Case Report

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Chondromyxoid Fibroma: An Unusual Tumour at An Atypical Location

Oncology Section

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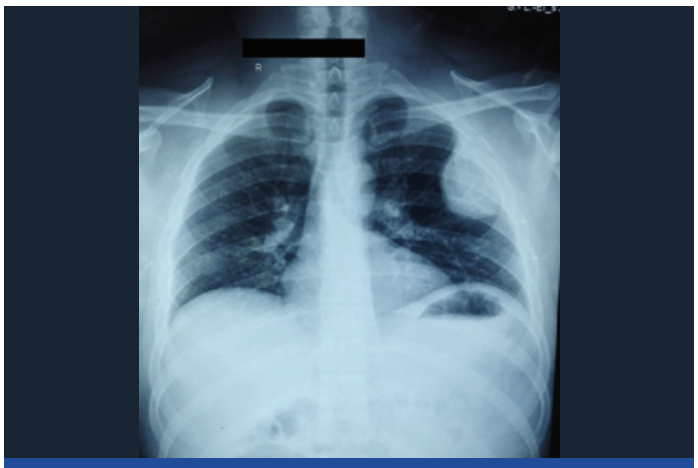
ABSTRACT

Rib tumours are mostly secondaries arising from breast or prostrate malignancies. Among primary rib tumours, osteochondromas are reported as the commonest cause. Chondromyxoid fibromas are primary benign rib tumours that are seldom seen, occurring almost exclusively at the metaphyseal ends of large tubular bones. Here a case of chondromyxoid fibroma of rib, its clinical and radiological features, management and prognosis, is discussed which has only an occasional mention in literature.

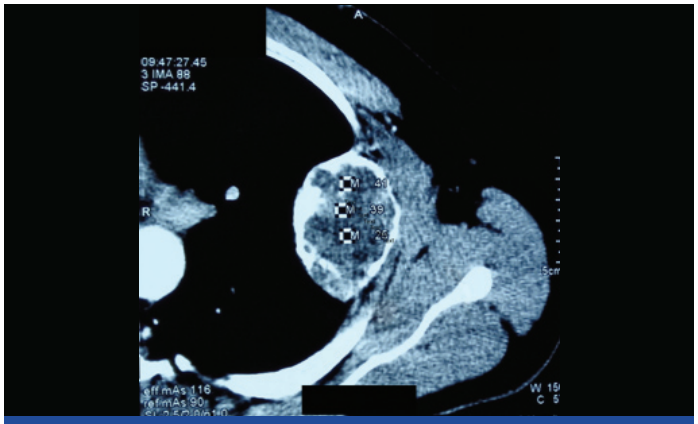
Keywords: Benign rib tumours, Primary rib tumours, Secondary tumours

CASE REPORT

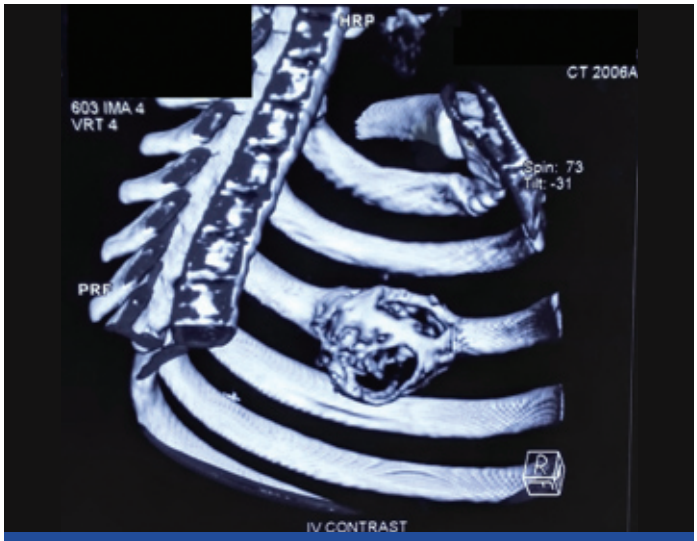
A 22-year-old male came to the surgical outpatient department to obtain fitness for a job interview. He was clinically normal. On obtaining a postero-anterior chest radiograph, a dense shadow was noted in his left 3rd rib [Table/Fig-1]. After reviewing the chest radiograph, a contrast enhanced computed tomography (CECT) of the thorax was ordered to delineate the lesion [Table/Fig-2,3]. With a preoperative diagnosis of a primary rib tumour probably of benign origin, the patient was advised an excision biopsy of the lesion. He underwent a left sided posterolateral thoracotomy followed by wide local excision of the left 3rd rib. Intraoperatively, a grayish white smooth nodule measuring approximately 7.0x5.5x4.0cm lesion on the inner aspect of the left 3rd rib projecting anteriorly was identified and excised with a wide margin [Table/Fig-4]. The wall of the tumour was thin with an eggshell like appearance. On microscopic examination, the tumour showed lobules of chondroid areas and spindles with osteoblastic multi-nucleated giant cells. There were focal areas of calcification with no evidence of malignancy that proved the diagnosis of Chondromyxoid fibroma [Table/Fig-5]. Patient was discharged on the fifth postoperative day and has been on regular follow-up with no signs of recurrence.



