Superficial Extraskeletal Ewing's Sarcoma of Forefoot with Inguinal Lymph Node Metastasis and Lung Metastasis: A Rare Case

Abstract

Ewing's sarcoma is the 2nd most common paediatric bone tumour but can also be seen in extraskeletal sites. Superficial extraskeletal ES is a rare entity and is rarely seen in foot. Majority of these patients are females in second decade of life, presenting with small, single mass, mobile, sometimes painful. They usually have a favourable outcome due to infrequent metastasis.

Keywords: *Ewing's sarcoma, Ewing's sarcoma family tumor, extraskeletal, superficial*

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Introduction

Ewing's sarcoma family tumor (ESFT) is the second-most common primary bone tumor in childhood after osteosarcoma[1] but is also extraskeletal (30%).[2] Extraskeletal ES is well described in deep soft tissues which is frequently found in paraspinal muscles, chest wall, and lower extremities, but in subcutaneous sites, it is rarely explained.[3] Female predominance with median age of diagnosis at 17 years has been described for superficial extraskeletal ES.[3] The superficial variant is usually less aggressive and rarely metastasizes.

Clinically, they present as a single superficial mass, 2–3 cm, mobile, sometimes painful, occurring over an average period of 5 months without a history of any trauma.[4]

Microscopic features show tumor of small, round blue cells, and cytogenetics shows undifferentiated tumor that shares common neuroectodermal precursor cell, arrested at different stages of differentiation.[5] The most common translocation seen is between EWS gene on chromosome 22 and FLI1 gene on chromosome 11 or ERG gene on chromosome 21.[6] The proto-oncogene c-myc is expressed in ESFT, whereas n-myc is not amplified.[7]

Unlike, deep counterparts of extraskeletal ES, superficial ES generally, display a favorable clinical outcome with 10-year probability of survival rate of 91%, which is due to small size, early detection, complete surgical removal, superficial location, and preventing metastatic spread.[3]

Here, we reported a rare case of superficial extraskeletal ES with inguinal lymph node involvement and lung metastasis.

Case Report

A 21-year-old male presented with complaints of swelling over the plantar aspect of the right forefoot which he noticed 2 years ago. The onset of swelling was insidious and was not painful. The swelling was of the size of peanut initially and was gradually increasing but had no discharge or bleeding. Thus, the patient landed to some local hospital after 2 years where he was investigated and biopsy was taken. The bleeding did not stop after biopsy, and hence, he came to our hospital.

The findings of general physical examination were unremarkable. Local examination revealed swelling of 5 cm × 5 cm at the plantar surface of forefoot. It was hard with irregular surface and margins but was not fixed to the underlying bone. The swelling then ulcerated which bleed on touch. The movements and pulsations of the foot and limb were normal.

X-ray foot Antero-posterior (AP), lateral, and oblique views [Figure 1] showed normal cortical outline and opacity of foot bones. On the lateral view, soft-tissue density lesion was seen in relation to metatarsal heads, no calcification or lucent areas were seen.

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Magnetic resonance imaging right foot [Figure 2] showed intensely T2 enhancing lobulated mass in the plantar aspect of the right forefoot extending up to subcutaneous tissue with minimum joint effusion right ankle. This lesion was hypointense on T1 and did not show suppression on fat saturation sequences.

Biopsy [Figure 3] shows diffuse sheets, nests, and cords of tumor cells having pleomorphism, round, oval, irregular, or elongated nuclei, hyperchromatic to coarse chromatin, variably conspicuous nucleoli, and moderate amount of eosinophilic cytoplasm. Features were suggestive of undifferentiated tumor, and immunohistochemistry (IHC) was advised.

On IHC, tumor cells were immunoreactive for vimentin, CD99, and TLT-1 and negative for cytokeratin, leukocyte common antigen, synaptophysin, and desmin. Overall features were suggestive of ES.

Positron emission tomography-computed tomography (PET-CT) [Figure 4] was done which revealed an ill-defined intensely hypermetabolic soft-tissue nodule in the plantar aspect of right foot suggestive of primary mitotic pathology. Few nonhypermetabolic to hypermetabolic pulmonary nodules were noted in the left lung field, suggesting pulmonary metastasis. Few fluorodeoxyglucose-avid right inguinal lymph nodes were noted suggestive of lymphatic metastasis.

Single fraction radiotherapy at 600cGy was given to stop the bleeding, and then, neoadjuvant chemotherapy was started with vincristine adriamycin cyclophosphamide (VAC) alternating with ifosfamide etoposide (IE) for eight cycles which was to be followed by PET-CT for response assessment.

Discussion

ESFTs are the second most common bone tumor predominantly affecting males in the second decade accounting for 2% of pediatric malignancies.[1] Extraskeletal ES can be seen in any part of the body and constitutes for 30% of ES,[2] but superficial ES is very rare. It is predominant in females and is seen at a median age of 17 years.[3] Superficial extraskeletal ES is mainly located on the lower limbs (38%), upper limbs (26%), head (20%), and trunk (16%).[3] Despite lower extremity is a relatively frequent site of superficial extraskeletal ES, very few cases of superficial ES of the foot have been reported. In a study of 14 cases of superficial extraskeletal ES, only one case was reported in the foot.[8]

Owing to its rarity and nonspecificity on histology, diagnosis is difficult and may mimic Merkel cell carcinoma, cutaneous lymphoma, clear cell sarcoma, malignant primitive neuroectodermal tumor, small cell carcinoma, rhabdomyosarcoma, and poorly differentiated adnexal tumors.

