Case 13550

Eurorad

Ewing sarcoma of rib

Published on 22.04.2016

DOI: 10.1594/EURORAD/CASE.13550
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Bones Thoracic wall Thorax
Procedure: Diagnostic procedure
Procedure: Contrast agent-intravenous
Procedure: Computer Applications-3D
Imaging Technique: CT
Special Focus: Neoplasia Calcifications / Calculi Case
Type: Clinical Cases
Authors: Pradosh Kumar Sarangi1, Sasmita Parida2, Jayashree Mohanty3
Patient: 12 years, male

Clinical History:

A 12-year-old boy from India presented with right-sided chest pain, intermittent fever and cough with expectoration for the past five months. He had no contact history of tuberculosis. His leukocyte count was 12000/mm^3 and ESR (Erythrocyte sedimentation rate) was 70mm/hr at the time of examination.

Imaging Findings:

He was initially seen by a general practitioner who started him with intravenous antibiotics and anti-tubercular therapy suspecting it to be a case of tubercular empyema as it is endemic in India. Chest radiograph showed opacified right hemithorax and rib destruction was not obvious at that time. There was no improvement in the patient's condition after a month of treatment and the patient was referred to our hospital for a CECT thorax.

Noncontrast CT scan showed a large heterogeneous soft tissue mass with calcific foci occupying almost the whole right hemithorax associated with pleural effusion. There was lytic destruction of right 3^{rd} rib with bone expansion without any significant periosteal reaction (Fig. 2, 3). The lesion was noted extending to the mediastinum with mediastinal shift to the left. It was abutting the right lateral chest wall without any extrathoracic extension. The mass lesion showed heterogeneous enhancement on contrast-enhanced CT (Fig. 4). Histopathological examination of the mass revealed Ewing sarcoma.

Discussion:

Ewing sarcoma (ES) is named after James Stephen Ewing, American pathologist (1866-1943), which he described as diffuse endothelioma or endothelial myeloma. It belongs to the Ewing sarcoma family of tumours (ESFT), which are small round blue cell tumours sharing common microscopic appearances and karyotyping abnormality [non-random t(11;22)(q24;q12) chromosome rearrangement resulting in formation of EWS-ETS fusion gene] [1, 2, 3].

Approximately 10% of all primary malignant bone tumours are Ewing sarcomas [4]. ES is the second most common primary malignant tumour of the bone after osteosarcoma in children and adolescents [3]. Ewing tumour is the most common cause of a malignant chest wall mass in children. The typical patient with Ewing sarcoma, notwithstanding the site of origin, presents between 10 to 25 years of age [2, 3, 4]. The male-to-female ratio is 2:1 for patients with primary Ewing sarcoma of the rib [4]. Most common sites affected are femur (21%), ilium (12%– 13%), tibia (8%–11%), humerus (10%), fibula (7%–9%), ribs (8%), and sacrum (6%). In long bones the most affected site is

metadiaphysis (44%), followed by middiaphysis (33%) and metaphysis (15%) [2].

Primary Ewing tumour of the rib usually presents with a clinical history of chest pain sometimes associated with a palpable chest wall mass .The rib lesions may show predominantly lytic (82%), mixed lytic sclerotic (9%) or sclerotic (9%) patterns [4, 5]. Thirty five percent of the affected ribs show an “expanded” contour. The lateral portion of the rib is most commonly affected [4]. Periosteal reaction in rib lesions is relatively insignificant, as compared with other bones involved by Ewing tumour. Ewing sarcoma of the rib penetrates the bone cortex rapidly with resultant formation of a soft tissue mass. The intrathoracic component is larger than the extrathoracic component and intraosseous involvement. The intrathoracic component sometimes obscures subtle bone changes on chest radiograph and can be mistaken for empyema when fever and pleural effusion are present. Computed tomography simplifies the diagnosis in such cases and can show precise extent, relationship to the adjacent structures, intraspinal extension and metastatic spread to the lungs and mediastinal lymph nodes [2, 4, 5, 6].

Treatment includes a combination of neoadjuvant chemotherapy followed by en bloc resection of rib and radiotherapy. The five year disease-free survival rate is now approximately 75% [2, 5].

