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Case Report

Pathology Section

Extraskeletal Osteosarcoma- A Case Report

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ABSTRACT

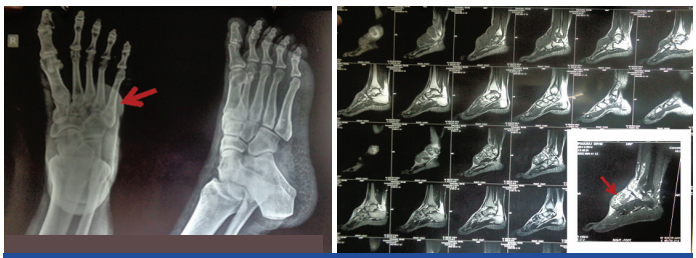
Osteosarcoma is the most common primary malignant bone tumour with a bimodal peak. But its extraskeletal variant is very rare. It arises exclusively in soft tissue without at any skeletal attachment. It accounts for 2% of all soft tissue sarcomas. It is usually diagnosed as an incidental finding during imaging. Here, we are presenting a case of extraskeletal osteosarcoma diagnosed incidentally during imaging in a 30 years old male for its rarity.

Keywords: Dorsum of foot, Malignant tumour, Soft tissue

CASE REPORT

A 30-year-old well-built male presented with a swelling in the right foot for a year. It was noticed while imaging for a trivial trauma. On examination, it was a large irregular mass measuring 8x4x3 cm involving the right foot. The mass extended from the ankle to the middle 1/3rd of the dorsum of the foot. External surface was smooth with irregular margins and with restricted mobility. The sensation was intact, no bruit over the swelling, no distal neurological deficit and no nodes were palpable.

Radiological investigations included chest X-ray which was normal. X-ray of the dorsum of the foot showed a soft tissue mass [Table/Fig-1] and MRI of the dorsum of the foot [Table/Fig-2] showed a large marginated, multi lobulated mixed intensity signal in the deep sub- cutaneous and intermuscular plane without adjacent bone and joint involvement. It was reported by the radiologists as a deeply infiltrating soft tissue mass. Suggestions were Aggressive fibromatosis and rabdomyosarcoma. Pre-operative trucut biopsy was sent. It showed pleomorphic spindle cells arranged in sheets. It was reported as spindle cell sarcoma. Following which wide local excision of the mass with reverse sural artery flap and split skin grafting was done and was sent to our department for histo-pathological examination.



[Table/Fig-1]: X-ray shows a soft tissue mass (Red arrow) involving the dorsum of the foot. **[Table/Fig-2]:** MRI shows an infiltrating soft tissue mass involving the dorsum. (Inset) MRI picture of the soft tissue mass (Red arrow)

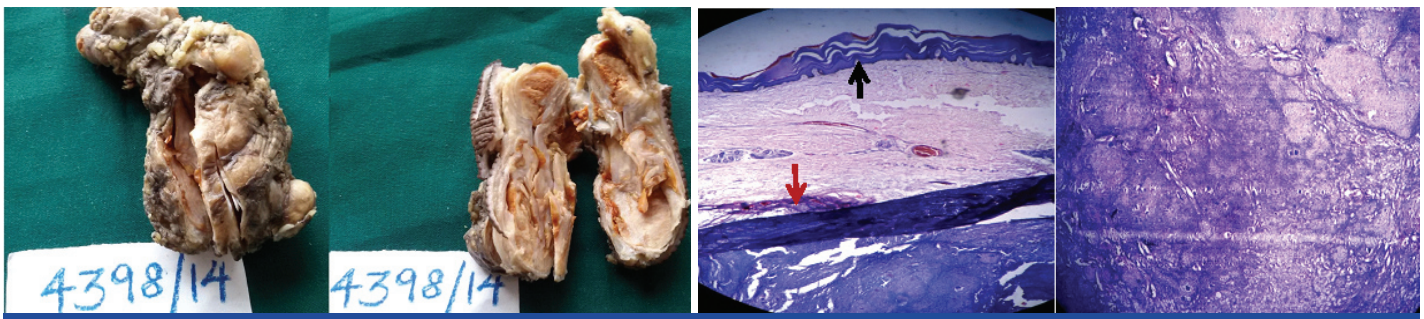
GROSS EXAMINATION

We received the specimen in the Department of Pathology. It was a skin covered soft tissue mass measuring 9x4x3 cm. Skin measures 9x2.5 cm. The external surface of the mass was nodular. Cut surface showed a firm tumour measuring 7.5x4x3 cm. The tumour had a variegated appearance with grey-white and yellow areas [Table/Fig-3].

