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CASE REPORT

Primary Osteosarcoma of Rib in a Child—an Uncommon Case Report

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Introduction

The WHO defines osteosarcoma as a malignant neoplasm in which the neoplastic cells produce bone [1]. Conventional osteosarcoma represents 75 to 85% of all osteosarcomas. It is a high-grade malignant tumour that occurs predominantly in the metaphysis of the long bones of adolescents and young adults. Osteosarcoma can be classified into osteoblastic, chondroblastic and fibroblastic variants depending on predominant component. Chondroblastic osteosarcoma represents 10–15% of all osteosarcoma [2].

Case Report

A 12-year-old boy presented at general surgery outpatient department with a large swelling over his anterior chest wall for the last 1 year which was gradually increasing in size. On local examination, the mass was firm, non-tender and fixed to chest wall on right side. Computed tomography of the chest revealed an expansile lytic lesion measuring 7 × 3.5 × 2.5 cm, over anterolateral aspect of the sixth rib (Fig. 1).

Complete surgical excision was done, and the specimen was received at the histopathology department. Gross examination revealed a greyish-black mass measuring (7 × 3 × 2 cm), and cut surface was friable in appearance. Microscopically, the mass showed predominantly cartilaginous differentiation. Cartilage cells were marked anaplastic towards the periphery of lobules. Hypercellular splindling of spindle cells is present. Osteoid matrix was seen lined by malignant tumour cells (Figs. 2, 3 and 4). The diagnosis was supported by immunohistochemistry which was negative for BCL2 and S100 (Figs. 5 and 6).

Discussion

Osteosarcomas are malignant mesenchymal neoplasm that rarely occur in the thorax. The rib, scapula and clavicle are the most frequent sites of origin [3]. Osteosarcoma affects the ribs in 1 to 3% of cases [4]. Cases originating from the ribs are infrequent and have been reported mainly in the paediatric population [5, 6]. The youngest patient with osteosarcoma of the rib reported in the literature to this date is a 7-year-old girl [7]. In our case, the patient was 12 years old. The commonest presentation of osteosarcoma of the rib is pain and a palpable chest wall mass [8]. In the present case, the child presented with chest wall swelling.

Osteosarcoma originating from such rare sites poses numerous diagnostic and therapeutic challenges [9]. The diagnostic 'sunburst-'type radiological feature was not observed in the present case. Computed tomography (CT) examination is of utmost importance for diagnosing and differentiating rib tumour from pleural and lung lesion. CT also assesses the depth of extension of tumour to underlying structures.

According to the WHO classification, the chondroblastic osteosarcoma is defined as a histological entity characterized by predominant presence of chondroid matrix, which tends to exhibit a high degree of hyaline cartilage and is intimately associated with the non-chondroid element (osteoid or bone matrix) [10]. Extensive chondroid differentiation may often be confused with chondrosarcoma. Unni suggests that in an adolescent patient, chondroid tumours should be considered chondroblastic osteosarcoma, unless proven otherwise [11]. Other differential diagnoses include Ewings sarcoma, rhabdomyosarcoma, primitive neuroectodermal tumours (PNET) or Askin tumours, other sarcomas and metastatic lesions in the ribs [8].

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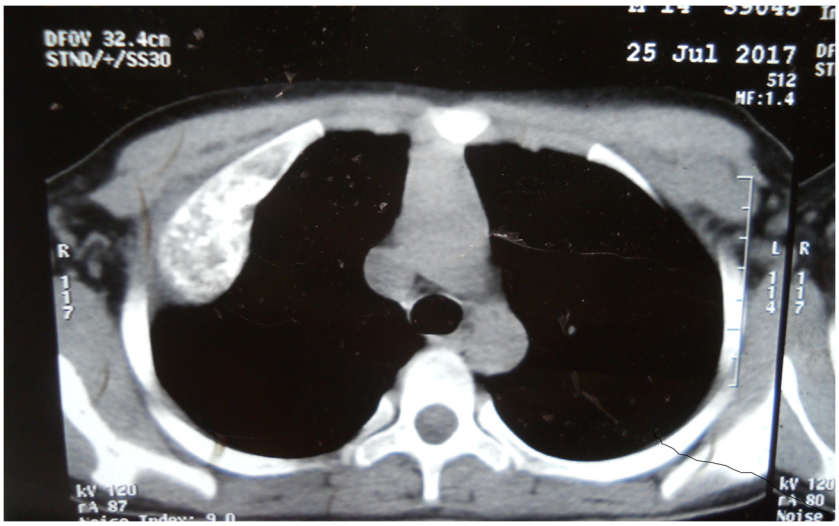

