Case Report

Orthopedic Section

Calcaneal Ewing's Sarcoma with Skip Metastases to the adjacent Tarsal Bones

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ABSTRACT

Ewing's sarcoma (ES) is a malignant non-osteogenic primary tumour of the bone, which is mainly seen in the diaphysis of the long bones and in the flat bones of the pelvic girdles in young patients. In 30% of the cases, Ewing's sarcoma is multicentric in origin and in 14-50% of the cases, multiple metastases are present at the time of diagnosis. Ewing's Sarcoma of the calcaneum has been infrequently reported in literature. Skip metastases to the adjacent tarsal bones have been reported even more rarely. We wish to report a case of Primary Ewing's sarcoma of the calcaneum with skip metastases to the adjacent tarsal bones, which was diagnosed by clinicoradiological examination and confirmed by histopathology.

Keywords: Ewing's sarcoma, Calcaneum, Skip metastases

INTRODUCTION

Ewing's sarcoma is a malignant non-osteogenic primary tumour of the bone, which is mainly seen in the diaphysis of the long bones and in the flat bones of the pelvic girdles in young patients. Originally, James Ewing described it in 1921, as a tumour arising from undifferentiated osseous mesenchymal cells; however, recent studies suggest that Ewing's tumour may be neuroectodermally derived from the primitive neural tissue [1].

All these lesions (Ewing's sarcoma and PNET) are now included in the same classification, the Ewing's sarcoma family of tumors (EFTs). The EFT, in 85% of the cases, is associated with translocation t(11;22)(q24;q12). This fusion of the EWS gene on 22q12 with the FLI1 gene on 11q24, results in a chimeric fusion transcript, EWS-FLI1. In another 10-15% of the cases, the translocation t(21;12)(22;12), resulting in the EWS-ERG (Ets-related gene) fusion, is seen. The remainder of 1-5% of the cases shows other complex translocations. The peak incidence of Ewing's sarcoma is noticed in the second decade of life, with a male preponderance of 1.6:1. It is an extremely anaplastic, round cell tumour, primarily arising in the intramedullary portion of the bone and metastases are not unusual [2].

Ewing's sarcoma demonstrates a predilection for the trunk and the long bones. In the truncal skeleton, the pelvis predominates, followed by the scapula, the vertebral column, the ribs and the clavicle. Of the long bones, the most common site is the femur, followed by the humerus, the tibia and the bones of the forearm, in that order. As opposed to osteosarcoma, Ewing's sarcoma of the long bones tends to arise from the diaphysis, rather than from the metaphysis.

In 30% of the cases, Ewing's sarcoma is multicentric in origin [3]. In 14-50% of the cases, multiple metastases are present at the time of diagnosis [4].

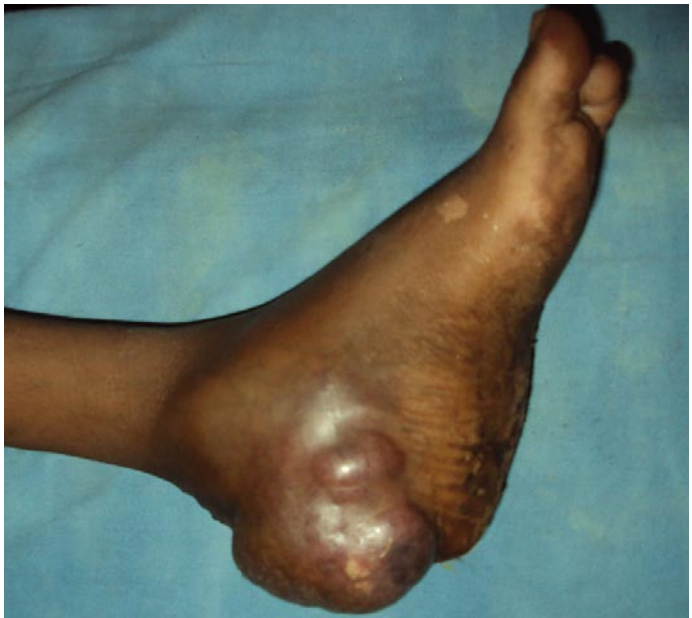
The management of Ewing's sarcoma includes multiagent neoadjuvent chemotherapy, followed by enblock excision of the tumour mass. Surgical resection improves the local control of the disease. Operative treatment may be particularly applicable in the foot [5].

