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EWING'S SARCOMA OF FOOT: A CASE REPORT AND BRIEF REVIEW OF LITERATURE

Clinical Research

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ABSTRACT

We present this case of Ewing's sarcoma of the foot in a 30 year old male, because of its unusual presentation and clinical course. The patient reported at our centre at a very advanced stage of disease. Lab investigations, X-ray and imaging studies were suggestive of a sarcomatous pathology but FNAC from the primary site as well as from distant metastasis clinched the diagnosis of small round cell sarcoma. The biopsy from the lesion in the amputated foot established the diagnosis of Ewing's sarcoma. The patient was managed by multi-disciplinary including surgery, neoadjuvant chemotherapy and radiotherapy

KEYWORDS

Ewing's Sarcoma; Malignant bone Tumor ; Foot; Metastasis.

Case

A 30 year young man presented with history of a swelling in left foot, which was rapidly increasing in size for last 8 months. To begin with, the pain was dull and continuous, relieved by analgesics. For the last two months, the pain had become severe making the patient unable to bear any weight on that limb. The patient had been on regular analgesic and antipyretics for last one month. The patient had history of intermittent low grade fever and weight loss for last two months.

The Patient had past history of a swelling developing on the medial aspect of same foot about 18 months ago. That time, the swelling was operated by a surgeon in primary health center. The wound healed by primary intention and swelling was reduced markedly. The patient received a short course of medicines and the surgical wound healed by primary intention. The patient remained apparently well for six months when the swelling reappeared on foot and started increasing in size rapidly. The patient also developed a swelling in the groin region of the same limb about 6 months ago which has also started growing in size over last 2-3 months. The treating doctor at Primary health center on X-ray of the feet suspected tumoral lesion in the foot and referred the patient to a higher center for further management. The patient reported to us after eight months of the recurrence of foot swelling.

Clinical examination at the time of presentation to our center, revealed a large swelling involving whole of the dorsum of the foot and extending into middle and anterior part of planter surface as well. All the toes including great toe were spared off the swelling. The swelling appeared warm but signs of acute inflammation were absent. The skin over the dorsal surface was scaly [Fig 1A]. Few dilated veins were present all over the swellings. Two thinned out callosities (4x5 cm size each) on planter surface were present, suggestive of patient being able to bear weight and walk some time back [Fig 1B]. On medial surface it appeared shiny and stretched out [Fig1C]. The swelling had variable consistency in different parts, ranging from firm to soft and solid to cystic. At places there were areas of hypo-pigmentation. A healed scar was present on medial aspect of foot suggestive of old surgical intervention.

The examination of the groin region of the same limb showed multiple lymph nodes masses vertically along medial side of femoral vessels as well as lymph node masses placed horizontally in sub inguinal region. Those in longitudinal plane were large sized varying in size from 5 to 8 cm. These were firm, discrete, non-tender, non-matted, and extended up to upper one third of medial part of thigh. The horizontally placed sub inguinal lymph nodes varied in size from 2 to 3 cm. and were firm to rubbery in consistency, non-tender and non-matted. Systemic examination did not reveal anything except for mild anemia. The patient was subjected to further investigations.

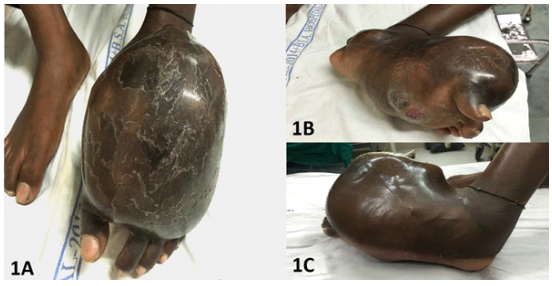

Fig 1 [1A- Skin over the dorsal surface scaly without signs of acute inflammation; 1B- Two thinned out callosities on planter surface; 1C- Dilated veins present over lateral side of swelling]

Lab Investigations: Blood investigations showed WBC count to be raised with increase on absolute neutrophil counts. The ESR was raised (95 mm/1st Hour, Wester green's Method). The Kidney function test, liver function tests were within normal limits. The alkaline phosphatase was within normal range. Serology for HIV1 and HIV2 was non-reactive. There was presence of anemia with Hemoglobin of 9.4 gm% and decreased MCV (74.9).