TEACHING POINT
Whenever a young patient presents with pleural effusion, especially in tuberculosis endemic countries like India, it is worthwhile to perform histopathological examination before starting the patient on antituberculosis treatment.
Differential Diagnosis List: Ewing sarcoma of rib, Osteosarcoma, Chondrosarcoma, Tuberculous empyema with chest wall involvement, Thoracic actinomycosis

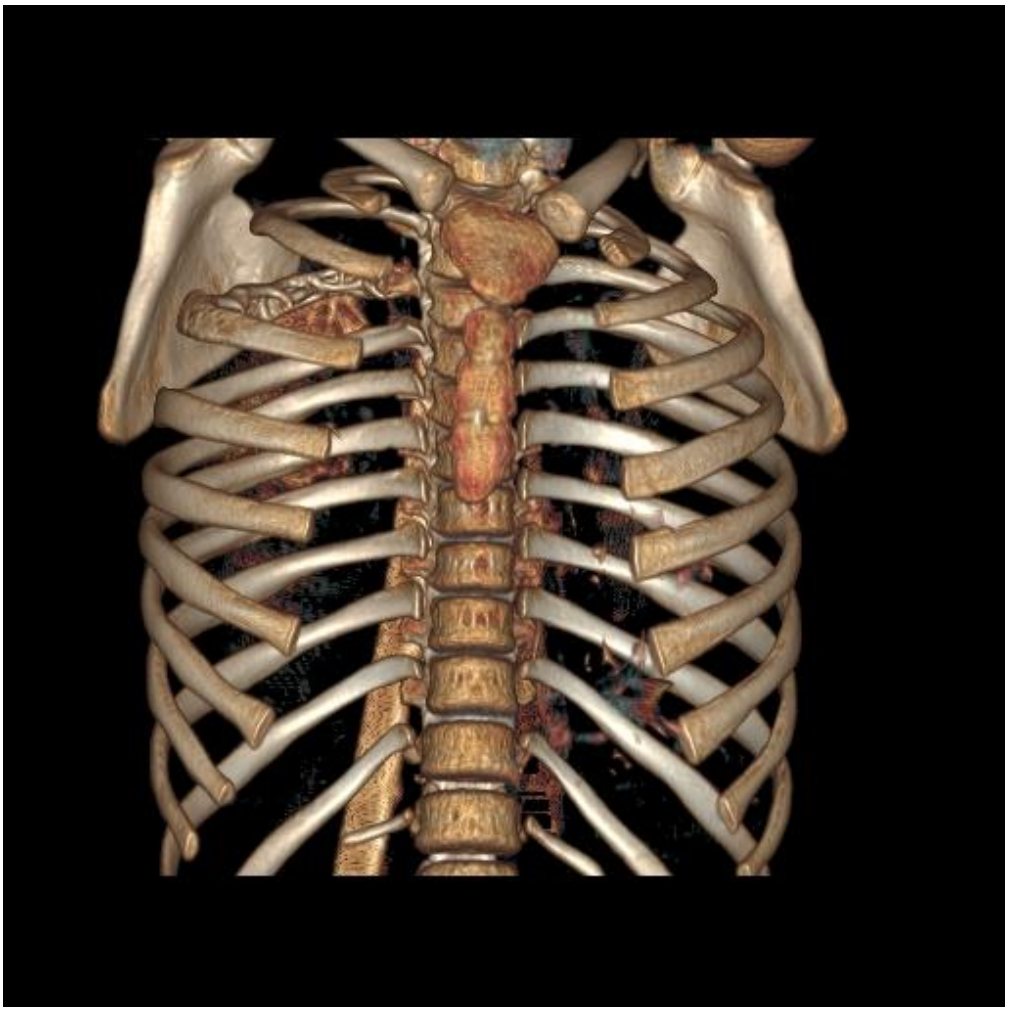
Final Diagnosis: Ewing sarcoma of rib

References:

Ewing J (1972) Classics in oncology. Diffuse endothelioma of bone. James Ewing. Proceedings of the New York Pathological Society, 1921. CA Cancer J Clin 22(2):95-8 (PMID: 4622125)
Murphey MD, Senchak LT, Mambalam PK, Logie CI, Klassen-Fischer MK, Kransdorf MJ (2013) From the radiologic pathology archives: ewing sarcoma family of tumors: radiologic-pathologic correlation. Radiographics 33(3):803-31 (PMID: 23674776)
Burchill SA (2003) Ewing's sarcoma: diagnostic, prognostic, and therapeutic implications of molecular abnormalities. J Clin Pathol 56(2):96-102 (PMID: 12560386)
Moser RP Jr, Davis MJ, Gilkey FW, Kransdorf MJ, Rosado de Christenson ML et al (1990) Primary Ewing sarcoma of rib. Radiographics 10(5):899-914 (PMID: 2217978)
Levine E, Levine C (1983) Ewing tumor of rib: radiologic findings and computed tomography contribution. Skeletal Radiol 9(4):227-33 (PMID: 6867771)
Widhe B, Widhe T, Bauer HC (2007) Ewing sarcoma of the rib--initial symptoms and clinical features: tumor missed at the first visit in 21 of 26 patients. Acta Orthop 78(6):840-4 (PMID: 18236193)

Figure 1

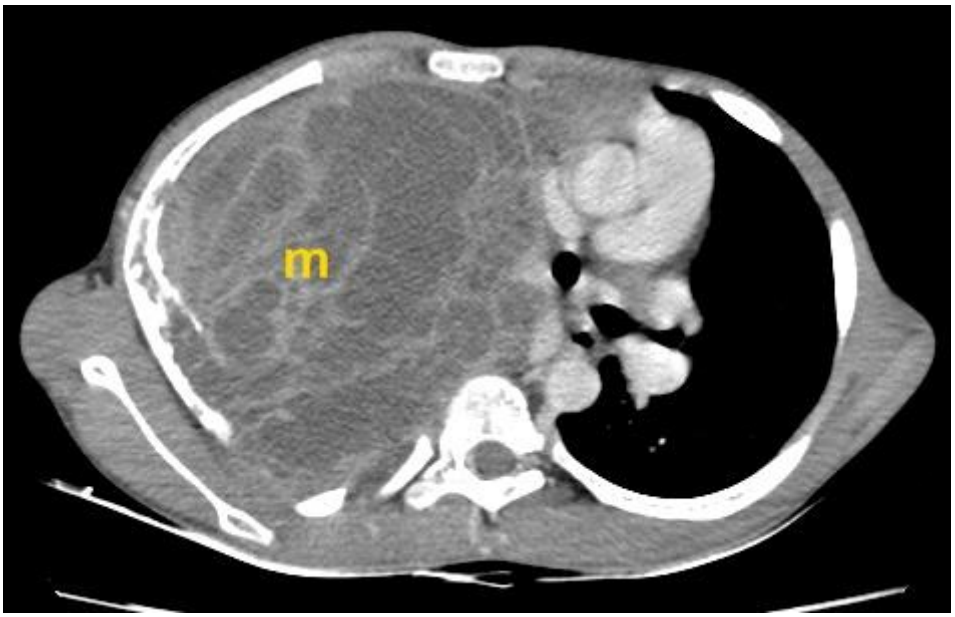
a



Description: Right third rib destruction (posterior and lateral segment). **Origin:** Dept of radiodiagnosis,SCB medical college, cuttack,