Postoperative radiotherapy is given if there is doubt of tumour residua. The prognosis is poor and the tumour commonly metastasizes to the lungs and to other bones. The most unfavourable prognostic factor in Ewing's sarcoma is the presence of distant metastases at diagnosis. Other unfavourable prognostic factors include an age which is older than 10 years, a size larger than 200 ml, more central lesions (as in the pelvis or spine), and poor response to chemotherapy [6].

Ewing's sarcoma is of rare occurrence in the small bones of the hand and the feet. We wish to report a case of Primary Ewing's sarcoma of the calcaneum with skip metastasis to the adjacent tarsal bones, which was diagnosed by clinicoradiological examination and confirmed by histopathology.

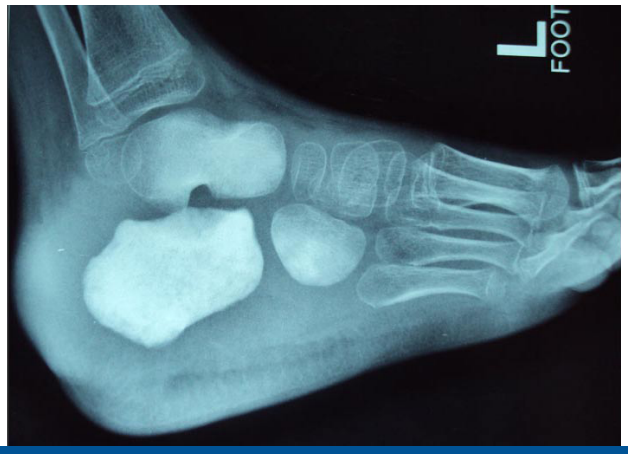
CASE REPORT

An 8-year-old boy presented with a swelling of seven months duration in the region of his left calcaneum. It started with a small painful swelling in the region, which went on to rapidly increase in size, attaining its present size at the time of admission [Table/Fig 1].



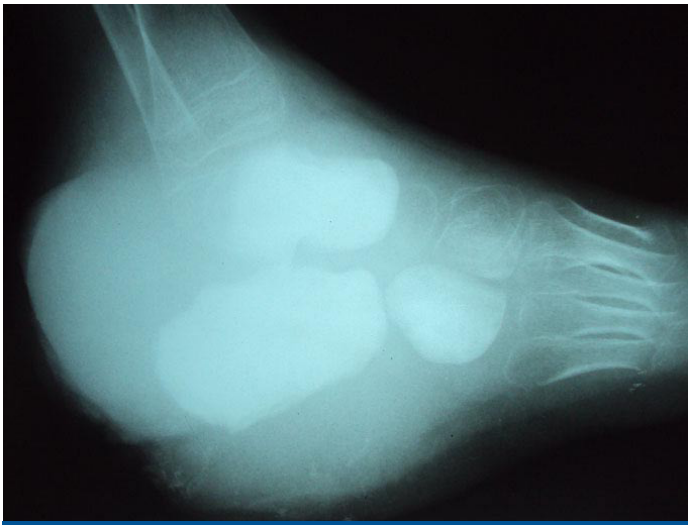
[Table/Fig 1]: Clinical photograph of patient's left heel showing huge swelling with fungating mass

On examination, there was a huge swelling present all around the heel with fungating mass. Overlying skin was stretched and shiny, with visible dilated subcutaneous veins. The rest of the skeletal survey was normal. All the blood parameters were normal, except the alkaline phosphatase level, which was found to be raised. The initial radiograph of the calcaneum, taken four months after the appearance of the swelling, showed a sclerotic lesion in the calcaneum, with soft tissue mass and minimal focal increase in the density of the talus and the cuboid [Table/Fig 2].



[Table/Fig 2]: Radiograph of patient's left heel at four months of disease showing sclerotic lesion of calcaneum with soft tissue mass and minimal focal increase in density of talus and cuboid

The initial radiograph raised doubts about the skip metastases to the adjoining tarsal bones. The radiograph of the ankle, taken at presentation (seven months after the swelling), showed the clear cut involvement of the adjacent tarsal bones – the talus and the cuboid [Table/Fig 3].



[Table/Fig 3]: Radiograph of patient's left heel at seven months of disease clearly showing sclerotic lesions in calcaneum, talus and cuboid with soft tissue mass

Fine needle aspiration cytology from the swelling was consistent with the diagnosis of Ewing's Sarcoma. CT scan of the chest was normal. The patient was advised adjuvant chemotherapy and above–the-knee amputation after explaining the prognosis of the disease, but the patient and his parents refused. Hence, the patient was managed with chemotherapy only.

