

Case Report

DIAGNOSING EWING'S SARCOMA OF THE RIB IN CHILDREN – IS IT QUITE CHALLENGING??

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ABSTRACT

Ewing's sarcoma is a primary malignant bone neoplasm in children which accounts for 25 % of all malignant bone tumours. It more commonly occurs in Pelvic bones & Lower extremities. Approximately 10 % of all Ewing's Sarcoma cases arise from the ribs. Ewing's Sarcoma in the Thoraco-pulmonary region is called as Askin's tumour. We present a case of Ewing's Sarcoma of the 8th rib on the right side in a 11yr old boy.

Key Words: Ewing's Sarcoma, Askin's Tumour, Ribs

INTRODUCTION

Ewing's sarcoma is the fourth most common primary malignancy of bone & second most common in patients younger than 30 years of age. It usually occurs in diaphysis of long bones, with male predominance 1.6:1. In 1921, Sir James Ewing was the first to describe it as non-osteogenic primary tumour of the bone characterized by the presence of small round cells which he termed as "Endotheliomyeloma of bone" (Ewing, 1922). In pediatric age groups common malignant tumour involving chest wall are Ewing's sarcoma, rhabdomyosarcoma and chondrosarcoma (Jugens, 1994). Askin & his colleagues described malignant small round cell tumour of bony chest wall during childhood, hence Ewing's sarcoma of ribs is named as Askin's tumour (Cabezali *et al.*, 1994). Pain & swelling are the most presenting symptoms, sometimes can be associated with intermittent fever. Any haemorrhagic pleural effusion is usually rare presentation. Though Ewing's sarcoma is one of commonest chest wall malignant tumour, Physicians & Orthopaedicians have found it challenging in diagnosing this condition early, which could affect the prognosis of the patient. So, we present a case of 11yr old boy, diagnosed to have Ewing's sarcoma of right side 8th rib, which could have been diagnosed at a much earlier stage.

CASES

11 years old boy was brought with c/o Pain over the right chest wall (Lateral aspect) on & off for past 6 months, which was associated with fever (low-grade) on & off, Loss of weight inspite of good appetite. For the last 20 days mother of the boy had noted a swelling over the right chest wall which increased in size. He had no c/o cough, sputum expectoration or difficulty in breathing. They had already consulted certain physicians (atleast 3 to 4 times) during initial phase of chest pain, but were neglected as a musculocutaneous pain. On examination, Patient was afebrile & no evidence of any cardio or respiratory system abnormality. Chest examination revealed a swelling of size 5 X 3 cms in the right side of the chest wall over Mid-Axillary line extending along the course of 8th rib with well defined posterior margin & ill-defined anterior margin. Swelling was warm & tender on touch, surface of swelling was smooth, Bony-hard in consistency & immobile but moves along with ribs during respiration. Skin over the swelling was normal & skin was pinchable & moveable. Blood parameters were near normal, except for elevated ESR – 26 mm. (Figure 1) Plain X-ray of chest showed permeative lytic lesion along the course of the 8th rib. (Figure 2) Contrast enhanced CT chest showed destructive lesion in the lateral aspect of 8th extending both anteriorly & posteriorly with associated large intra & extra-thoracic soft tissue component of size 12x4x4.6 cms suggestive of small round cell tumour. There was no evidence of focal metastasis or mediastenal Lymph node enlargement. A open biopsy of the swelling over 8th rib was performed & sent for HPE which revealed (Figure 3) sheets of small to medium sized cells with vesicular nuclei, few

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conspicuous nucleoli & scant eosinophilic to vacuolated cytoplasm which were consistent Ewing's sarcoma or Primitive Neuroectodermal Tumour (PNET). Immunohistochemical studies were done & the tumour cells were found to be CD99 positive & LCA negative. Finally the case was diagnosed as Ewing's sarcoma of the 8th rib. Patient was referred to other hospital for chemotherapy & further management.

