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Case Report

A tragic end of rare variant of osteosarcoma - A case report

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ABSTRACT

Osteogenic sarcoma, a malignant mesenchymal neoplasm of long bones, has many histological variants. One such uncommon variant in which the cells have epithelioid morphology is epithelioid variant of osteosarcoma. The present case highlights the importance of knowledge for these rare variants.

1. Introduction

Osteogenic sarcoma, a malignant mesenchymal neoplasm of long bones, has many histological variants one such uncommon variant in which the malignant cells have epithelioid morphology and exhibit divergent immunohistochemical profile of epithelial and mesenchymal differentiation has been designated as epithelioid osteosarcoma. It is usually seen in older individuals and presents with resistance to chemotherapy. The varied cyto-architectural and immune-phenotypic properties of this tumor pose a diagnostic challenge on fine needle aspiration cytology, biopsies and one such case has been reported here.[1]

2. Case Report

A twenty-one-year-old male college going student presented with an atraumatic, painful swelling in the right lower thigh since four months. There were no systemic complaints. Local examinations revealed a tender swelling, measuring 10x5cms in the posterior and lateral aspect of right lower thigh with local rise in temperature. Radiological examination showed an ill-defined, expansile lytic lesion involving the epi-meta-diaphyseal region of distal one third of femur with soft tissue involvement, suggested a primary bone tumor possibly giant cell tumor. Estimation of serum alkaline phosphatase was found to be 780U/L (33-96U/L). A clinical diagnosis of giant cell tumor/aneurysmal bone cyst was made. Fine needle aspiration cytology revealed highly cellular lesion with singly scattered, clusters of plump epithelioid cells with marked nuclear pleomorphism and hyperchromasia. Tumor osteoid was not evident. A diagnosis of malignant neoplasm possibly osteosarcoma was made and biopsy confirmation was asked for. Incisional biopsy showed sheets of epithelioid tumor cells with scant questionable tumor osteoid invading bony trabeculae. Immunohistochemistry (IHC) neoplastic cells were intensely positive for cytokeratin and focally positive for S-100 and CD 30. Further CD 34, CD 31, CD 117 ALK, CD1a, MPO, EMA, desmin and CD 23 were all negative thus ruling out epithelioid variants of other sarcomas. Intense cytokeratin positivity warranted work up for exclusion for a metastatic carcinoma. Subsequent metastatic evaluation by means of CT scan revealed no other primary and only local uptake was noted on bone scan. Patient received three cycles of chemotherapy

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with doxorubicin and cisplatin followed by above knee amputation, as limb salvage surgery was not feasible due to widespread nature of tumor. Amputation specimen grossly demonstrated grey-white fleshy tumor with areas of necrosis involving lower femur. Histopathological examination revealed tumor osteoid was noted very focally and viable areas showed sheets of round to polygonal cells with abundant eosinophilic cytoplasm and highly pleomorphic hyperchromatic nuclei. Final diagnosis of epithelioid osteosarcoma of distal one third of femur with 40% tumor necrosis was made. Patient was discharged and was asked for follow-ups. After 3 years, the patient presented with tiny nodules arising from the amputation stump site. Further, fine needle aspiration cytology was done and revealed same picture of epithelioid osteosarcoma. Radiological work up revealed tumor deposits in lung, vertebrae and brain. While still the work up and further management was being planned patient deteriorated and succumbed to epithelioid variant of osteosarcoma.

