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Pathology

FNAC DIAGNOSIS OF EWING'S SARCOMA CALCANEUM WITH SKIP METASTASIS IN THE SMALL TARSAL BONES - A RARE CASE REPORT

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ABSTRACT Ewing's sarcoma is the second most common malignant primary bone tumor of childhood and adolescence affecting mainly the diaphysis of long bones and flat bones. This tumor is extraordinarily rare in small bones of the feet and presents as a swelling with atypical radiological features of cystic and lytic lesion with scant periosteal reaction. Moreover, skip metastasis to adjacent bones is even rarer. A rare case of Ewing's sarcoma calcaneum with skip metastasis in the small tarsal bones is being reported. Radiological lesion was not typical of Ewing's sarcoma, and a CT scan report was misinterpreted as chronic osteomyelitis. Based on radiologist's report he was treated with antibiotics for about two years without any response. He was finally diagnosed as Ewing's sarcoma on fine needle aspiration cytology.

KEYWORDS : FNAC Ewing′s sarcoma, calcaneum, skip metastasis, FNAC of bone tumors

INTRODUCTION

Ewing's sarcoma is a malignant non-osteogenic primary tumor of the bone, which is mainly seen in the diaphysis of the long bones and the flat bones of the pelvis in young patients. In 1921, James Ewing described it as a tumor arising from undifferentiated osseous mesenchymal cells; however, recent studies suggest that Ewing's tumor may be neuroectodermal in origin, derived from the primitive neural tissue.[1]The overall incidence of Ewing sarcoma has remained stable over the past 25 years . This study presents a rare case of calcaneal Ewing sarcoma as this tumor is responsible for only 3% to 5% of cases in the bones of the hands and feet.[2]

Fine needle aspiration is the simplest, most economical and reliable technique to diagnose any accessible palpable or deep seated mass in the body. Here we report a case of primary Ewing's sarcoma of the calcaneum with skip metastasis in other short bones of the right foot such as talus and adjoining metatarsal bones. The patient presented with gradually enlarging swelling in the right ankle joint and was misdiagnosed clinicoradiologically as an infective pathology. The diagnosis of Ewing's sarcoma was established on fine needle aspiration cytology and the patient was treated accordingly.

CASE REPORT:

A 26- year- old farmer from the nearby village was referred to the orthopedic outpatient department of a tertiary care hospital with a progressive swelling in the region of right ankle joint. He narrated the history of a gradually enlarging painful small swelling in the region, which started rapidly increasing in size recently (Fig 1a). It was associated with boring pain in the heel.

The X-ray of the right ankle joint presented that the cortical outline of the right calcaneum, talus and small tarsal bones were irregular and small irregular lytic areas were seen in their substances with irregular heterogeneous overlying skin tissue. Based on imaging findings radiologist's diagnosis was ? infective lesion (Fig 1b). Hence, initially the patient was managed on the lines of chronic infection i.e. chronic osteomyelitis in his native place, based on radiological diagnosis. When there was no response to the antibiotic therapy, he was referred to this tertiary care hospital.

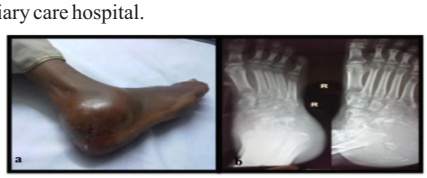

Fig 1: (a) Clinical photograph showing swelling at the ankle joint. The overlying skin is shining. Note a scar over the swelling; (b) X-ray of the ankle joint showing osteolytic lesions in calcaneum and tarsal bones with periosteal reaction.

He attended the out-patient-department (OPD) of the orthopedic surgery, from where orthopedic surgeon referred the patient to the cytopathology laboratory in the department of pathology for fine needle aspiration cytology (FNAC), before starting the treatment and to confirm the radiological diagnosis. On examination of the swelling it was observed that the overlying skin was stretched and shining. Visibly dilated subcutaneous veins were also seen. The consistency of the swelling was soft to firm on palpation. A scar mark was present on the surface of the swelling probably somebody tried to aspirate it.

CYTOPATHOLOGICAL EXAMINATION:

Fine needle aspiration was carried out with a 21 gauze needle attached to a 20 ml disposable syringe. After thorough examination of the swelling and properly cleaning the overlying skin FNAC was performed. On FNAC about 0.5 ml blood mixed thick fluid was aspirated. Multiple smears were prepared and stained by conventional H:E stain, May Grunwald Giemsa (MGG) stain and Periodic Acid Schiff's (PAS) stain for cytological examination.