Radiography: X-rays showed complete destruction of 2nd, 3rd and 4th meta-tarsals. X-ray showed soft tissue mass with sclerosed ruminants of 1st and 5th metatarsals being present.2nd metatarsal was completely destroyed while only small part of destroyed 3rd and 4th metatarsal was visible. There was evidence of pathological fracture of 1st metatarsal [Fig 2A and 2B].

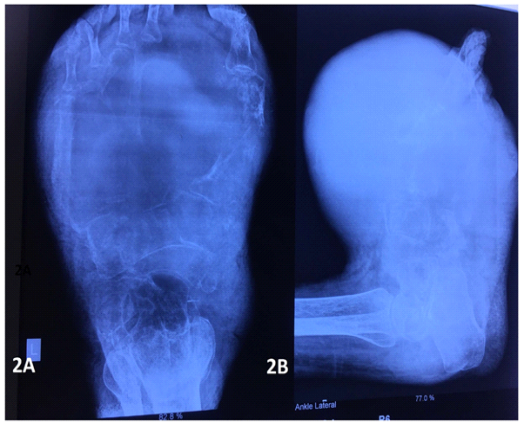

Fig 2 (2A- X-ray AP view of foot showing near complete destruction of 2nd, 3rd and 4th meta-tarsals and sclerosed ruminants of 1st and 5th metatarsals being present);(2B-Large soft tissue swelling with destruction of bones involving mid foot)

MRI: The MRI of foot showed a large multi lobulated soft tissue lesion (201mm x128mm x172mm) arising in the foot with complete loss of osseous structures viz. tarsals and metatarsals and the joints between them, muscles and tendons. Only posterior part of calcaneus and phalanges were visualized. Mostly solid, this tumor had component with heterogeneous enhancement and increased vascularity. At places, there were cystic areas with fluid level suggestive of hemorrhages. The MRI was suggestive of a large sarcomatous tumor. The contrast study showed a very large tumor with solid component (201mm X 128mm X 172mm approximately) with heterogeneous enhancement and increased vascularity. The MRI pictures were suggestive of sarcoma but no definitive opinion was possible [Fig3A].

CECT Chest and Abdomen:

There were large multiple discrete lymph nodes in the left inguinal, external iliac, obturator internus and common iliac location. Small lymph nodes at para-aortic and retrocaval location were also seen [Fig 3B]. There was a large metastatic area in supra splenic area. Bony metastases were also seen in body of L5 vertebra with pathological fracture and retropulsed posterior-superior cortex, causing mild spinal canal stenosis and inferio- posterior corner of body of D10 vertebrae. There were bony metastases in posterior superior iliac bone, right 6th rib and right 9th rib also. There were lung metastases in left lower lobe (22x17mm) and left upper lobe (10x14mm) [Fig 3C].

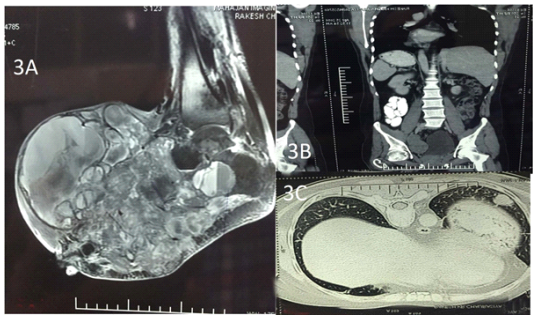

Fig3 (3A-MRI Foot showing large multi lobulated, mostly solid with component of heterogeneous enhancement and increased vascularity and at places cystic areas with fluid levels suggestive of hemorrhages. Complete loss of tarsals and metatarsals. Posterior part of calcaneus and phalanges visualized); (3B-CECT abdomen showing metastasis in supra splenic area, small lymph nodes in para-arotic and retrocaval location); (3C-lung metastasis in left lower lobe)

FNAC: FNAC from foot swelling as well as from lymph nodes from groin area showed blood mixed aspirate. The smears were cellular and comprised of small round malignant cells, suggestive of Ewing's sarcoma.