Fig. 1 CT scan showing lytic lesion of the rib

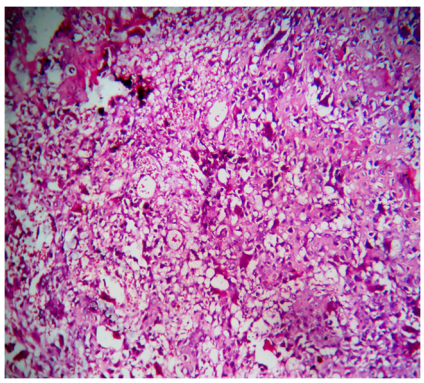

Fig. 2 Picture showing osteoid H&E (X100)

The prognostic significance of chondroblastic differentiation has been controversial, with conflicting reports of association with better [12] as well as worse survival [13].

For osteosarcoma of the rib, a wide surgical excision followed by adjuvant chemotherapy increases the chance of a relapse-free survival of the patient [7]. Surgery should include resection of the full thickness of the chest wall with wide margins that may include the adjacent ribs, intercostals muscles, pleura and vertebrae [8].

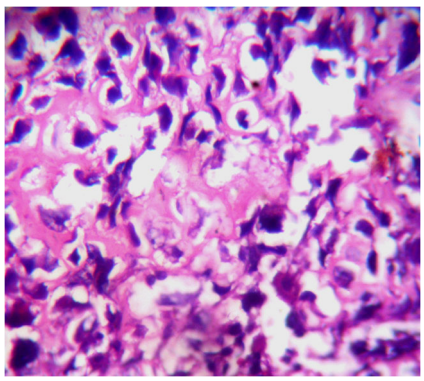

Fig. 3 Picture showing osteoid lined by malignant cells (X400)

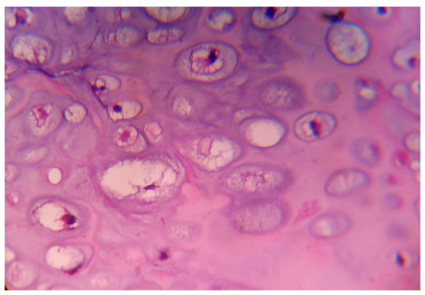

Fig. 4 Picture showing cartilaginous differentiation (X400)

Conclusion

We presented a rare case of primary chondroblastic osteosarcoma in the rib of a child. In paediatric and adolescent age groups, chondroblastic osteosarcoma is an important differential diagnosis of chest wall swelling. Though diagnostic difficulties are common, combination of radiological, histopathological and immunohistochemical features aids in diagnosis. Early diagnosis and prompt treatment are the cornerstone of management in chondroblastic osteosarcoma.

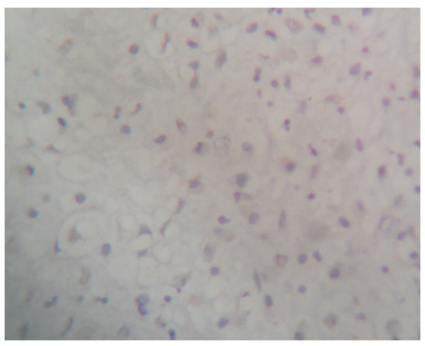

Fig. 5 Picture showing S100 negative stain (400)

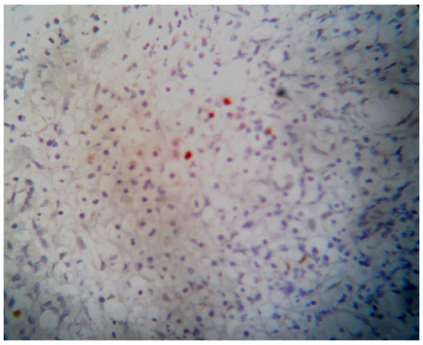

Fig. 6 Picture showing bcl2 negative stain (400)

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict interest.

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