Case Report

Primary rib osteosarcoma: A rare entity

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ABSTRACT

Osteosarcoma (OS) is the most common primary bone tumor. Predominantly it is seen in the long bones of the body and rarely in the flat bones. Primary bone tumor arising from rib is most commonly chondrosarcoma, and rare reports of OS are there in the literature. Management of OS is multi-modal, including surgery and chemotherapy. However, radiotherapy plays a role in OS of flat bones when a complete resection is not possible due to cosmesis or surrounding vital structures. Here, we report a case of primary OS from rib, which was managed by surgery and chemotherapy.

Key words: Chemotherapy, osteosarcoma, rib

INTRODUCTION

Osteosarcoma (OS) is the most common primary bone tumor. It occurs during the period of rapid growth as seen in puberty, but can also occur in children and in elderly individuals. OS mainly arises from the long bones such as tibia, femur, and humerus. Rarely does it arise from flat bones such as clavicles, ileum, sacrum, ribs.[1] Flat bone OSs are more common in the adult population but though uncommon, it can occur in the pediatric population. Primary OS in ribs is a rare entity. Here, we report a case of primary OS arising from the rib in an adult.

CASE REPORT

A 19-year-old male patient presented to the out-patient department with a history of swelling in the left side of the chest. The swelling was painful, and it was insidious in onset and progressive in nature. There were no other systemic complaints. For the above complaints, patient was evaluated, and chest radiograph revealed a homogenous opacity in the left lung field as shown in Figure 1. Computed tomography (CT) scan of the thorax was done, and it showed a mass arising from the left 2nd and 3rd rib [Figure 2]. Possibility of chondrosarcoma was considered. Staging work up was normal, and the patient was taken up for wide excision of the involved ribs along with chest wall reconstruction. Histopathological examination (HPE) of the specimen revealed conventional OS of the left 2nd and 3rd ribs with areas of osteoid along with negative margins [Figure 3]. Since there was characteristic presence of osteoid, immunohistochemistry studies were not contemplated. Patient was started on adjuvant chemotherapy consisting of ifosfamide, adriamycin and cisplatin for a total of six cycles along with myeloid growth factor support. At present, he is doing well and on regular follow-up.

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DISCUSSION

Osteosarcoma is the most common malignant neoplasms of the bone with a peak incidence in the second decade with the most common site being the metaphysis of the long bones as the tumor has a special predilection for the growing end of the long bones. Around 10% of OS arises from the flat bones, pelvis being the most common site. Chest wall involvement by OS has been reported up to 3% of cases.[2] Tumors of flat bones can occur as a result of metastasis or as secondary to radiation or chemotherapy based treatment, but primary OS is extremely rare.[3]

Osteosarcoma arising from such rare sites poses a diagnostic dilemma to the primary treating doctor including the radiologist and the pathologist. The most common symptom due to the chest wall OS is a pain and a

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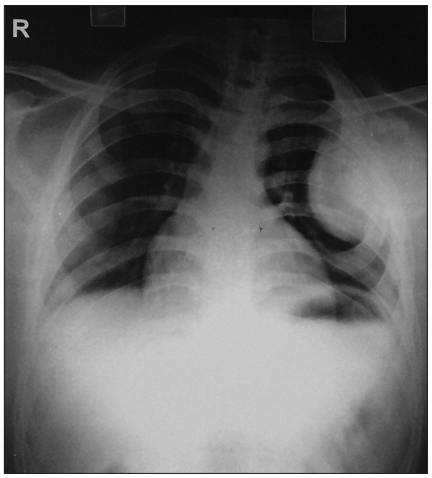

Figure 1: Chest X-ray showing a homogenous opacity in left lung field suggestive of a nonparenchymal lesion

