently lost to follow-up. Eventually, when the symptoms worsened and three biopsies later, he was diagnosed with extraskeletal low-grade OS with high amplification of both MDM2 and CDK4. The tumor, in his case, was not amenable to resection. He was started on chemotherapy, but his performance status worsened, and eventually, he succumbed to his illness. There was a delay in his diagnosis, leading to a fatal outcome.

In view of the rarity of this entity, the right approach towards the patient is not known and few patients underwent upfront surgical excision [1, 9, 10] and others resorted to neoadjuvant chemotherapy first [12, 13].

In the only case series by Burt et al. [8], primary treatment was resection in 31/38 patients (82%). Seven patients had resection of the primary tumor and the synchronous metastases to lung, diaphragm (1 patient), and cervical lymph node (1 patient). Adjuvant chemotherapy was used in 15/31 patients (48%) and adjuvant radiation therapy in 3/31 patients (10%). Of the 31 patients undergoing resection, 10% had local recurrence. The overall 5-year survival was 15% in this study; the overall median survival was 12 months. Survival was maximum in patients who underwent resection plus adjuvant chemotherapy.

In our case, the histology was giant cell-rich type. GCRO is defined as an OS, in which more than 50% of the tumor consist of numerous uniformly distributed osteoclastic giant cells amidst oval or spindle mononuclear cells embedded in a fibrovascular stroma [5]. It forms 3% of all

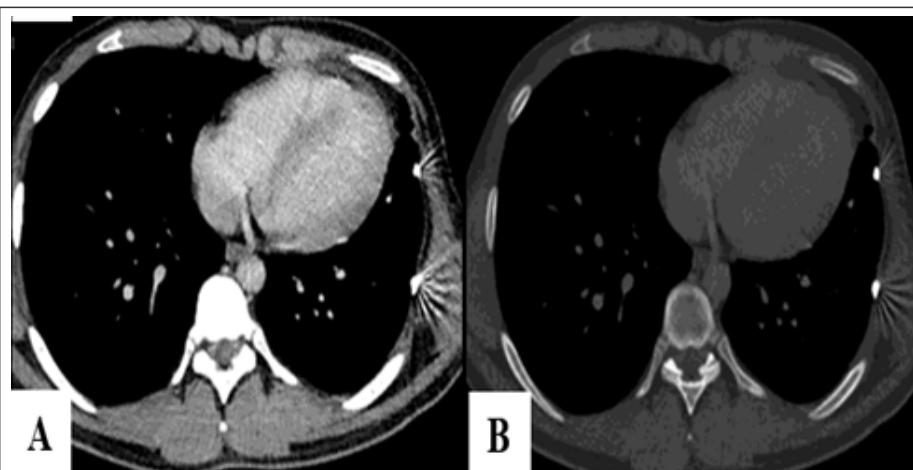


Figure 6: (a and b) Axial computed tomography after 2 months of surgery showed surgical clips in situ with no evidence of recurrent or residual mass along the surgical site.

OS and has a clinical course similar to high-grade OS. Again, it poses a diagnostic dilemma and requires expertise.

Our patient was not fit for upfront surgery, so he was started on neoadjuvant chemotherapy. He responded to the chemotherapy with a resolution of pleural effusion and reduction in the size of the lesion and later on underwent surgery with reconstruction of the chest wall. However, such OSs have a poor prognosis [8].

Moreover, the HPE of this patient showed only 40% necrosis which is again a poor prognostic factor [14]. He subsequently had recurrent disease.

Conclusion

Our case reinforces that OS of chest wall requires a high index of suspicion. Flocculent calcification on imaging can be a clue to the diagnosis. A thorough histopathological review by an expert sarcoma pathologist is

needed. OS is a chemoresponsive disease and symptomatic patients not amenable for upfront resection can be stabilized with the use of neoadjuvant chemotherapy before proceeding to the surgery. However, the data on the disease is very limited to guide the management in these rare cases.

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