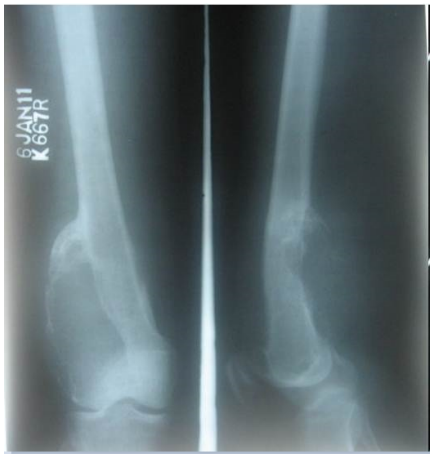

Fig. 1: X-ray showing expansile lytic lesion involving distal femur

3. Discussion

Conventional osteosarcomas are most common sarcomas of bone, constituting approximately 35% of skeletal malignancies. Classified into histological types like osteoblastic, fibroblastic, chondroblastic and others depending on the predominant matrix pattern.[1] Epithelioid osteosarcoma, subtype of osteoblastic osteosarcoma accounts for only 5.7% of all osteosarcomas, occurs in higher age groups. Our case was manifested in adolescent age. Malignant cells resemble epithelial cells, round or polyhedral rather than spindle shaped and arranged in sheets, nests or even gland like pattern mimicking metastatic adenocarcinoma. Tumor osteoid may be sparse or focal.[2] The epithelioid cells exhibit positive reactions for cytokeratin and epithelial membrane antigen but not vimentin.[3,4] Osteonectin and osteocalcin are more specific

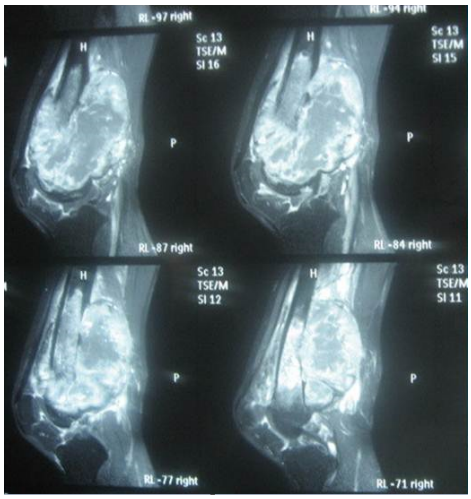

Fig. 2: CT image showing hyperintense mass with areas of necrosis

Fig. 3: Gross specimen showing grey white fleshy mass destroying the cortex and medulla of distal femur with extensive necrosis

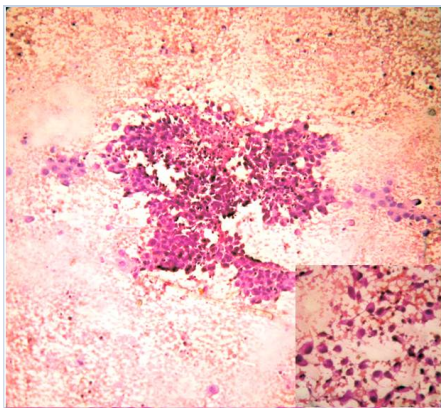

Fig. 4: FNA smears showing clusters and singly scattered tumor cells (H&E, x100) Inset showing epithelioid cells in singles (H&E, x400)

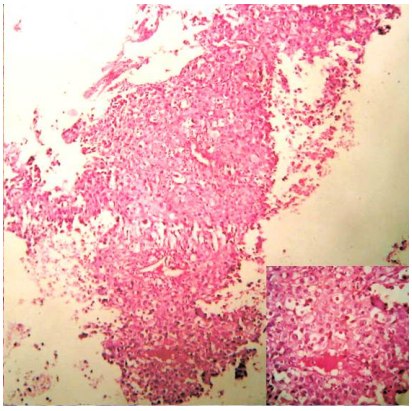

Fig. 5: Biopsy showing diffuse sheets of neoplastic epithelial appearing cells (H&E, x100) Inset showing polygonal cells with moderate eosinophilic to clear cytoplasm (H&E, x400)

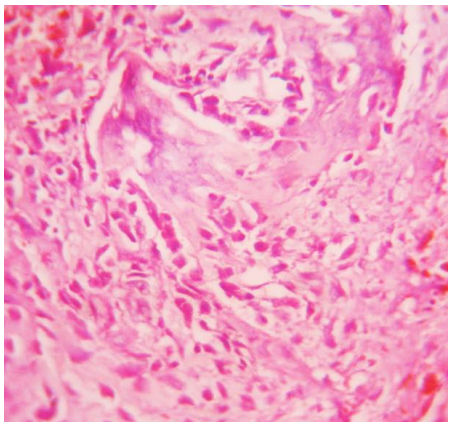

Fig. 6: Photomicrograph showing osteoid surrounded by tumor cells (H&E, X100)