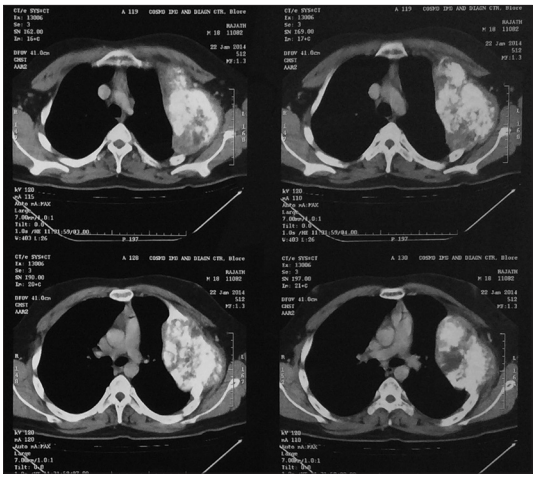

Figure 2: Computed tomography scan shows a heterogenous calcified lesion in left 2nd and 3rd rib with areas of necrosis

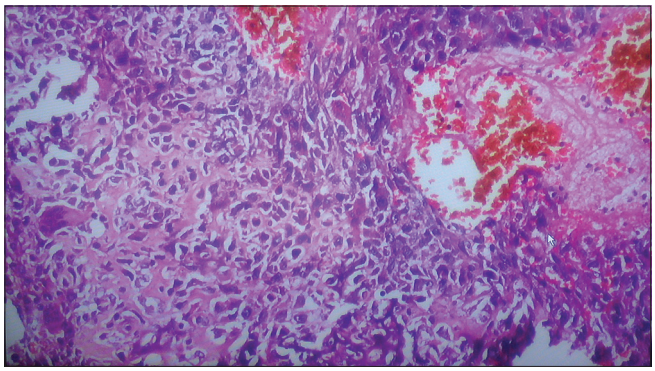

Figure 3: Histopathological examination shows pleomorphic tumor cells with areas of eosinophilic homogenous osteoid suggestive of osteosarcoma

chest wall swelling. The radiologic finding in these patients will be homogenous opacity in the involved region. The characteristic radiological findings of OS like periosteal reaction, destruction of the rib and calcifications may be difficult to visualize because of the limitations of chest radiograph compared to those of the long bones.[4] The lesion may be misinterpreted as an extra osseous chest wall tumor, teratoma, calcified mesothelioma or lung cancer. CT scan and magnetic resonance imaging helps in identification and characterization of the mass. Often the epicenter of the mass is localized to the rib. It also provides additional diagnostic information about the involvement of adjacent structures like lung, pleura and also for spinal cord involvement in OS arising from the ribs.

Because of the site of the tumor being flat bones and radiologic changes, it can be confused with more common bone tumors arising from these sites such as osteochondroma, chondrosarcoma, and fibrosarcoma.[5] Hence, an accurate HPE is essential for making a proper diagnosis and appropriate management. The tumor cells are spindle-shaped and are pleomorphic in nature. The clinching diagnostic clue is to detect the presence of osteoid, which is histologically, a dense, pink, amorphous intercellular material.[6] Diagnostic hurdles may be encountered when the osteoid production is scant, and a careful search for the same and extensive sampling of the tumor is warranted. A combined effort with radiological and clinical correlation is trustworthy and may serve to avoid pitfalls in the diagnosis.

Management of OS is multi-modal. Staging investigation is essential to rule out any metastatic disease. CT scan thorax to detect pulmonary metastases and a bone scan to rule out other bone involvement is a must. The primary mode is the wide local excision. In the rib, surgical management includes complete excision of the involved rib with repairing of the defect with an appropriate mesh. Though OS is relatively radioresistant, radiotherapy has been tried with some benefits in cases of OS of head and neck region, axial skeletons when complete resection is not fully feasible. If margins are involved, addition of radiotherapy has been tried, which has some beneficial effect. Adjuvant chemotherapy is an essential component of the treatment as it has increased overall survival of patients with OS.[7] The chemotherapeutic agents with maximum activity against OS are high dose methotrexate, cisplatinum, doxorubicin, and ifosfamide. Combination of chemotherapeutic agents was administered to this patient, and presently he is doing well and on regular follow-up.

Osteosarcoma though is a common tumor arising from the long bones, can arise from flat bones like in ribs as seen in this case. The main modality of treatment is surgical excision of the tumor, followed by adjuvant systemic chemotherapy. This can lead to improvement in cure rates. In cases of incomplete excision, additional radiotherapy can be tried to improve the outcome of patients.

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