Microscopic examination of the smears revealed that the smears were hyper-cellular consisting of dimorphic cell population of small and medium sized round cells (Fig 2a). There were small rounded lymphoid cells with scanty cytoplasm and little larger cells with moderate amount of cytoplasm (Fig 2b). Most of the cells were dissociated but few cords, loose clusters and pseudo-rosettes (Fig 2c) were present. The later cells were PAS positive (Fig 2d). Based on these cytomorphological features a cytodiagnosis of Ewing's sarcoma calcaneum was established.

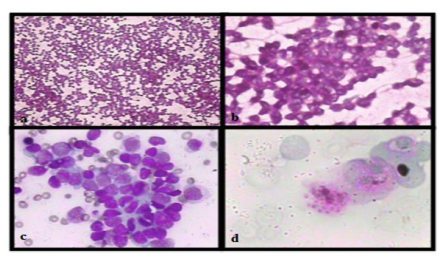

Fig 2: Microphotographs of FNAC smears: (a) Hypercellular smear consisting of small rounded cells with scant cytoplasm (MGG X 10), (b) Dimorphic cell population with an attempt to loose clustering (MGG X 40), (C) Pseudorossete formation (MGG X 80), (d) PAS positive cells (PAS X 100).

DISCUSSION:

Ewing's sarcoma is a primary malignant bone tumor usually seen in the diaphysis of long bones and rarely in the flat bones of young patients. Since 1921, Cook has reported 29 cases of Ewing's sarcoma of the calcaneum.[3] Ewing's sarcoma may involve the small bones of hands and feet and even the os calcis, though this is rare. Dahlin et al reported 165 cases of Ewing's Sarcoma, among these only four cases occurred in the feet.[4] Reinus et al in the Intergroup Ewing's Sarcoma Study (IESS) reported 12 cases of Ewing's sarcoma (ES) involving bones of hands and feet out of a total of 377 patients.[5] In the present case the age of patient was 26 years and was treated with conventional and anti-tubercular drugs because the radiological picture was not typical of Ewing's sarcoma which is described as 'onion peal appearance'. Imaging techniques were suggestive of chronic osteomyelitis due to presence of osteolytic areas with periosteal reaction in the calcaneum and adjoining small tarsal bones. In the early stages of the disease, the disease needs to be differentiated from osteomyelitis, since both may produce periosteal reaction with bone destruction. However, in the present case the diagnosis was made on fine needle aspiration only due to presence of classical cytomorphological features of Ewing's sarcoma. In a cytological study of 14 cases of Ewing's sarcoma, Dahl et al (1986) concluded that the smears have characteristic appearance and that fine-needle aspiration cytology can be used for primary diagnosis and also for chromosomal analysis to reveal the typical 11:22 translocation of Ewing's sarcoma.[6]

The cytosmears were cellular with dimorphic cell population of small round cells. These smears were distinguished from lymphoma group of tumors due the cell arrangements, besides being dissociated the cells were present in loose clusters, cords and even in pseudo-rosettes.

Where as in lymphoma all the cells are dispersed. On PAS staining the larger cells were positive for glycogen[7,8]. On immunocytochemical staining Ewing's sarcoma cells are positive for CD99, cytokeratin and MIC[2].

Shirley et al 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 calcaneus, the prognosis for disease free survival was excellent. The location of the lesion is important, since in the reported cases in the literature and in the IESS series, lesions of the calcaneum fared poorly.[9] Therefore FNAC plays a significant role in early diagnosis of Ewing's sarcoma[10].

Interesting features in the present case were-

i. Age of the patient, he was 26 year old, little older for this tumor as mentioned in the literature.[9]
ii. The site of the tumor is rare as only in 3-5% cases in bones of hand and feet have been reported in the literature. Ewing's sarcoma of calcaneum with skip metastasis in small tarsal bones is further rare.[2]
iii. Imaging picture was misinterpreted, leading in the delay in instituting the proper treatment, as has been the experience of other workers.[1]
iv. Fine needle aspiration cytology smears were characteristic of Ewing's sarcoma giving a final diagnosis of Ewing's sarcoma as already mentioned in the literature.[7]

CONCLUSION:

FNAC is a very economical, quick and an out- door procedure in the diagnosis of Ewing's sarcoma family of tumors. It is well accepted by the clinicians as well as the patients. Accurate diagnosis can be made in other bone tumors too when the adequate material is available. FNAC is also useful in long term follow up.

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