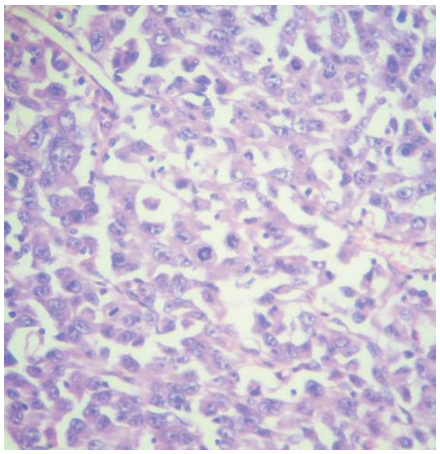

Fig. 7: Photomicrograph showing mitotically active neoplasm with cells exhibiting moderate amount of eosinophilic cytoplasm and vesicular nucleus (H&E, X400)

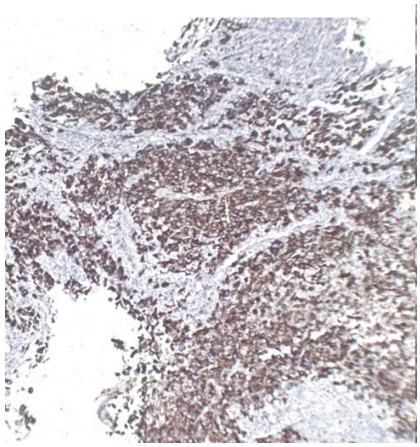

Fig. 8: Immunohistochemical stain showing tumor cells exhibiting intense positivity for cytokeratin(x100) & (x400)

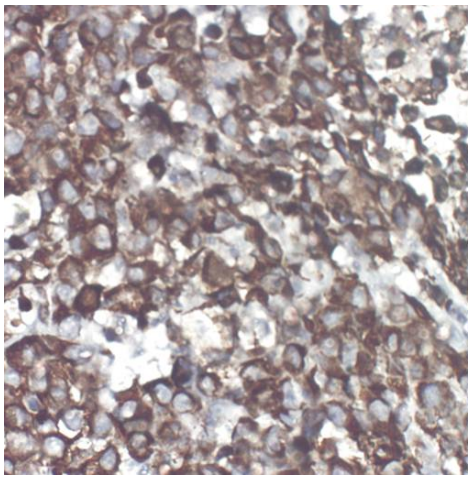

Fig. 9: Immunohistochemical stain showing tumor cells exhibiting intense positivity for cytokeratin (x100) & (x400)

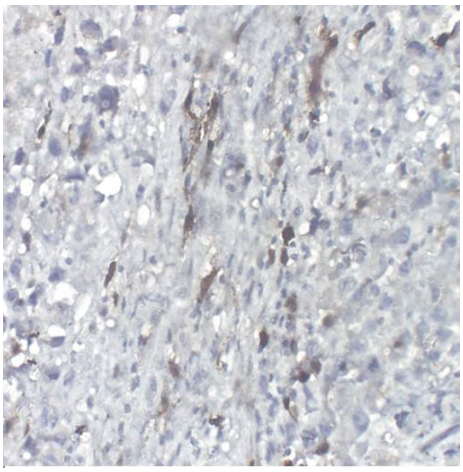

Fig. 10: Immunohistochemical stain showing focal positivity for S-100 (x100)

markers for osteoblastic differentiation. The 5-year survival rate for patients with epithelioid osteosarcoma treated with surgery with or without chemotherapy is 13.5%. As in our index case who succumbed to the entity within 3 years of diagnosis. This is in direct contrast to 75% ten-year survival rate of conventional osteosarcoma treated with surgery and chemothareapy.[1]

4. Conclusion

The present case highlights the importance of diligent and careful search for osteoid matrix. Panel of immunohistochemistry markers aids to differentiate this entity from other sarcomas and metastatic carcinomas, thus, helping in diagnosing this uncommon variant of osteosarcoma which poses an extremely poor prognosis.

5. Source of Funding

None.

6. Conflict of Interest

None.

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