DISCUSSION

Ewing's Sarcoma is the second most common primary malignant tumours in children. It usually originates from diaphysis, but can present as skip lesions also. It commonly affects the Shafts of Long bones, followed by Pelvis & sacrum. Approximately 10 % of the primary ewing's sarcoma occurs in ribs, Primary malignant bony chest wall tumours are usually uncommon which include Chondrosarcoma, Ewing's sarcoma, solitary Plasacytoma, lymphoma & desmoid tumours (Jugens, 1994). The commonest presentation of Ewing's Sarcoma of a rib is pain and palpable mass (Mirra, 1989). Sometimes it may be associated fever, pathological fracture, breathlessness due to pleural effusion / atelectasis. Radiological features of ewing's sarcoma includes permeative lesion with lytic, moth-eaten & mottled appearance of the medullary cavity of the bone, invasion of overlying cortex reflects rapid bone destruction, Periosteal new bone formation may extend along the shaft in case of long bobs which appears as fusiform layers of bone around the lesion giving "onion peel" appearance, tumour extends into surrounding soft-tissues, with radiating streaks of ossification gives a "sunray appearance" & "codman's triangle". In case of Ewing's sarcoma of ribs there will be typical permeative with lytic, moth-eaten or mottled appearance of the involved ribs (Wilner, 1982). Histological features of Ewing's sarcoma of long bones & ribs are similar which includes (Mascimento, 1980).

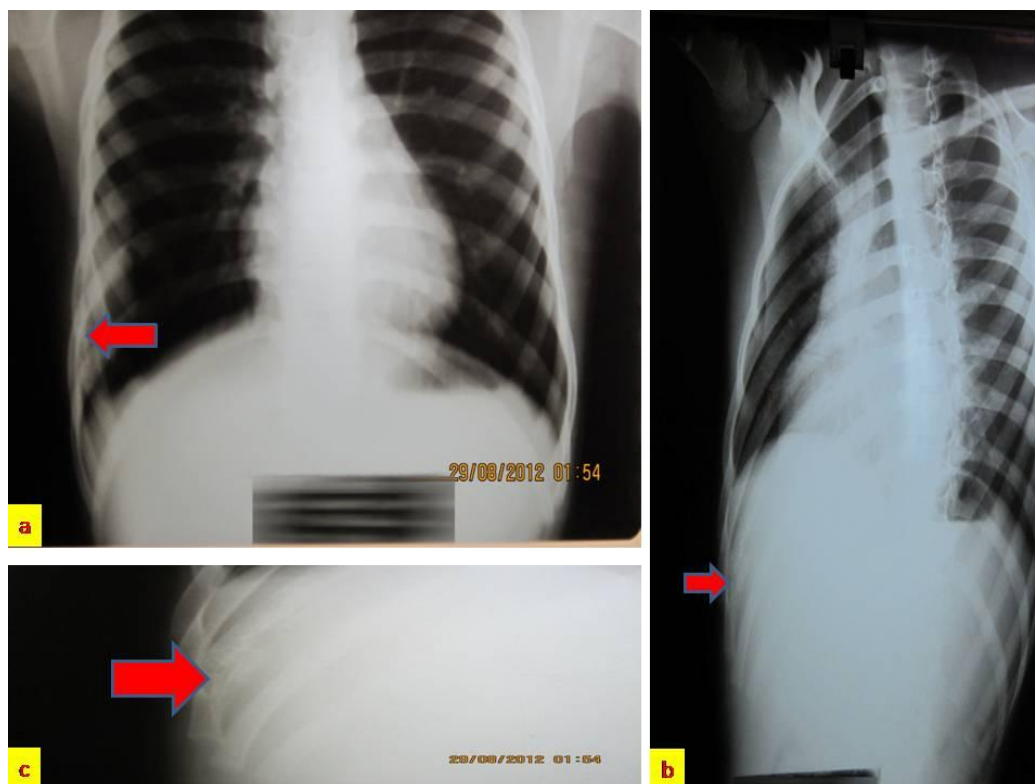


Figure 1: (a) shows destructive lesion of 8th rib on the right side. (b) Shows left lateral view of chest showing an erosive destruction of the right side 8th rib. (c) Is enlarged view of previous x-ray showing the destructive lesion along the course of the solitary rib