DISCUSSION

Ewing's Sarcoma of the calcaneum has been infrequently reported in literature. Since 1921, Cook has reported 29 cases of Ewing's sarcoma of the calcaneum in the literature [7]. These rare cases are usually misdiagnosed, leading to treatment delay, which is detrimental to the outcome of the disease. Clinicoradiologically, Ewing's sarcomas can be misinterpreted as osteomyelitis, cartilaginous tumours, giant cell lesions, lymphomas, and osteosarcomas [8] and their differentiation often requires extensive evaluation by using different imaging modalities. The classical radiological presentation of Ewing's sarcoma is a destructive lesion in the diaphysis of the long bones, with an onionskin periosteal reaction. A universal feature of the disease is the presence of a large soft tissue mass which is relatively larger than the extent of the bone changes. Sometimes, the only apparent change in a radiograph is the presence of a mass, cortical erosion or periosteal reaction being hardly visible [9].

The radiographical features in the hand and feet involvement are generally those of typical Ewing's sarcoma: permeation, soft-tissue mass, and, frequently, coupled with a sclerotic reaction. However, with the exception of sclerosis, the features suggesting bone reaction and slow tumour growth in these patients were distinctly unusual, as compared to Ewing's sarcoma in general [10].

The lack of a lamellated or speculated periosteal reaction and the absence of cortical thickening were more commonly seen in the ES of the hands and feet than in other locations [11].

The absence of a periosteal reaction and the lack of cortical thickening were also noted in our patient. The lesions affecting the tarsal bones, more often demonstrate atypical radiographical features [8].These atypical radiographical appearances may play a role in the reported delay in the diagnosis of Ewing's sarcoma within the tarsal bones. CT or MR will optimally delineate the osseous and the soft tissue extent of the tumour, which is often much greater than may be appreciated on conventional radiographs.

In Ewing's sarcoma, the metastatic pattern may be pulmonary involvement alone, bone or bone marrow spreading alone, skips metastases, or combined metastatic disease [12],[13].

The imaging features of the local spread of Ewing's sarcoma involving the small bones into the adjacent bone, have been described in the literature very infrequently. Agarwal et al has reported calcaneal Ewing's sarcoma with metastases to the ipsilateral tibia and the fibula [14].

Shirley et al [15] reviewed 10 patients with ES of the foot: five in the calcaneus, one in the talus, two in the metatarsals and two in the phalanges. With the exception of those patients with lesions in the calcaneum, the prognosis for disease free survival was excellent. The location of the lesion is important, since in the reported cases in the literature, the lesions of the calcaneum were found to fare poorly [15].

In our patient, it was not possible to clearly determine the exact local extent of the tumour on conventional radiography in the beginning of disease. However, the initial radiograph raised doubt about the skip lesions in the talus and the cuboid [Table/Fig 2]. As the patient reported late to our institution, conventional radiography at presentation, revealed the clear cut involvement of the talus and the cuboid, along with the lesion in the calcaneum. MRI, due to its superior contrast resolution and multi-planar capabilities, is more sensitive than other imaging techniques, especially for the investigation of tumour spread to the bony structures and the bone marrow. Hence, MRI should always be performed in the analysis of Ewing's sarcoma, since it allows the accurate evaluation of the tumour extent, which is critical for its management [7],[16],[17].

According to a retrospective study concerning 235 patients with non-metastatic Ewing's sarcoma of the bone, 15 patients with a skip lesion were identified at diagnosis. But the skip lesions were located in the adjacent juxta-articular bone in only 2 cases [18], as was seen in our patient. Skip lesions in patients with otherwise non-metastatic skeletal Ewing's sarcoma, may be of the same consequence as the molecular detection of the marrow metastases and possibly confer a worse prognosis. Newer imaging modalities (for example PET) and careful staging work-up may indicate that skip metastases in Ewing's sarcoma are more common than previ-

ously suspected [19].

CONCLUSION

This case report confirms that the routine radiological management of Ewing's sarcoma should include conventional radiography, CT or MRI of the affected part to rule out skip lesions, as skip lesions are often missed out initially. CT scan of the chest should also be done to detect lung metastases. CT and MRI are essential in the determination of the true extent of the tumour. It is important to bear in mind that the most unfavourable prognostic factor in Ewing's sarcoma, is the presence of distant metastases at the time of diagnosis. Early recognition of an atypical appearance and the location of Ewing's sarcoma are necessary for its adequate treatment.

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DECLARATION ON COMPETING INTERESTS: No competing Interests

Date of Submission: **14/09/2010**
Date of Review: **27/11/2010**
Date of Acceptance: **07/11/2010**
Date of Publication: **06/02/2011**