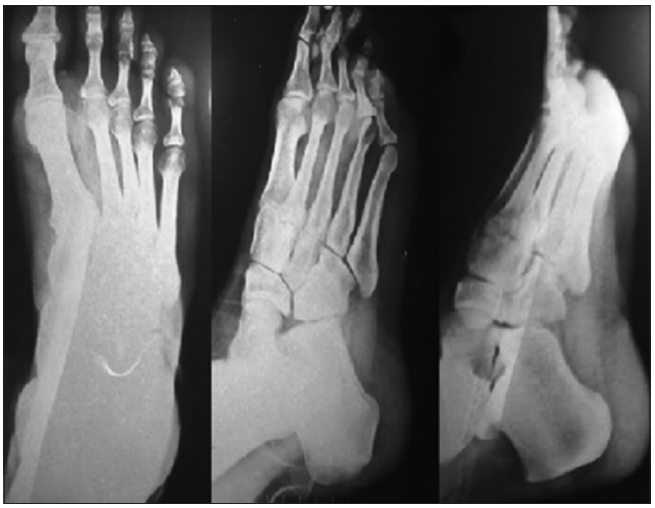

Figure 1: X-ray anterior, lateral, and oblique view

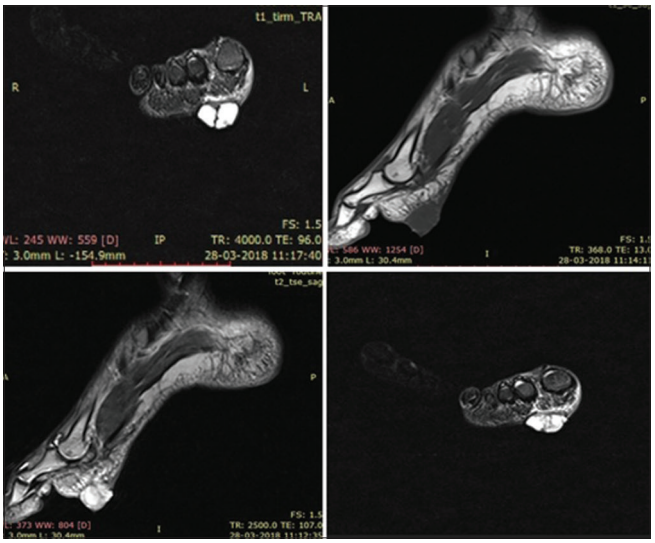

Figure 2: Magnetic resonance imaging right foot

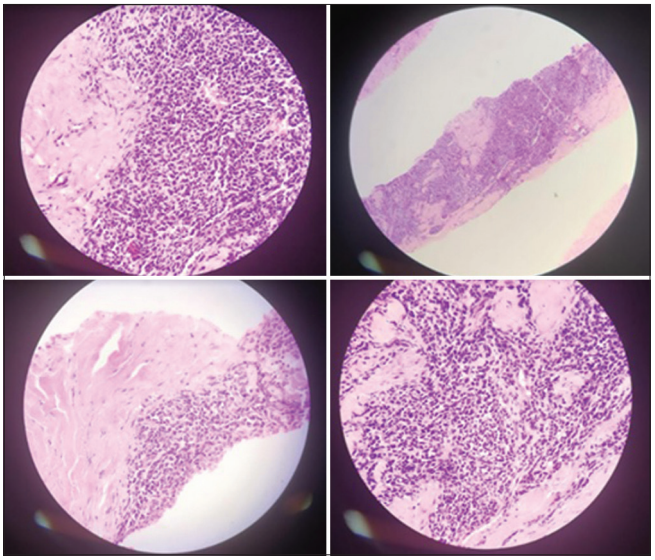

Figure 3: Histopathological images

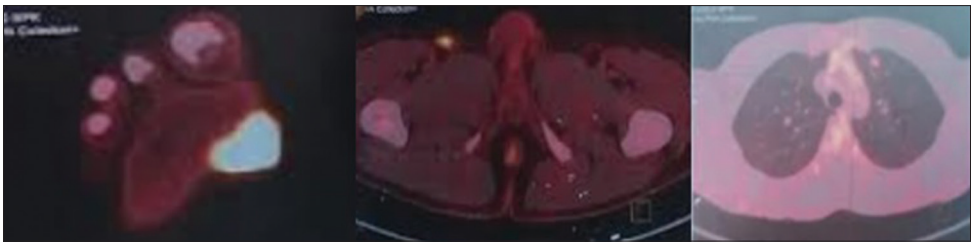

Figure 4: Positron emission tomography images

In a study, dissemination to regional lymph nodes was seen more commonly in extraskeletal primary tumors than in primary bone tumors.[2] The regional lymph node involvement with extraskeletal tumors was 12.4% and 3.2% for primary skeletal tumors.[2] The presence of metastasis is very rare, with only three cases described in the literature,[9] one with positive lymph nodes and other two with distant metastasis. The majority of these patients did not present evidence of metastasis during minimum 16-year follow-up.[9]

In our patient, in addition to rare tumor location, metastases were seen in inguinal lymph node and lungs. These rare cases can be usually misdiagnosed or lead to delay in the diagnosis. Thus, clinicians should retrieve adequate history and investigate such patients thoroughly. It should be kept in mind that superficial extraskeletal ES can occur at unusual sites and though rare, can present with metastasis also.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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