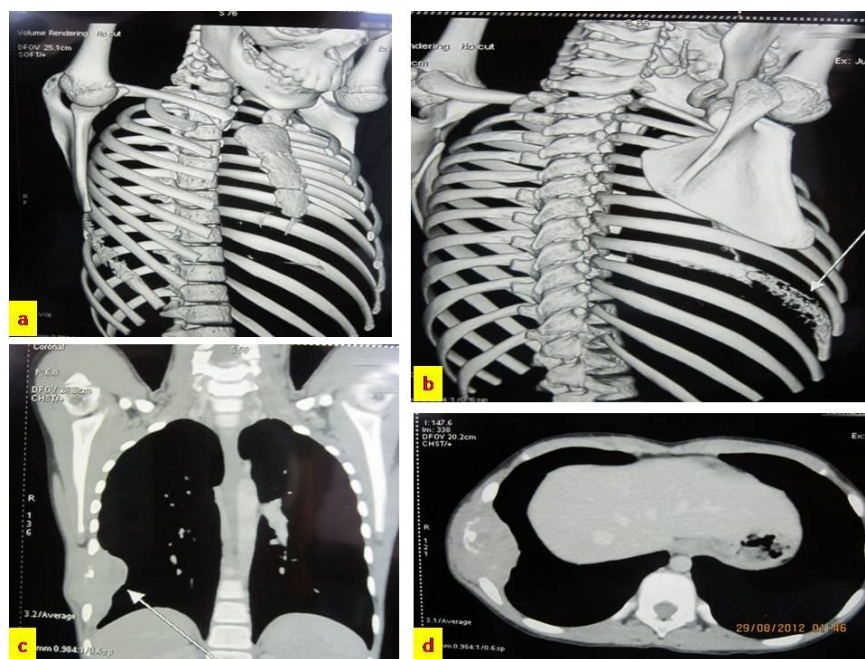


Figure 2: (a) & (b) shows 3D reconstructed view of chest showing destruction of the 8th rib from posterior to anterior third margin. (c) & (d) ct images showing extra & intra-thoracic soft tissue involvement, without any involvement of lung parenchymal tissues