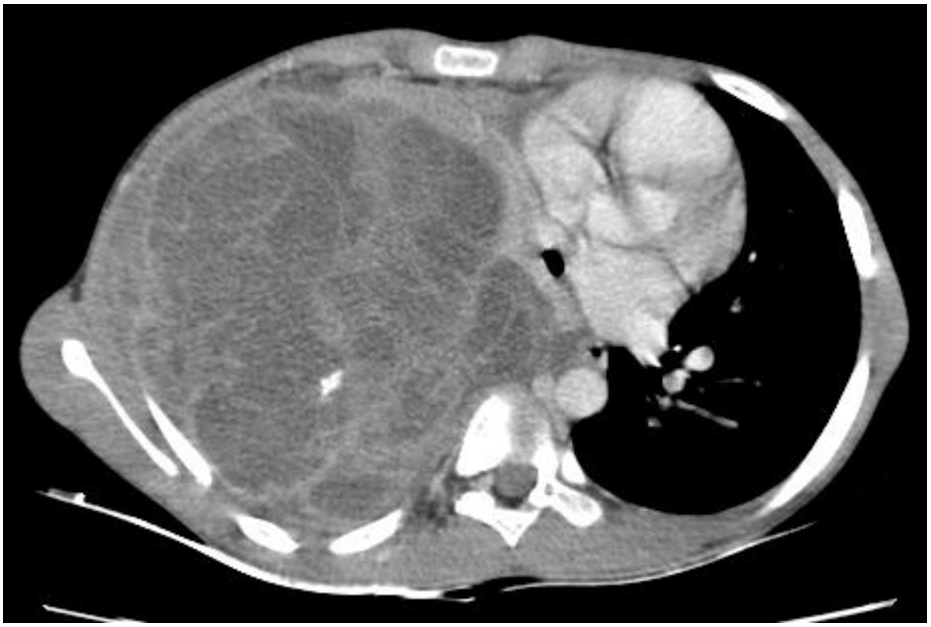
Figure 2

a



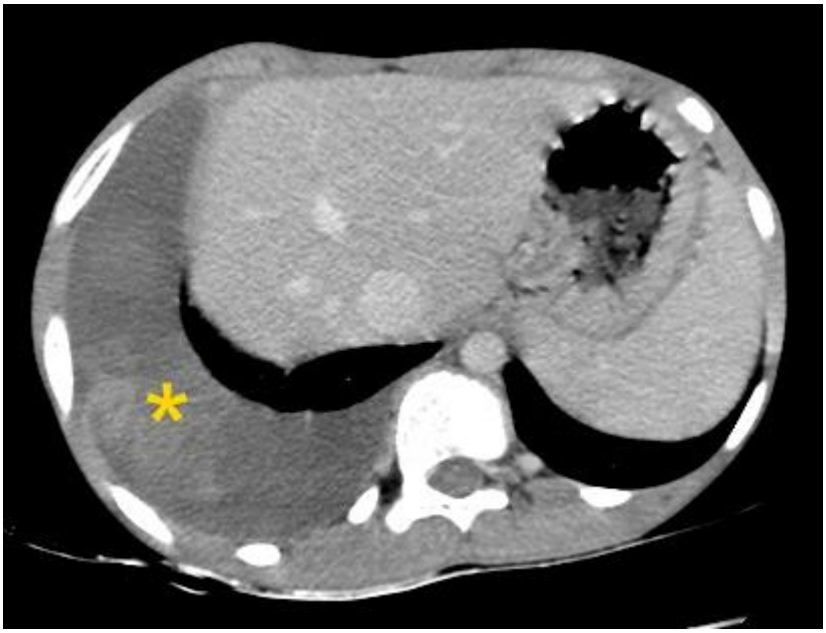
Description: Note the heterogeneous enhancement of the mass (m) with mediastinal extension.
Origin: Department of radiodiagnosis, SCB medical college, Cuttack, India

