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Case Study

Chondrosarcoma of chest wall invading lung: A rare case report

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ABSTRACT

Chondrosarcoma is the most common primary malignancy of the chest wall but occasionally primarily arise within the lungs and bronchi. Sometimes tumors arising from the ribs and sternum manifest as large intrathoracic masses with only minimal osseous involvement. In this case report a 60 year old male, presented with swelling in the right mammary area. Radiologically there is an irregular expansile lobulated soft tissue mass lesion arising from the right third anterior rib. Histopathology was suggestive of chondrosarcoma.

Keywords: chondrosarcoma, intrathoracic mass, resection, chemoradiation.

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INTRODUCTION

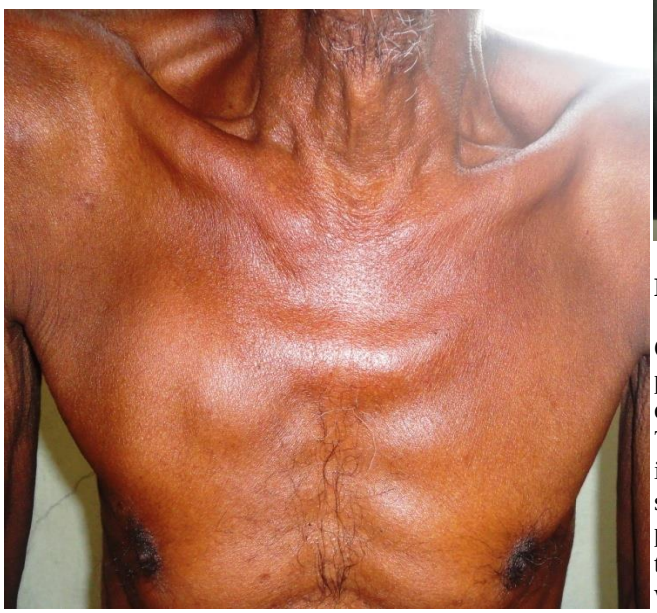
Chondrosarcoma is the single most common malignant tumor of the chest wall, most often, it occurs anteriorly at the costochondral junction[1]. They may also arise from sternum and scapula. The commonest reported incidence of these tumors is in 3rd and 4th decade and is more frequent in men with male to female ratio of 1.3:1[6]. This is a locally invasive slowly growing tumor and usually presents with pain in a previously asymptomatic mass. It is known to recur locally and may have late metastasis.[2]

CASE REPORT

A 60 year old male, presented with the swelling in the right mammary area and dry cough from 6 months. The swelling was insidious in onset, associated with pleuritic chest pain, right shoulder pain and shortness of breath. His past medical history was not significant .He is a chronic smoker, agriculture worker by occupation.

On examination there was bulging noted in right mammary area (Fig 1).There was dull note on percussion and diminished breath sounds were noted. Chest X-ray PA view showed right lung mass with multiple bilateral metastatic lesions (Fig-2).Ultrasonography of the chest showed large right sided intrathoracic mass lesion with calcifications.

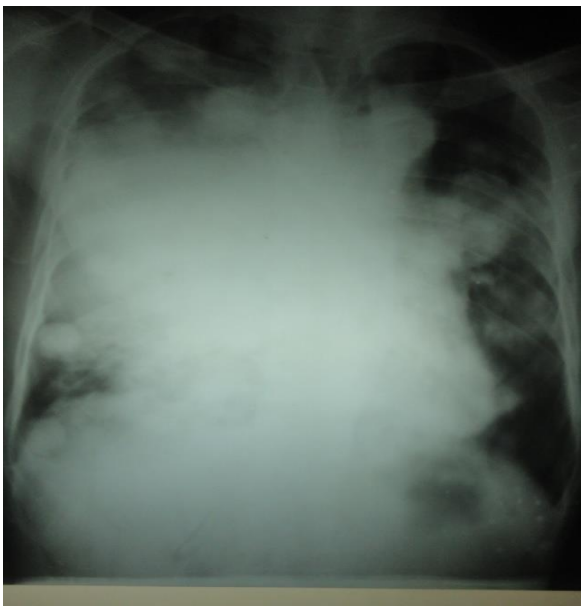
Figure:1 showing bulging in the right mammary area



A computed tomography (CT) scan of the chest revealed irregular expansile lobulated soft tissue mass lesion measuring 13×14.9×18 cm arising from the right third anterior rib and showing lytic lesions in right third rib (Figure 3).The lesion was involving and compressing adjacent lung parenchyma, extending in to anterior mediastinum and compressing SVC, right atrium, ascending aorta. Multiple metastatic lesions were seen on both lungs, largest measuring 5× 4.9 cm.