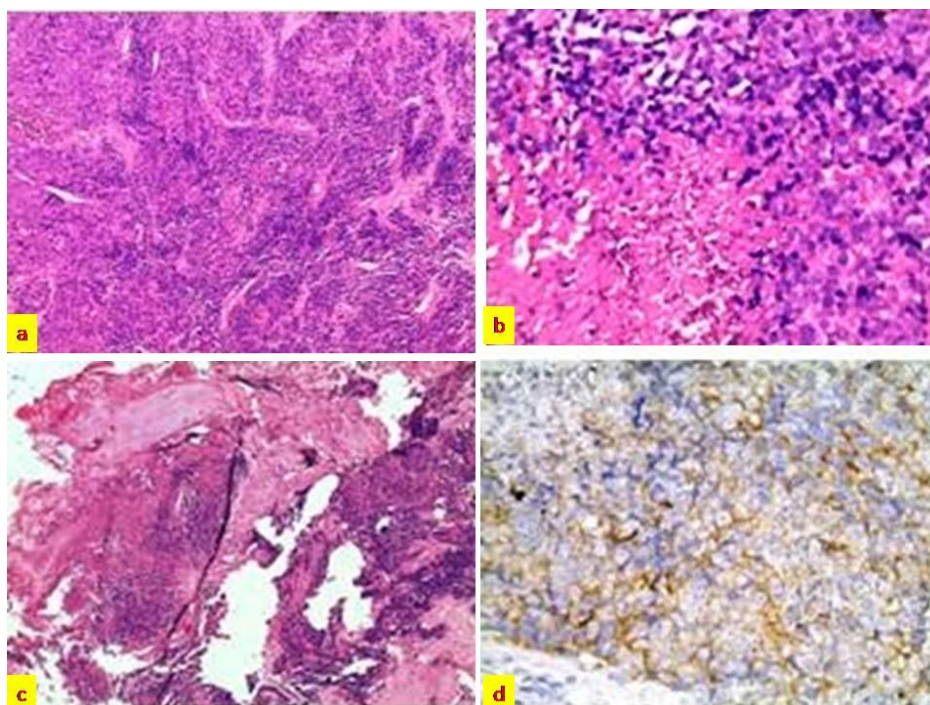


Figure 3: Histological pictures of the excised tumour. (a) Sheets of small round blue cells (b) Small round Blue cells with areas of necrosis. (c) Rib cartilage cells with adjacent neoplastic cells (d) CD 99 positive cells

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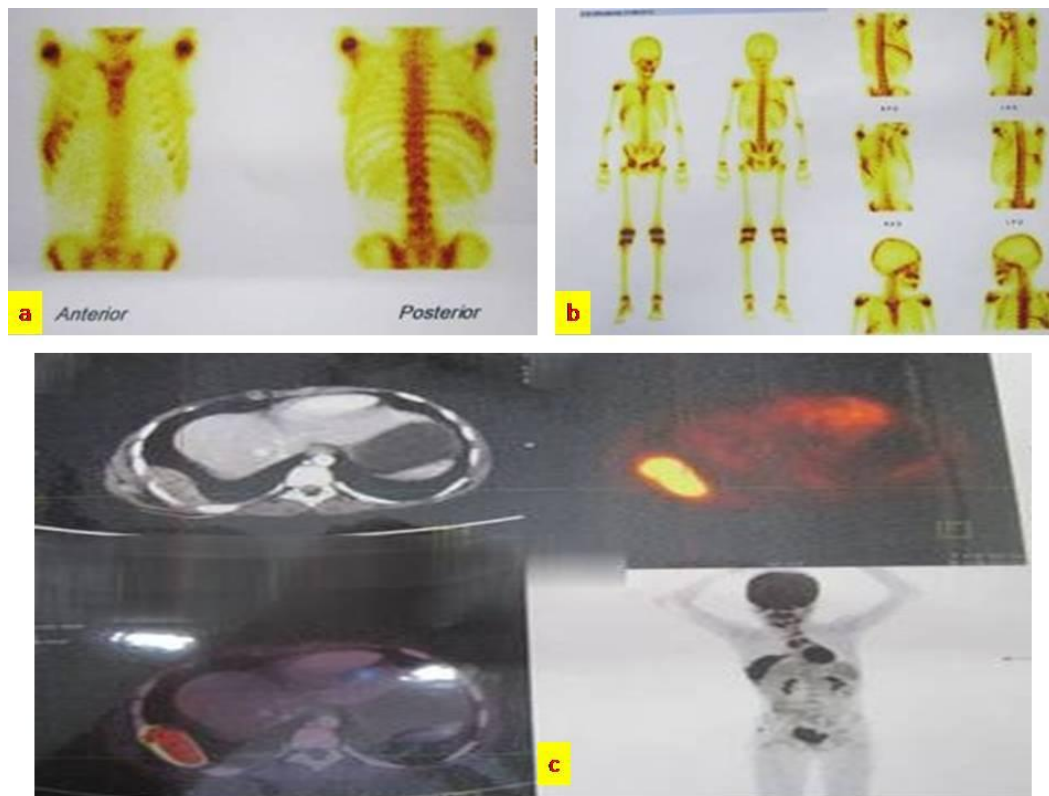


Figure 4: (a) & (b) whole body scan images showed increased uptake in the right chest wall 8th rib alone. (c) PET-CT image of the whole body showed mediastinal node metastasis at the time of chemotherapy

1. Compact sheets of small blue polyhedral cells which are haematoxylin stained with pale scant cytoplasm & ill-defined boundaries.
2. The nuclei are uniform round or oval & contain scattered chromatin.
3. Presence of glycogen makes it periodic acid-schiff (PAS) positive
4. Devoid of reticulin fibres
5. Immunohistochemical tests shows t(11,22) (q24, q12) is the most common translocation diagnostic of ewing's sarcoma, in > 90 % of cases (Genet, 1992), CD 99 positive & LCA negative.