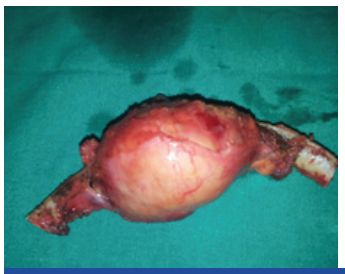
[Table/Fig-1]: Chest radiograph (PA view) showing the tumour

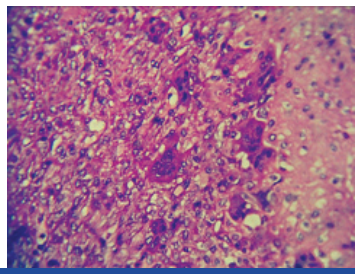


[Table/Fig-2]: CECT chest (trans-axial view) showing the tumour on the inner aspect of the rib



[Table/Fig-3]: CECT reconstruction image showing the tumour





[Table/Fig-4]: Specimen post resection **[Table/Fig-5]:** HPE of the tumour (x10) showing giant cells with normal chondroid tissue

DISCUSSION

Chondromyxoid fibroma (CMF) is the least common bone tumour with an incidence of less than 1% and only around 500 cases having been reported till 2000 [1]. CMF was coined and reported first by Jaffe and Lichtenstein in 1948 [2]. It has been suggested that chromosome 6 abnormalities may be involved in the formation of this lesion [3,4]. Patients with this entity present usually with pain, swelling or cough [5,6] and sometimes can be completely asymptomatic or may present with a pathological fracture [3]. There is no racial or sexual predilection although males might have a slightly higher incidence. The youngest and the oldest reported patient were of ages 3 and 87 respectively at the time of diagnoses [1].

Serial number	Location	Age of patient	Sex	Symptoms	Treatment
1	Left 1st rib	60 years	Male	Persistent cough	En bloc resection of the first and second ribs, with distal clavicle and left upper part of the sternum.
2	Right 2nd rib	15 years	Female	Mild chronic back pain	En bloc resection of the tumour
3	Left 4th rib	25 years	Male	Left upper back pain	Excision with a 2cm margin
4	Left 5th rib	40 years	Female	Incidental finding	Excision with a 2cm margin
5	Right 2nd rib	15 years	Female	Mild back pain	Excision with a 2 cm margin

[Table/Fig-6]: Site of occurrence age, sex, symptoms and management of rib CMF [6-10]

The tumour is known to arise from the metaphyseal end of long bones like tibia, predominantly from the upper end and the femur and very rarely from fibula, calcaneum, metatarsals, phalynges, scapula, ribs and vertebrae [7]. It rarely arises from the rib with only five cases reported as of 2013 [Table/Fig-6].

CMF is believed to have a cartilaginous origin. The tumour does not normally breach the periosteum, but however is located eccentrically with a thin shell of periosteal bone. The cut surface may appear greyish-white or bluish grey resembling cartilage having small cystic cavities. Microscopically, the tumour has lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle-shaped or round cells with a varying number of multinucleate giant cells of different sizes [4,7]. Immunohistochemically, the tumour resembles a chondroblastoma [4].

Radiologically, the tumour is not easy to diagnose. Radiographs might show an expensile ovoid lesion with a radiolucent pattern and well defined sclerotic margins, septations and bulging thinned out overlying cortex [6]. CT scanning is the best imaging modality for detecting sclerotic margins and ridges and matrix mineralization, and CT findings can depict the cortical integrity of the lesion. CT scans may show calcification within the tumour that is not visible on conventional radiographs; therefore, CT findings may increase the suspicion that a lesion is cartilaginous [2,3,6]. On MRI, depending on the varying amounts of myxoid and cartilage tissue, the center of the tumour is hyperintense on T2-weighted spin-echo images and STIR sequences. Along with the typical lobulated pattern, the images are distinctive of a tumour of cartilaginous origin. The highly vascularized connective tissue at the border of the lesion accounts for a rim of moderate to high signal enhancement on T1-weighted images after gadolinium-diethylene triamine penta-acetic acid (Gd-DTPA) [5]. Nuclear imaging has been used but literature is limited [2]. Either CT or MRI is preferred for diagnosis and the differentials: fibrous dysplasia, aneurysmal bone cyst, chondroblastoma, chondrosarcoma, periosteal chondroma, intraosseous schwannoma and periosteal haemangioma should be borne in mind [3,5,7].

En bloc resection with negative surgical margins is preferred over curettage in treating this condition. Recurrence as a potential complication must be remembered. Amputation must be considered in cases with local recurrence and with soft-tissue involvement in critical areas. Radiation is not recommended in both, cases with resectable tumours and those with recurrences due to the fear of radiation induced malignancy [5]

CONCLUSION

Chondromyxoid fibroma of rib is an extremely rare tumour. Awareness of this diagnostically challenging condition is required as accidental fatal misdiagnosis of chondrosarcoma may be made. Treatment should involve en bloc resection with negative surgical margins, as the tumour is notorious for recurrence.

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