An ultrasound guided fine needle aspiration of the lesion prepared for rapid cytological evaluation showed chondrocytes with moderate vacuolated cytoplasm with hyperchromatic nuclei and some binucleated cells in chondromyxoid background, suggestive of chondrosarcoma(Fig:4).

Fig:2 Chest X-ray PA view showing mass with metastatic lesions



DISCUSSION

Chondrosarcomas are malignant tumors that produce chondroid matrix. Most are >4 cm in diameter, and some grow as large as 25 or 35 cm.[3,4] The most extensive tumors occur in the flat and irregular bones, including the pelvis, ribs, and scapula, where they can grow to a large size before producing symptoms. Chondrosarcomas of the thorax usually manifest as large chest wall masses with bone destruction and soft tissue extension[6]. Within the chest, anterior ribs are most commonly involved, at the costochondral junction.

Fig:3 CT scan chest showing metastasis

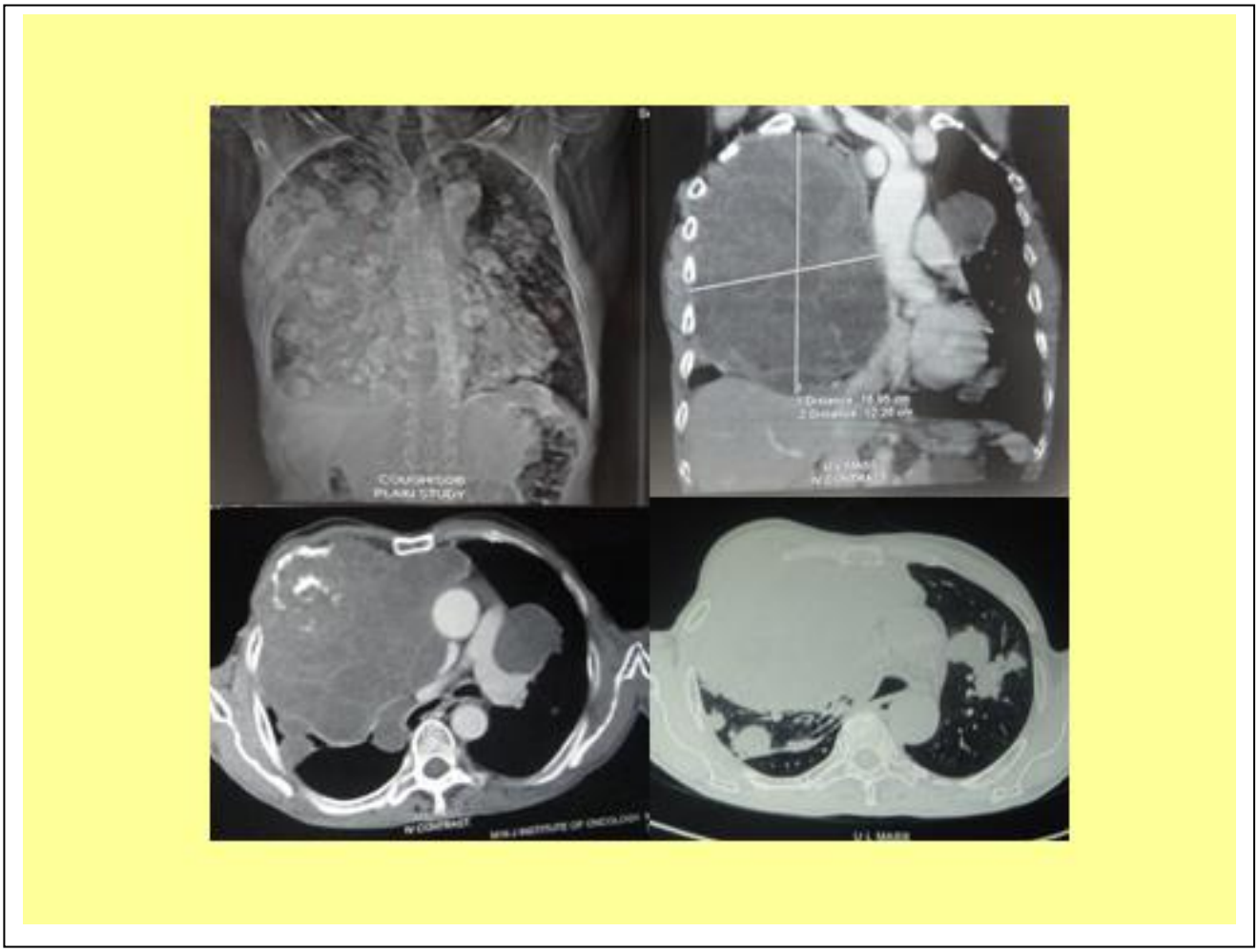
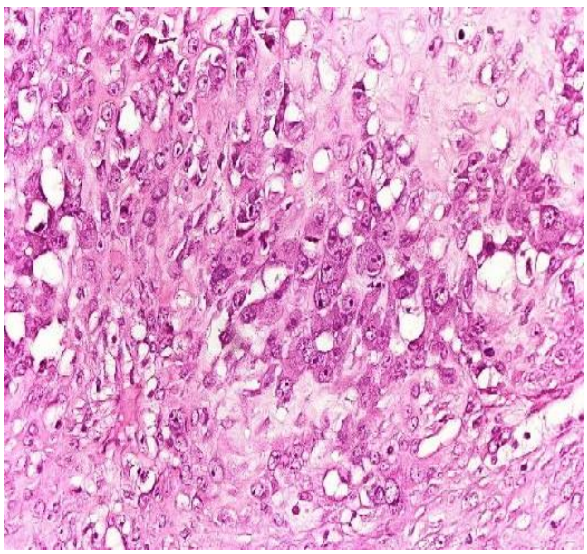