Differential diagnosis of Ewing's sarcoma include multiple entities of diverse cause such as metastasis, lymphoma, myeloma, neuroblastoma, osteosarcoma, Pancoast tumour, Pleural – based masses, pneumonia, eosinophilic granuloma & osteomyelitis (Rose, 1983; Omel, 1973; Senac, 1986). This long list of diagnoses can be readily narrowed down with proper combination of clinical, radiological & histological assessment. For instance the average age of presentation of Ewing's sarcoma is around 12.4 yrs. So neuroblastoma could be ruled out which occurs at a much younger age group. Metastases, lymphoma, myeloma & Pancoast tumour occurs in relatively older age group. Pancoast tumour occurs in apex of the lung (extra-osseous origin). Osteomyelitis & Eosinophilic granuloma don't have extensive soft tissue involvement as Ewing's sarcoma does. However if Ewing's sarcoma is confined to ribs alone then it will have permeative, moth – eaten appearance which will be indistinguishable from eosinophilic granuloma or osteomyelitis. Sometimes Ewing's sarcoma with soft tissue component can be difficult to differentiate from pleural based masses. In such cases careful radiological assessment may reveal intra-osseous involvement of the solitary rib. But in case of doubt it always better to evaluate with CT or MR imaging. Enchondromas & Fibrous dysplasia of ribs are unlikely to be confused with primary ewing's sarcoma of

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the rib due to typical features of Lobulated configuration with mineralization & flecks of calcification in tumour matrix in case of enchondromas, expansile lesion of rib in case of fibrous dysplasia (Kransdorf, 1990). Osteosarcoma of rib is very uncommon, but sometimes the telangiectatic variant of osteosarcoma is sometimes mistaken for lytic Ewing's sarcoma of the rib (Issacs, 1986). Histologically other small blue cell tumours can be differentiated with Immunohistochemical tests: Lymphoma – LCA positive & PAS negative, Rhabdomyosarcoma – positive for desmin & myoglobin makers, Neuroblastoma – positive for neuroectodermal markers, Eosinophilic granuloma is Reticulin positive, PAS positive. So diagnosing Ewing's sarcoma at an earlier stage is of prime importance as it's directly related to prognosis of the patient. Since the tumour is chemosensitive, which also decreases the size of tumour & provides a 75% long term survival rate (20 years) if the tumour is localized to a particular region. Ewing's sarcoma is an aggressive tumour which is associated with bone marrow & Pulmonary metastasis, in which the later one has a slightly better prognosis. Sometimes they can be associated with lymph nodal & liver metastasis also. In case of metastasis the treatment outcome is usually poor & the 5 yr survival is only 25 %. So in case of suspected Ewing's sarcoma of any part of the bone rapid diagnosis & Aggressive management with chemotherapy followed by surgical intervention for accessible sites is the order of the day.

Ewing's sarcoma is one the aggressive primary malignant tumours of bone which if diagnosed early, patient (mostly children) will have a much better prognosis. In our case since the patient presented to us only after 6 months, PETCT showed mediastinal nodal metastasis at the time of starting chemotherapy (Figure 4), which would grossly affect the prognosis in our patient. So in order to have an earlier diagnosis most of the Radiological education must be focused on:

- i) Recognition of radiological abnormality in x-rays, CT & MR imaging
- ii) Any unexplained chest pain without any h/o trauma, with c/o of fever on & off & constitutional symptoms should be evaluated for Ewing's sarcoma.

So in this report we have used a better educational approach that begins with a specific disease - the Ewing's sarcoma of the rib in detail & familiarize the reader with all the radiologic manifestations of this disease. This approach should facilitate the earlier correct diagnosis which will result in effective treatment & more favourable outcome for the patient afflicted with primary Ewing's sarcoma of the rib (Thomas, 1983).

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