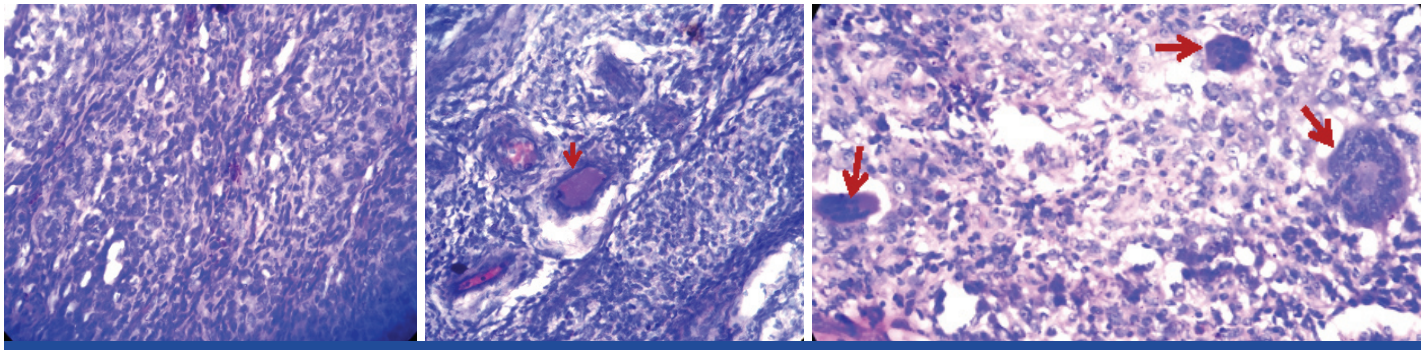
MICROSCOPY EXAMINATION

Sections studied show structure of skin and an underlying tumour tissue with cells arranged in sheets and nests [Table/Fig-4]. The tumour cells show hyperchromatic nuclei with eosinophilic cytoplasm with neoplastic osteoid formation [Table/Fig-5,6] many osteoclasts type giant cells are seen [Table/Fig-7] Circumferential and posterior surgical margins are free of tumour infiltration.

Based on the microscopic findings and in correlation with the radiological findings the diagnosis of extraskeletal osteosarcoma was made. Surgery with wide local margin is the treatment of choice. In tumours with negative surgical margins, limb salvage procedure is recommended. Even though adjuvant radiotherapy and chemotherapy are recommended, extraskeletal osteosarcoma is chemo resistant when compared with osseous osteosarcoma. Prognosis is poor with five year survival rate ranges from 10- 46%. Most of the patients die due to recurrences or distal metastasis. Our case had a tumour measuring 7cm which is slightly more than the size that is associated with better prognosis. However, he had fibroblastic morphology, which is associated with good prognosis. Long term follow-up for recurrence and metastasis was recommended for our case.



[Table/Fig-3]: Show a skin covered variegated tumour (Red arrow). **[Table/Fig-4]:** Shows structure of skin (black arrow) and an underlying tumour tissue (Red arrow) arranged in sheets and nests (H&E, 4X) Shows the tumour tissue arranged in sheets and nests (H&E 10X)



[Table/Fig-5]: Show tumour tissue arranged in sheets having eosinophilic cytoplasm and hyperchromatic nuclei (H&E, 40X). **[Table/Fig-6]:** Show tumour cells with osteoid (Red arrow) (H&E,40x). **[Table/Fig-7]:** Show tumour tissue and many osteoclasts like giant cells (Red arrows) (H&E, 40X).

DISCUSSION

Extra skeletal osteosarcoma is a rare (1-2%) malignant mesenchymal tumour of soft tissue [1-6]. They present as progressively enlarging soft tissue mass with no specific signs or symptoms. The duration of symptoms varies with an average of 6-8 months following the initial symptoms. Predisposing causes include mechanical injury [1,3] in 2/3rd of the patients and previous history of radiation [2,3] in 1/3rdof the cases. These tumours are large and deep seated [5]. The tumour can present as a well-circumcised mass or with an infiltrating margin. The cut surface of the tumour is described as grey brown with multiple foci of necrosis and calcification.

Microscopically the tumour contains neoplastic osteoid, bone and occasional cartilage and there is variation in the relative proportion of these components [1,3]. Most of the cases have the picture of undifferentiated pleomorphic sarcoma (MFH) with neoplastic osteoid. The osteoid is deposited as fine lace like or coarsely trabecular pattern. In chondroblastic variant, the tumour tissue contains atypical cartilage with focal bone formation. In giant cell variant, they contain benign and malignant multinucleated osteoclast type of giant cells. Telangiectatic variant is very rare.

Osteocalcin is more specific with 82% sensitivity and 100% specificity for osteoblasts. Focal positivity for desmin, S-100 and cytokeratin may be found.

The differential diagnosis includes benign lesion like myositis ossificans and malignant tumour with metaplastic bone formation such as synovial sarcoma and epithelioid sarcoma [7]. However, distinguishing extraskeletal osteosarcoma from pleomorphic MFH like sarcoma with metaplastic bone is very difficult. Paraosteal osteosarcoma also present as soft tissue mass with ossified matrix, but that can be differentiated because the tumour is attached to the cortex by a broad base with cortical erosion. Differential diagnosis also includes periosteal osteosarcoma and high grade surface osteosarcoma.

The prognosis for extra skeletal osteosarcoma is poor and most patients die within two to three years. Approximately 50% of tumour shows local recurrence and 65% metastasize. The most common metastatic site is the lung [4] followed by liver, bones, regional lymph nodes and soft tissue. Tumour size, site, age, histological subtype and proliferation index has been proposed as prognostic variables [6]. Tumours with more than 5 cm size has unfavorable prognosis and fibroblastic histological subtype has a favorable prognosis. Treatment is combination therapy - limb salvage surgery followed by radiotherapy and chemotherapy.

CONCLUSION

Extraskeletal osteosarcoma is a rare malignant tumour of the soft tissue. Along with histological examination, radiological correlation is necessary as continuation with the nearby bone should be ruled out before diagnosing this tumour.

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