Fig:4 cytology showing chondrocytes , hyperchromatic nucleoli, binucleated cells



Pain is the most frequent presenting symptom, reported in 95% of patients[3,7]. It is often insidious and progressive, with an average duration of 1 to 2 years prior to presentation. The pain is typically described as aching at rest and worse, sometimes severe, at night. A palpable soft tissue mass or fullness is present in 30% to 80% of patients at presentation. Pathologic fractures are the presenting symptom in 3% to 17% of patients.

Grading of chondrosarcoma is imperfect since there is imperfect correlation between histologic appearance and biologic behaviour and since features such as size, location, radiologic appearances play a major role in determining the outcome of these lesions. The grading system has become subjective. Group of M.D.Anderson suggested grading as follows

Grade 1 – Tumors composed of inoccuous hyaline cartilage or sparse cellularity containing cells with dark pyknotic nuclei no larger than a lymphocyte.

Group 2A – Lesions are more cellular. More than 20 percent of the cells have nuclei larger than size of lymphocytes. Binucleation and nucleoli may be seen, but mitoses are not identified.
Grade 2B – lesions are more cellular, have numerous binucleate cells, occasional bizzare nuclei or spindling. Cellular areas correspond to the periphery of lobules. Mitoses may be found but are less than 1 per 10 high power fields.
Grade 3 - Lesions contain 2 or more mitoses per 10 high power fields.[8]
Grade 2A and 3 have capacity for distant metastasis.

Therapy consists of resection of the mass along with synchronous metastatic lesions[5] and chest wall reconstruction accompanied by chemotherapy or radiotherapy. Reconstruction often uses a combination of myocutaneous flaps and prosthetic materials[7]. The presence of a malignant pleural effusion is a contraindication to surgical resection. Even large tumors can be successfully resected with a multidisciplinary team approach involving an oncosurgeon, cardiothoracic and vascular surgeon, plastic surgeon, intensivist, anaesthesiologist and good nursing care[9]. Chondrosarcomas are known to be relatively radio resistant. Doses exceeding 60 Gy are recommended for effective treatment, which is often not tolerable for adjacent structures (e.g., neurologic tissue). Chemotherapy is also ineffective as they have poor vascularity and abundant extracellular matrix except for the mesenchymal subtype. No standardized proven approach exists for adjuvant chemotherapy in the setting of chondrosarcoma. Cisplatin and Adriamycin–based regimens seem to have the most efficacy.

The natural history and prognosis of chondrosarcoma is extremely variable. Overall 5-year survival rates for grade 1 chondrosarcomas are 90% to 94%[3,4]. For grade 1 chondrosarcomas with intact cortex and absence of soft tissue mass, consideration can be given to an intralesional procedure such as curettage with adjunctive ablation. However, if there are aggressive imaging features such as cortical breakthrough or soft tissue mass, or if the tumour is grade 2 or higher, wide surgical excision is required. 5 year survival rates for high grade tumours are very low. Poor prognostic factors include incomplete resection, metastases, and local recurrence, as well as age over 50 years[5].

In this case tumour originated from ribs and involved major part of the right lung and there were bilateral pulmonary metastasis and no evidence of extra pulmonary metastasis. Wide surgical excision with 4 cm margin and reconstruction of chest wall was not possible. Also there were multiple lung metastases excision of which was not possible. Palliative therapy was advised.

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