b



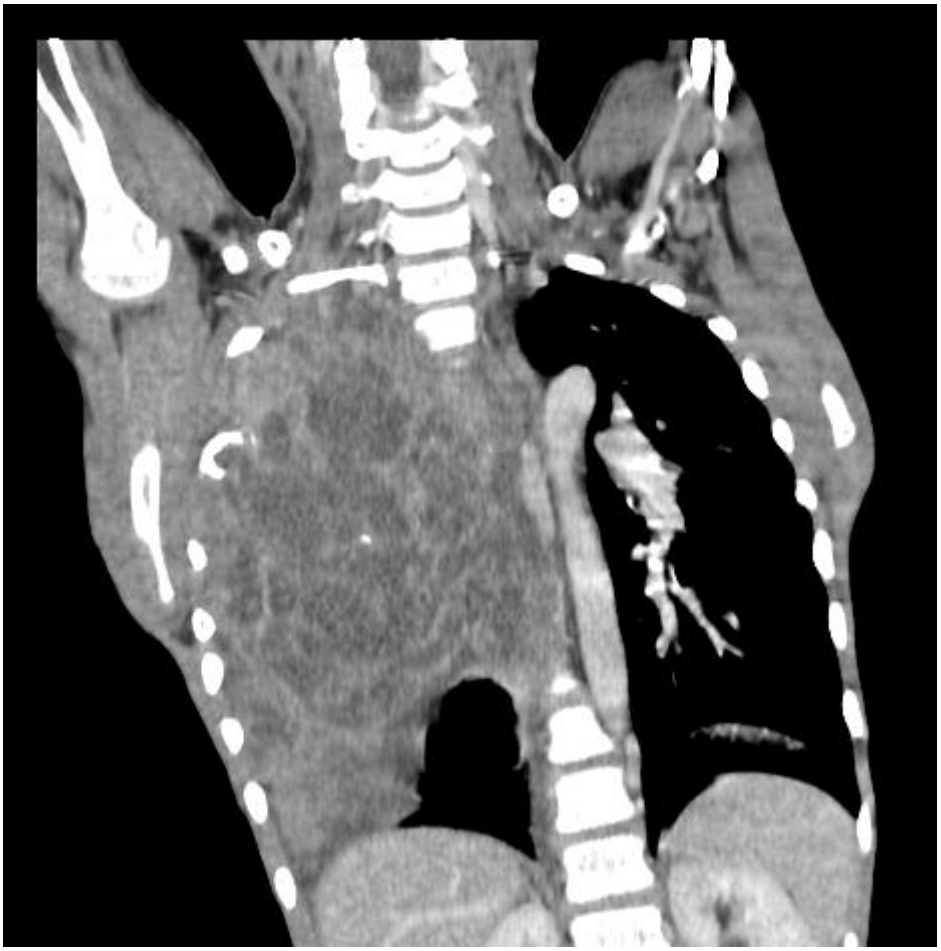
Description: Note the heterogeneous enhancement of the mass with mediastinal extension. **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

c



Description: Right pleural effusion (*) **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

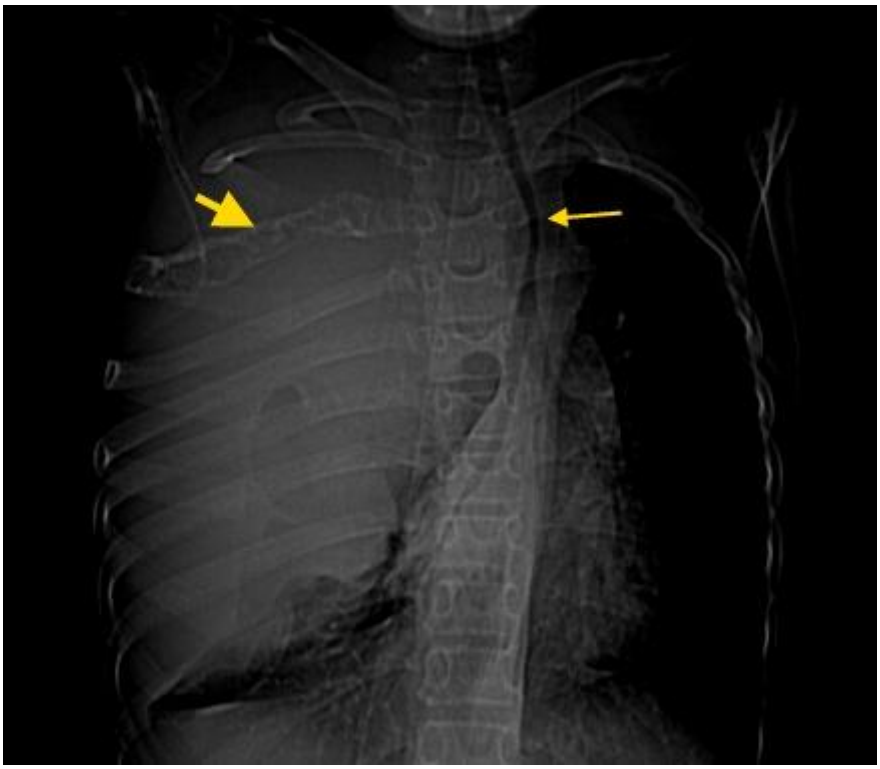
d



Description: Coronal image showing a large heterogenously enhancing mass occupying almost the whole right hemithorax. **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

