



Giant extraosseous Ewing sarcoma of the lung in a young adolescent female – A case report

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Extraosseous Ewing sarcoma is a rare soft tissue tumour that is histologically indistinguishable from the bone Ewing sarcoma. The translocation involving chromosome 22 along with CD 99 expression is pathognomonic and is useful in differentiating from other small round cell tumours. Primary lung involvement by this malignant tumour is very uncommon and up to this date only ten cases have been reported. We report a further case in a 15 year-old-female who presented with a huge lung mass causing an opaque haemithorax.

Keywords : extraosseous Ewing sarcoma; lung.

INTRODUCTION

Ewing's Sarcoma is a malignant neoplasm of bone. Very rarely it arises solely from soft tissue without bone involvement. It is termed as extraosseous Ewing sarcoma (EES) and is histologically indistinguishable from the bony Ewing sarcoma. The diagnosis is mainly based on microscopic histology and immunohistochemistry. The role of computed tomography (CT) scan is to define the extent of the tumour and to confirm that the soft tissue mass is totally extraosseous. Extraosseous Ewing sarcoma was first reported by Tefft *et al* (8). It usually involves the paravertebral region mainly at the lumbar and sacral level. A primary EES of the lung is exceedingly rare. Up to this date ten cases of primary lung EES have been reported (3,6). The giant nature of the tumour makes our case clinically unique.

CASE REPORT

A 15-year-old adolescent female presented with a 4-month history of a progressively increasing dry cough and right-sided pleuritic chest pain associated with mild swelling over the chest wall. On respiratory examination the air entry was markedly decreased and the percussion note was dull on the right side. There was no history of fever, haemoptysis, joint pain, rash or photosensitivity. No lymph node was palpable and the remaining physical examination was normal. Cardiovascular and abdominal examination did not reveal any abnormality. She had no significant past medical history and did not smoke.

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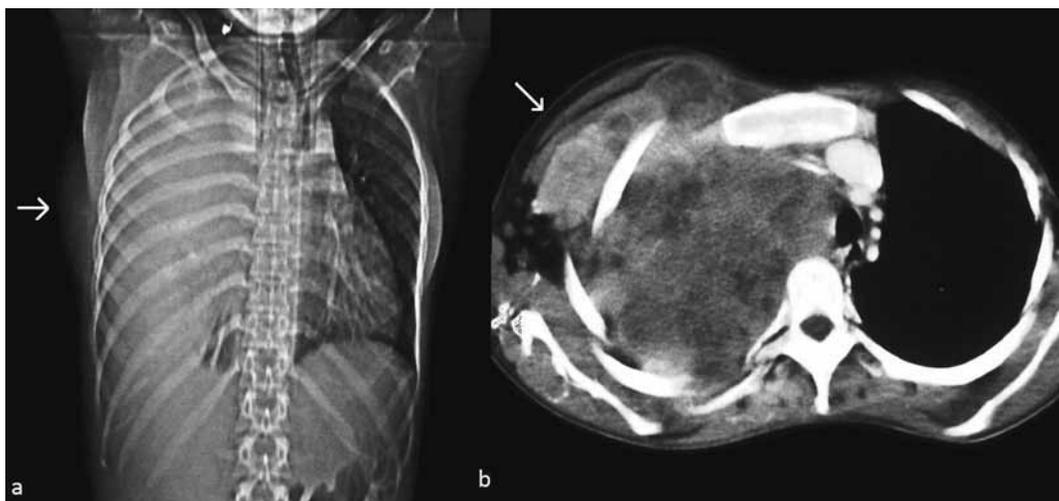


Fig. 1. — (a) Frontal CT scout view demonstrating an opaque right hemithorax with mediastinal shift towards the left. No evidence of any bony destruction was evident ; (b) CT of the chest revealing a lobulated lung mass infiltrating the chest wall.

Laboratory evaluations revealed a haemoglobin of 7 gm/dl, a white blood cell count of $8000/\text{mm}^3$ and a platelet count of $3 \times 10^5/\text{mm}^3$. Liver and renal functions were normal.

A chest radiograph demonstrated a completely opaque right hemithorax with mediastinal shift towards the left side. There was a soft tissue prominence on the right side ; however no evidence of any bony destruction was evident (Fig. 1a).

Computed tomography (CT) examination (Fig. 1b, 2a-b) of the chest revealed a heterogeneous lobulated mass with multiple hypodense areas involving the whole right hemithorax associated with a compressive atelectasis. No calcification was seen within the mass. There was some infiltration of the right antero-lateral chest wall but it was distinctively separated from the bone. All adjacent ribs and vertebrae were intact indicating thereby that the tumour had not arisen from the bone. The mediastinum was shifted towards the left side and a solitary pulmonary nodule was seen in the left lower lobe. Positron emission tomography (PET) scan showed increased uptake with standard uptake value (SUV) of 6.8.

