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Case Report

EXTRA OSSEOUS EWING'S SARCOMA- A CASE REPORT

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ABSTRACT

Extra osseous (extra skeletal) Ewing's sarcoma accounts for 1-2% of all soft tissue tumours in young adults with a predilection for males. The prognosis of patients aged <16 years of age is favourable. Here we report a case of 13 year old female patient who presented with a swelling in the right upper paravertebral region measuring 20×20×15cms with attached pleura and lung tissue. She was operated and histopathological study revealed it to be extra skeletal Ewing's sarcoma. However post operatively the patient received radiotherapy and was doing well.

INTRODUCTION

The Ewing's sarcoma is an uncommon malignancy, but it is second most common primary bone malignancy of childhood. It usually arises in the bone and is associated with soft tissue extension in 90% of cases [1]. It accounts for 1-2% of all malignant soft tissue tumours[2,3]. It is highly malignant tumour composed of small round cell[13]. It was first described by James Ewing in 1921 as 'diffuse mesothelioma of bone'[4]. In 1969, Taffe *et al* described a series of 5 patients with round cell tumour that arose in paravertebral soft tissues. They believed 4 of these tumours were related to Ewing's sarcoma but were unusual because of paucity of bony changes on plain film[5].

Subsequently, a number of cases were published describing the extra- skeletal variant of Ewing's sarcoma. It usually arises from paravertebral soft tissues of the trunk and the extremities. But here we present a case which had attached pleura and lung tissues which is very rare. Thus, we draw attention to the fact that Ewing's sarcoma should be thought in the differential diagnosis of any thoracic mass.

CASE REPORT

A 13 year old female patient presented with swelling in the right paravertebral region for past 3 months. On physical examination, a tender, hard swelling measuring 10 × 8 cms was palpated with restricted mobility. Chest X-ray revealed an opacity in the right lower zone (figure 1).

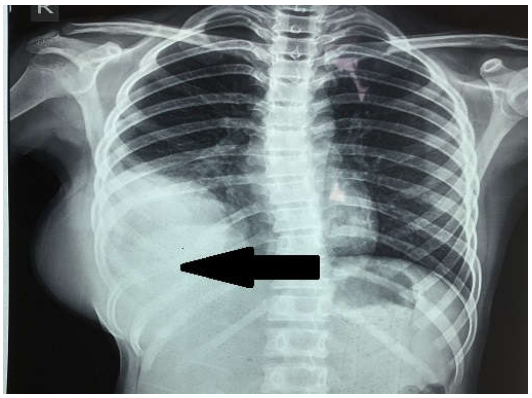

Figure 1- Chest X-ray of the patient revealed an opacity in the right lower zone (marked with arrow)

Fine needle aspiration revealed atypical mesenchymal tumour. CT scan of chest revealed a SOL measuring 20×20×15 cms arising from the right chest wall (figure 2). She was operated and a mass measuring 15×15×10 cms was resected from the right lower chest wall infiltrating the 8th and 9th ribs, densely adherent to diaphragm and the medial basal segment of lung along with overlying parietal pleura. 500ml of serosanguinous pleural fluid was also aspirated. The specimen was sent for histopathological examination. On gross examination, the

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mass was friable, measuring 12×11×7 cms with haemorrhagic and necrotic areas and attached lungs and soft tissues.

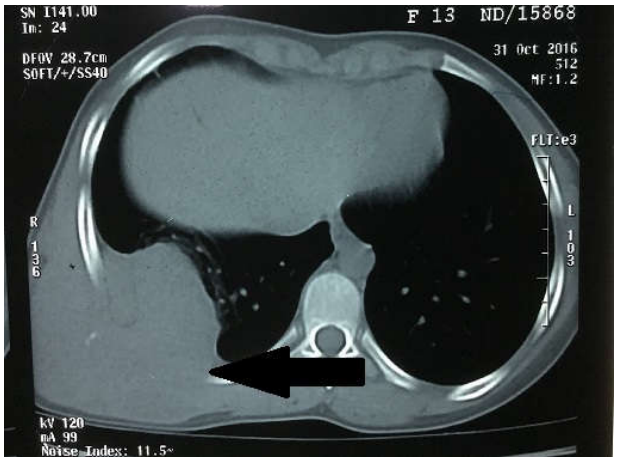

Figure 2 CT scan of the patient revealed a SOL measuring 20×20×15 cms arising from right chest wall (marked with arrow).

On microscopy, the tumour was solid comprising of uniform round cells with fine chromatin, scanty clear to eosinophilic cytoplasm and indistinct cytoplasmic membrane. Occasional small round cells with compact chromatin are also noted. Viable tumour cells are noted around blood vessels along with extensive geographical necrosis (figure 3, 4).

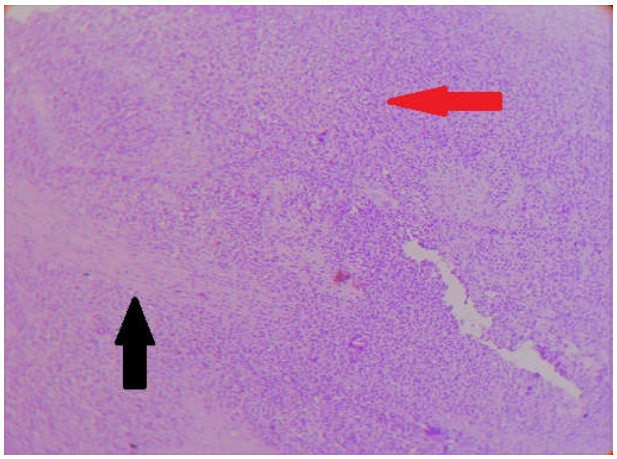

Figure 3 On histopathology, small blue round cell (marked with red arrow) with areas of necrosis (marked with black arrow) in Haematoxylin-Eosin section at 100× magnification.