Histopathology: The biopsy samples were taken from the amputated foot. The section showed small round malignant cell tumor, consistent with the diagnosis of Ewing's Sarcoma.

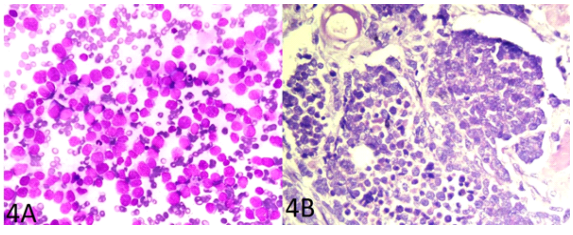

Fig 4 (4A- FNA smear showing small round cell sarcoma suggestive of Ewing's Sarcoma); (4B- Histopathology Section showing small round cell tumor consistent withdiagnosis of Ewing's sarcoma)

Follow up:

The patient was treated by multi-disciplinary approach. The patient was explained about the non-salvage stage of the foot and was operated upon to reduce tumor load by performing below knee amputation at the level of lower 1/4 of leg. Neoadjuvant chemotherapy was given, as per the European Intergroup Cooperative Ewing's Sarcoma Study (EICESS). Follow up after six cycles of chemotherapy showed significant reduction in size in the groin and abdominal lymph nodes in CECT studies. For Pulmonary lesions, additional radiotherapy was also given. The follow up at one year was uneventful. However, patient died after one and a half year of surgical amputation of the foot due to pulmonary metastasis and respiratory failure.

DISCUSSION:

Ewing's sarcoma, the second commonest primary malignant tumor of bone is usually seen in first two decades of life[1]. The ES usually arise in long bones (47%), Pelvis (26%) and chest wall (16%). Shirley et.al.[2] in their study reported Ewing's sarcoma arising in foot to be extremely rare, accounting for only 1-4 % of cases. Our patient had first surgery for foot swelling about 15 months ago, the records of which were missing. The patient remained symptom free for 6 months. He came for treatment to us after eight months of the reappearance of swelling. The late presentation and advance stage of disease in Ewing's sarcoma has been reported by many authors. M. Brotzmann et al.[3] did a study to elucidate whether a delay in diagnosis of malignant tumor of foot may reflect characteristic biological differences between Ewing's sarcomas of the foot and their counterparts at other skeletal sites. They observed a long overall delay (median of 18 months) in diagnosis of high-grade tumor like Ewing's sarcomas in foot is 2–6 times longer than the delays in diagnosis of Ewing's sarcomas located at other sites of the skeleton. These findings are consistent with other authors as well[1,4,7]. M. Brotzmann et. Al. argued that the rarity of bone tumors in foot is not the only cause of delay. It is because the symptoms such as pain and swelling are unspecific and frequently misinterpreted as being of inflammatory or posttraumatic nature. The variety of differential diagnoses explains the long delays in their diagnosis of these bone tumors. In general there was no striking difference in biological behaviour of Ewing's sarcoma in foot and elsewhere.

Most of the authors have reported that irrespective of location of the tumors, pain and swelling are the two most common presenting complaints in Ewing's sarcoma. Low grade fever is common and ESR is often raised[5]. The chief complaints in our patient also were the same. There are no specific laboratory tests for Ewing's sarcoma. However, elevated ESR, moderate anemia, leukocytosis or elevated level of LDH may be detected[1]. There was moderate anemia with decreased MCV and very high ESR in our patient.