Biopsy of the mass was done and histopathology (Fig. 3a) showed neoplastic proliferation of ovoid and spindle cells with vesicular nuclei and scanty

neoplasm in a pseudorosette pattern. The cells also contained periodic acid-Schiff stain (PAS) -positive inclusions. Immunohistochemical stain was positive for CD99 (Cluster of Differentiation 99) (Fig. 3b), S-100 protein and neuron-specific enolase (NSE). A CT guided biopsy of the left lobe nodule was consistent with a metastatic lesion.

Further clinical and radiological evaluation including a whole body CT scan, PET scan and bone scan revealed no evidence of an occult primary lesion. Clinico-radiological and immune-histological features were consistent with the diagnosis of extraskeletal Ewing sarcoma.

She was started on chemotherapy with Cyclophosphamide, Vincristin, Adriamycin, Epirubicin and Dacarbazine. Four courses of chemotherapy with a one-month interval were given along with two courses of radiotherapy. Unfortunately eight months later the patient died.

DISCUSSION

The group of Ewing sarcoma's comprises primary osseous and extraskeletal tumours. Primitive neuroectodermal tumours (PNET) and extra osseous Ewing sarcoma's (EES) are widely regarded as the same entity showing varying degrees of

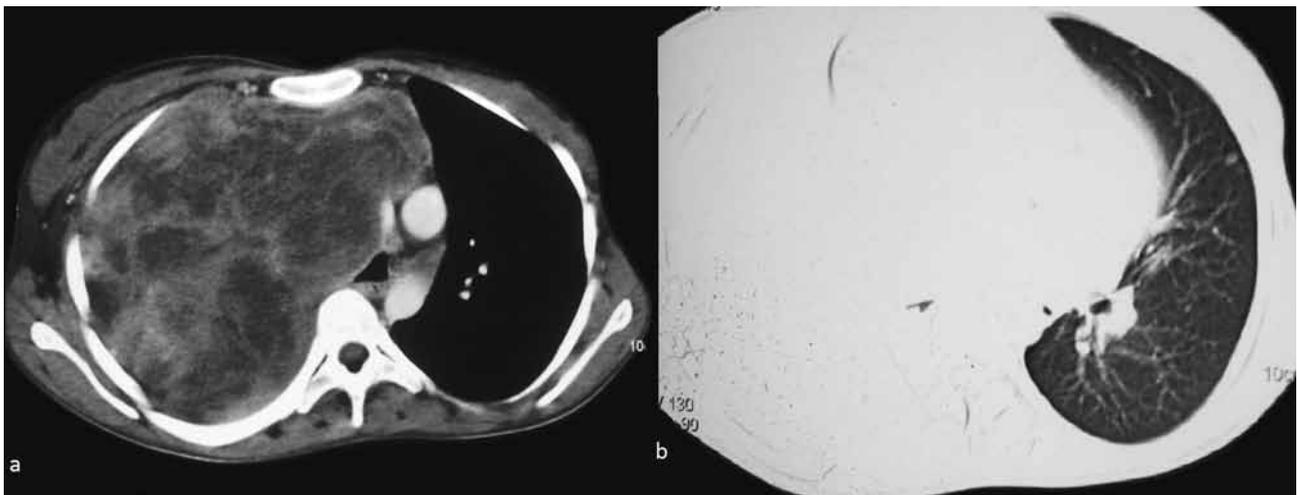


Fig. 2. — (a) CT scan of the chest demonstrating a large heterogeneous non-calcified mass filling the entire right hemithorax ; (b) CT with lung window setting showing a small nodule in the left lower lung field.