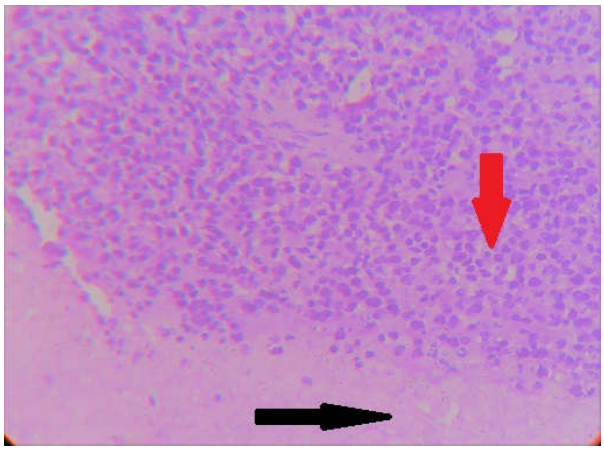

Figure 4 On histopathology, small blue round cells (marked with red arrow) and areas of necrosis (marked with black arrow) on Haematoxylin-Eosin section at 400× magnification.

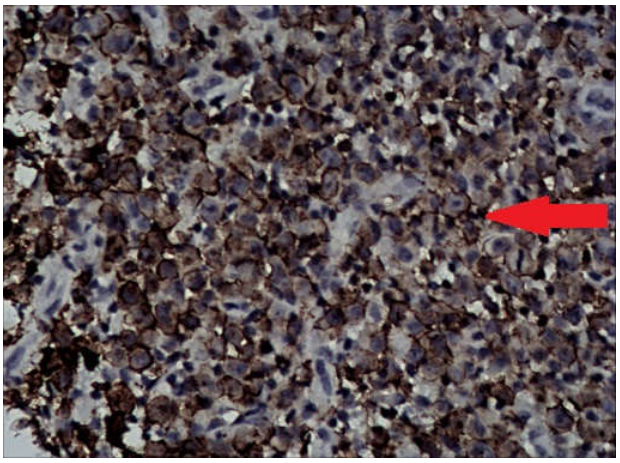

Figure 5- After immunohistochemistry, the small blue round cells showing CD99 membrane positivity (marked with red arrow) at 400× magnification.

FNCLCC grading:-

Tumour differentiation -3
Mitotic count- 1
Tumour necrosis- 2
Total- 6

Grade III (Poorly differentiated)
Immunohistochemistry for CD99 was done and the cells showed diffuse membrane positivity. (figure 5).

The diagnosis was confirmed as extra skeletal Ewing's Sarcoma.

The post-operative follow up was uneventful and she was reviewed after 2 weeks. She received radiotherapy thereafter and was doing well.

DISCUSSION

Extra skeletal Ewing's sarcoma is seen in young adults. Translocation t (11; 22) is found in the Ewing's sarcoma family tumours more so in the extra skeletal variant[6,1,2]. The gene from chromosome 22 which encodes the Ewing's sarcoma gene (EWS) and the gene from chromosome 11 (FLI-1) are involved to create the new fused gene (EWS-FLI-1) in the translocation t (11; 22). This newly formed variant encodes an altered protein which regulates other genes giving rise to Ewing's sarcoma[2]. Ewing's sarcoma has a predilection for males and commonly occurs in paravertebral region, lower limbs, chest wall, pelvis, retroperitoneal region, upper limb, head and neck[7].

Here we report a case of 13year old female patient having a mass in the upper part of back that was tender and had restricted mobility. It is very common for large, deep seated tumours to produce symptoms like these.

X ray of local parts reveals only soft tissue mass devoid of classical onion peel appearance[8]. Here X ray showed an opacity in the right lower zone. CT scans are of immense help in tumours located chest wall to detect the location and sometimes the spread. The CT scan revealed that it was a 20×20×15 cms mass arising from the chest wall. FNAC cannot reveal the detailed structural characteristics of the tumour.

Hence, histopathology still remains the mainstay of diagnosis. The histopathological examination in our patient revealed that

the tumour was composed of uniform round cells with scanty eosinophilic cytoplasm and indistinct cytoplasmic membrane with extensive necrosis. Thus having features identical to Ewing's sarcoma.

Electron microscopy reveals two types of malignant round cells[9]. One group called the chief cells which exhibit larger cell size with thin rim of pale cytoplasm, less hyperchromatic nuclei, nucleoli and diffusely dispersed chromatin nuclear details. The second group called dark cells are smaller in size, darker in colour with hyperchromatic and smudged nuclei[10]. Immunohistochemistry helps in confirming the diagnosis as was done in this case by CD99. These tumours characteristically also show vimentin positivity, S-100 negativity and intracytoplasmic glycogen[11].

Extra skeletal Ewing's sarcoma has a multimodality approach of treatment comprising of radiotherapy, chemotherapy and surgery[7,10]. The extra skeletal variety has a better overall prognosis compared to Ewing's sarcoma especially if patient's age is <16years[2]. Our patient was treated by surgery and radiotherapy and was doing well post operatively.

The differential diagnosis of Ewing's sarcoma include other small round blue cell tumours like neuroblastoma, embryonal rhabdomyosarcoma and lymphoma. Therefore, confirmation of the diagnosis of Ewing's sarcoma relies on positive staining for CD99 [12]. Here as CD99 was positive hence the diagnosis was confirmed.

CONCLUSION

Extra skeletal Ewing's sarcoma should be considered in any patient who has got a soft tissue mass in the trunk or extremities. A patient age <16 years is associated with favourable prognosis. The diagnosis can be confirmed by histopathology and immunohistochemistry. It is a curable by multimodality treatment.

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