In a study of 156 cases of Ewing's sarcoma, including three of the foot, Vohra [6] (1967) described the wide variation of the radiographic appearances and the similarity in appearance to osteomyelitis, osteogenic sarcoma, and metastatic deposits. X-rays of foot in our patient, at the time of presentation, showed destruction of bones of midfoot including metatarsals, with evidence of sclerosis in 1st and 5th metatarsals whereas all the phalanges were spared. The precise radiological diagnosis was not possible, however possibility of sarcoma was considered. Adkins et.al. [7] in their retrospective study of clinical course and results of the treatment in 16 patients (10 male and six female) who had Ewing's sarcoma of the foot from 1954 through 1992, found mean age to be 17 years (range 10-42 years). The tumor involved the metatarsals (six patients), phalanges (four), calcaneus (three), navicular (one), talus (one), and calcaneus and phalanx (one). Seven patients had metastatic disease at the time of diagnosis, and only one of these patients survived. None of the patients with pulmonary metastasis at presentation survived. Nine patients had localized disease at the time of diagnosis, and eight survived. Diagnosis was established at an average of 14 months from the onset of symptoms.

Our patient, a 30 year old male, had primary pathology in the foot. In view of advanced stage of disease at the time of presentation, it was not possible to comment on which small bone of midfoot was first to be involved. However, 2nd, 3rd, and 4th metatarsals were destroyed by tumor pathology. This was also observed in 6 of the 17 cases in the series of Adkins et.al. However, Sandrasagra[8] in his review of 433 cases of Ewing's sarcoma found involvement of metatarsal only in 2 cases.

In our patient, the MRI and contrast studies were not much helpful at precise diagnosis. However, these studies suggested an underlying sarcomatous pathology. CECT chest and abdomen showed distant metastasis in chest. There were bony metastases in lumber vertebras, iliac bone, right 6th and 9th rib. Most authors believe that radiological imaging alone is not sufficient for the diagnosis of Ewing's sarcoma[7,9]. MRI can narrow the differential diagnosis, but a specific diagnosis can rarely be established [10]. This was observed in our case as well. The diagnosis of Ewing's sarcoma should be confirmed by histopathology[7,9,10].

FNAC is of great help in diagnosis of Ewing's sarcoma. Fine-needle

aspiration of bone is a simple, reliable, and accurate diagnostic technique that can facilitate patient management and preoperative decision-making and/or avoid unnecessary invasive procedures for patients with primary or metastatic bone lesions. The overall accuracy is about 95% [11]. In our patient, the FNA smears, both from the primary site of tumor in the foot as well as from groin lymph nodes, showed small round malignant cells consistent with diagnosis of Ewing's Sarcoma. The biopsy sample from the amputated foot confirmed the diagnosis of Ewing's Sarcoma on Histopathology.

Modern multimodal therapeutic regimens for Ewing's sarcoma include a combination of chemotherapy, surgery and radio therapy[1,7]. Radiotherapy and surgery is the main the main stay of treatment for foot lesions; however control of primary lesion should be made on an individual basis[12]. In our patient, as the foot was non- salvageable, amputation at the level of lower one fourth of leg was carried out.

CONCLUSION:

Ewing's sarcoma generally involves diaphysis of long bone and axial skeleton. Involvement of foot bones is rare. In foot it is often very aggressive and diagnosis is often delayed. Patient having Ewing's sarcoma of foot at the time of presentation to the surgeon may already have local as well as distant metastasis, including involvement of lungs, bone or bone marrow, skip metastasis or a combination of all these features. Our patient had combinations of all these features. For early diagnosis of Ewing's sarcoma, FNAC is a very simple, cost effective procedure and helps in planning treatment protocol of the patient. Definitive diagnosis is by biopsy and histopathology. Further management requires multi-disciplinary approach including radiotherapy; neoadjuvant chemotherapy and limb salvage surgery wherever possible. For very advanced lesions, as was in our patient, limb salvage is often not possible and surgical amputation is performed which helps in local control of disease and early rehabilitation resulting into better quality of life.

Conflict of Interest: None.
Disclosure of grants or other funding: The study is from the Department of Orthopedics and Department of Cancer Surgery, Safdarjung Hospital, New Delhi (India). It is a Government of India owned institution and all the services are provided free of cost to the patients.

Consent: It is the policy of the hospital to take down written informed consent from the patient before carrying out any intervention procedure like FNAC, Biopsy etc. The informed consent for anaesthesia, below knee amputation and neo-adjuvant chemotherapy was also taken.

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