Figure 3

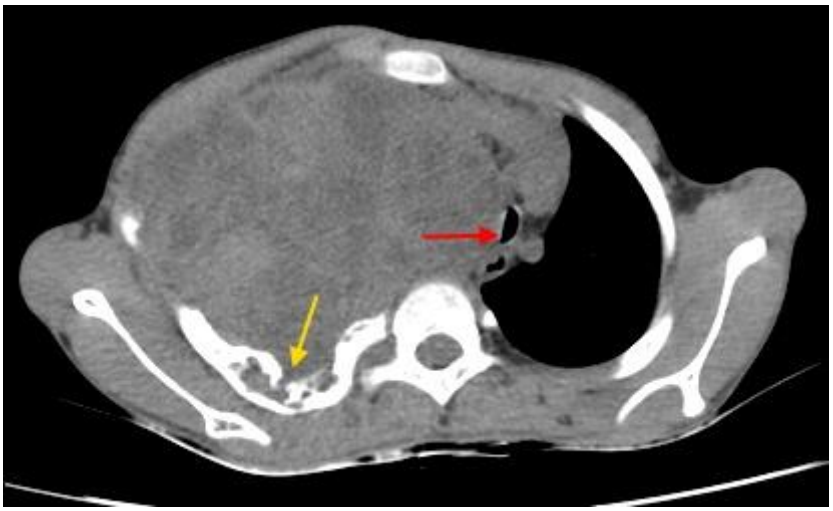
a



Description: Scanogram shows opacified right hemithorax with tracheal shift to left (long arrow) and destruction of right third rib (short arrow). **Origin:** Dept. of Radiodiagnosis, SCB medical college, Cuttack ,India

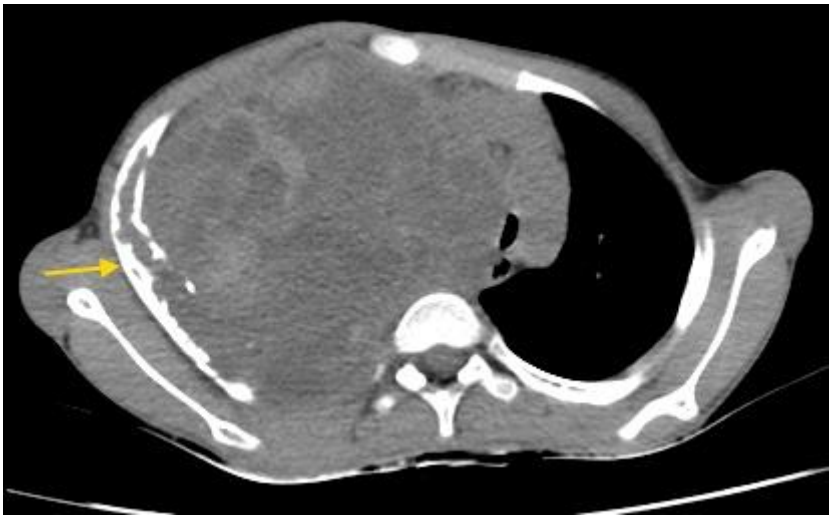
Figure 4

a



Description: Lytic bone destruction of right third rib (yellow arrow) with large heterogenous intrathoracic mass causing mediastinal shift to lthe eft (red arrow). **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