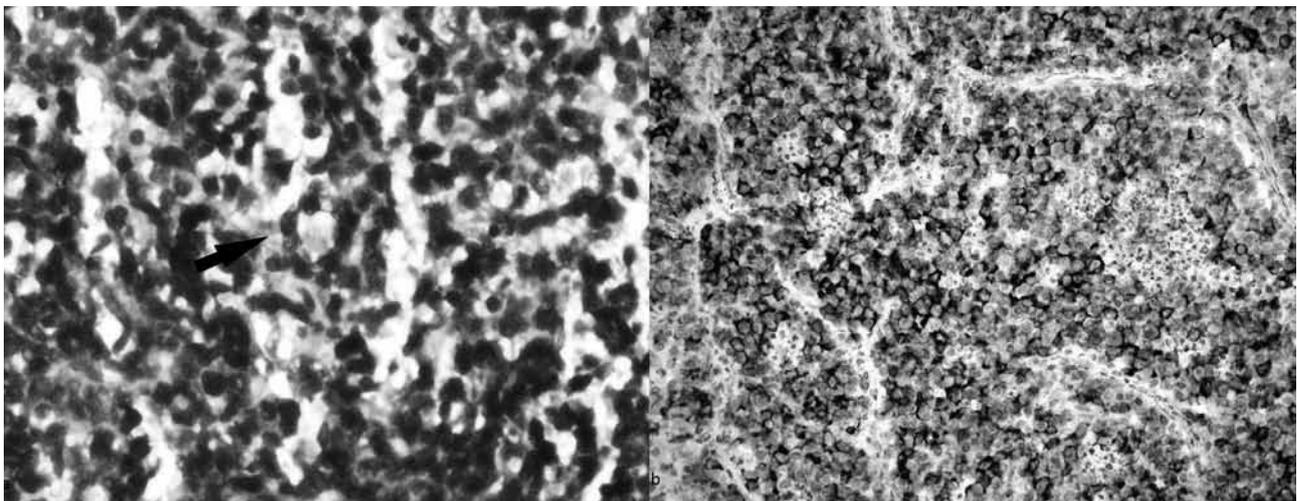


Fig. 3. — (a) Histopathology slides showing small round cells with vesicular nuclei and scanty cytoplasm in lobules separated by bands of fibrous connective tissue ; (b) Immunohistochemical stain for CD99 shows diffuse strong positivity in cytoplasmic membrane of tumour cells.

neuroectodermal differentiation (2). A common genetic translocation involving chromosome 22 is found in these tumours. The translocation $t(11;22)(q24;q12)$ is most common and this along with CD 99 expression is pathognomonic for these tumours and is useful in differentiating from other small round cell tumours occurring in childhood and adolescence. The histological differential diagnosis includes malignant lymphoma, rhabdo-

mysarcoma and neuroblastoma. Ewing sarcomas are positive for glycogen (PAS, 80%), neuron-specific enolase (60%), S-100 protein (50%), and MIC-2 marker (90%) while negative for leukocyte common antigen, epithelial membrane antigen, cytokeratin, desmin, vimentin, myoglobin, and glial fibrillary acidic protein (6).

EES is a rare soft tissue tumour that is histologically indistinguishable from the osseous form.

Immuno-histological findings along with the absence of bony involvement at the time of presentation is required for diagnosis.

Angervall *et al* in their series of 39 cases of EES reported the most common site to be the paravertebral region mainly at the lumbar and sacral level (1). Subsequently it was also reported involving the soft tissues of the orbit, vagina, kidney and the posterior mediastinum (5). A primary EES of the lung is exceedingly rare. The first case was reported by Hammer *et al* in 1989 (3). Since, ten cases of primary lung EES have been reported (3,6). We report the eleventh case. None of the previously described cases was so massive as to cause an opaque hemithorax. The giant nature of the tumour makes our case clinically unique.

The age of the patients with EES ranges from 20 months to 30 years, with a peak incidence at approximately 20 years of age. There is a slight male predominance. EES is usually manifested as a solitary soft tissue mass and may be superficially or deeply located. It is usually painless, but occasionally may cause local symptoms. Growth is rapid and distant metastases are frequent. Plain radiographs show EES as an extraosseous soft tissue mass without any calcified matrix. However, reactive changes such as periosteal proliferation, cortical thickening, bony erosion or sclerosis may occur.

CT scan is useful in delineating the extent of the tumour and to confirm that the soft tissue mass is totally extraosseous. Although the diagnosis of EES is primarily based on histology, some clinical and radiological features may help in the differential diagnosis from other sarcomas (4). Calcifications and fatty contents are unusual in EES. The presence of fat favours the diagnosis of liposarcoma and calcifications are commonly seen in malignant fibrous histiocytoma. Moreover, malignant fibrous histio-

cytoma is usually seen after the age of 45 and liposarcoma after the age of 30.

Since EES is an aggressive malignant tumour, the treatment of choice is an early surgical removal with intensive chemotherapy and radiation therapy to ablate any residual microscopic disease (7). Local recurrences and metastases are common. At the time of diagnosis approximately 25% of patients have metastases (2), and this is the most significant prognostic factor with a 5-year survival rate of less than 30% (6). The most common sites of metastases are the lung and the bone. Tumour size is not of much prognostic value. A younger age at time of diagnosis is associated with improved survival. The disease recurs in up to 40% of patients, which is associated with poor prognosis (2).

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