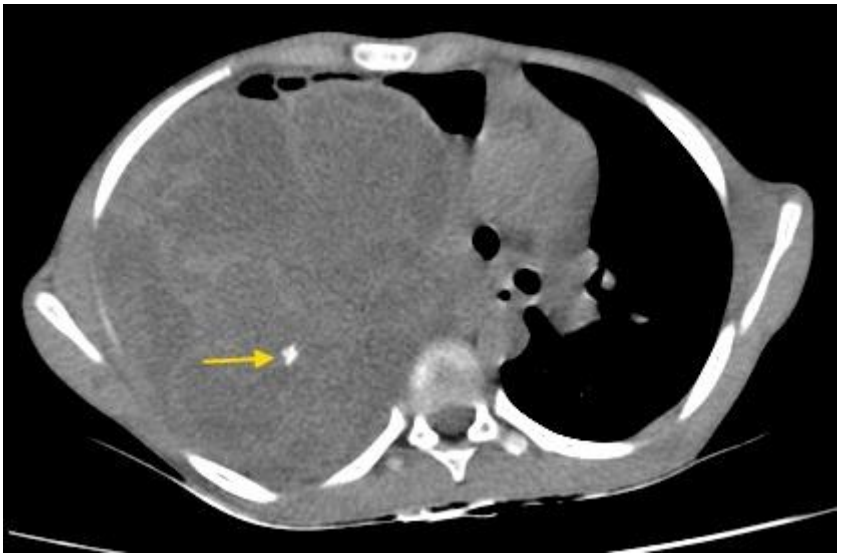
b



Description: Rib is expanded with cortical breach of inner margin.

Outer cortex is normal (arrow) without any extrathoracic extension. **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

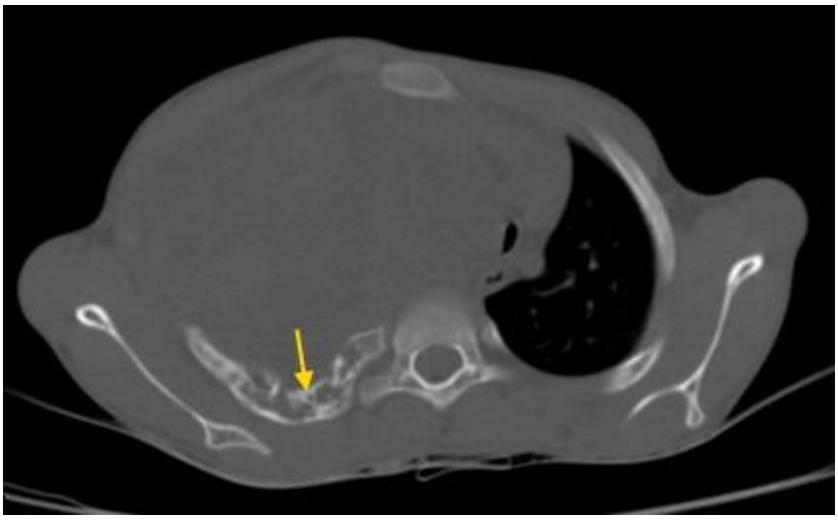
c



Description: Note the calcific focus (arrow) within the mass. **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

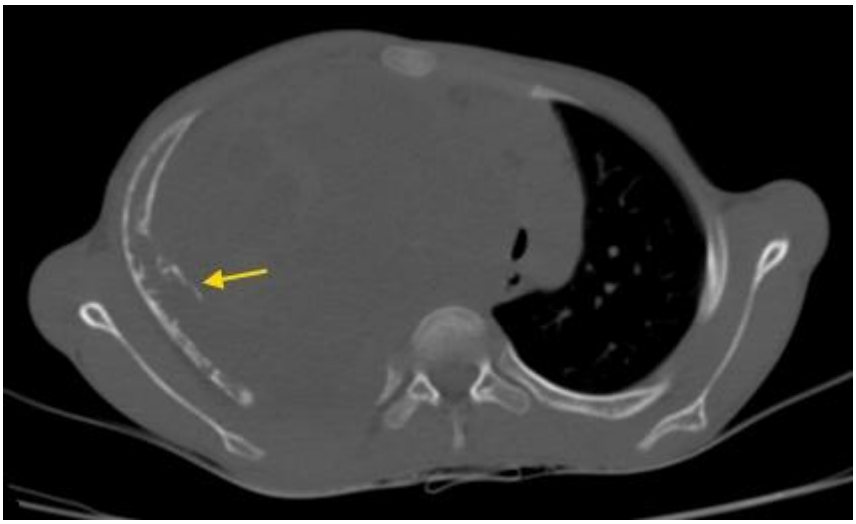
Figure 5

a



Description: Right third rib expansion and destruction involving the posterior segment (arrow). **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India

b



Description: Destruction of lateral segment of right third rib (arrow). Outer cortex is apparently intact without any extrathoracic soft tissue component. **Origin:** Department of radiodiagnosis, SCB medical college, Cuttack, India