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PRIMARY EWING'S SARCOMA OF THE PARANASAL SINUSES AND ORBIT

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ABSTRACT

Ewing's sarcoma is an uncommon malignancy occurring in childhood. It rarely involves maxillofacial region and particularly the orbital region in adults. We present a rare case of Ewing's sarcoma involving the maxilla together with left orbit and the complete clinical, radiographic and histological evaluation is done. The prognosis is poor due to the neoplasm's uncontrolled potential for metastatic spread, and therefore early and appropriate intervention is needed.

INTRODUCTION

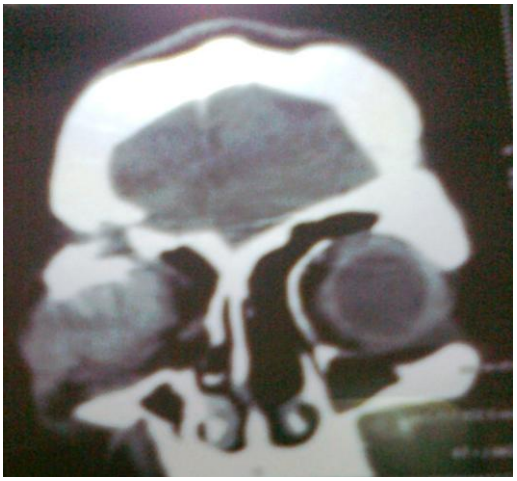
James Ewing was the first one to describe Ewing's sarcoma (ES), which was named after him in 1921. Ewing's sarcoma is a malignant small round-cell tumour typically occurring in the first and second decades of life[1] and classically involving the long bones of the limbs, the ribs, or the pelvis.[2] Less than 3% of all ES originate in the maxillofacial region, usually involving the mandible and maxilla.[3] Involvement of the orbit (ethmoidal, sphenoid sinuses, root of the orbit, lesser wing of sphenoid and temporal bone) is rare.[4]

The prognosis is poor due to the neoplasm's uncontrolled potential for metastatic spread, and therefore early and appropriate intervention is needed.

CASE REPORT

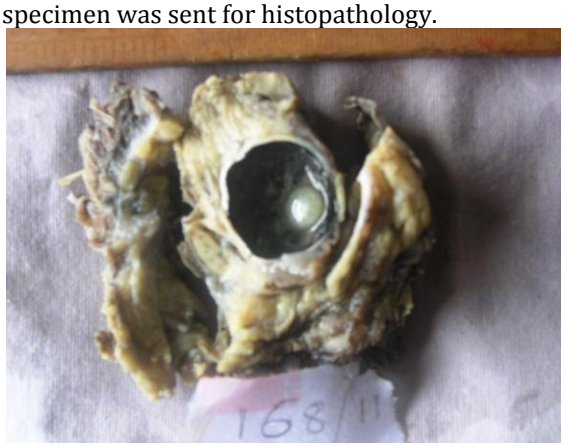
A 22 year old patient reported with a complaint of swelling and pain of 6 months duration on the left side of the face. He also experienced a progressive decrease in left nasal airway competence over the last 6 months. The general physical examination revealed no abnormality except for facial swelling which showed a gross facial asymmetry with a well circumscribed swelling measuring 8x6 cms on the left side of the face. On palpation, the swelling was tender, bony hard in consistency, nonpulsatile and non compressible. Examination of cardiovascular system, respiratory system and per abdomen showed no abnormality. Considering the patient's history and clinical observation, a malignant tumor of the maxilla was suspected, possibly osteosarcoma. A CT scan showed a destructive bone lesion involving the inferior wall of left frontal sinus and adjacent nasal bone with aggressive periosteal reaction and associated soft tissue extension.
Left nasal, ethmoidal craniofascial resection of the bone was done in 2009, followed by 5 cycles of chemotherapy.

Follow-up: 2 years later, the patient came with a history of swelling and tenderness of left lateral part of the orbit. He also complained of headache and visual loss. On ophthalmic examination, there was left axial proptosis. A CT scan showed a hyperdense soft tissue mass in the left orbit reaching upto the optical canal and inferior orbital fissure plate. Reoccurrence of the Ewing's Sarcoma was considered.



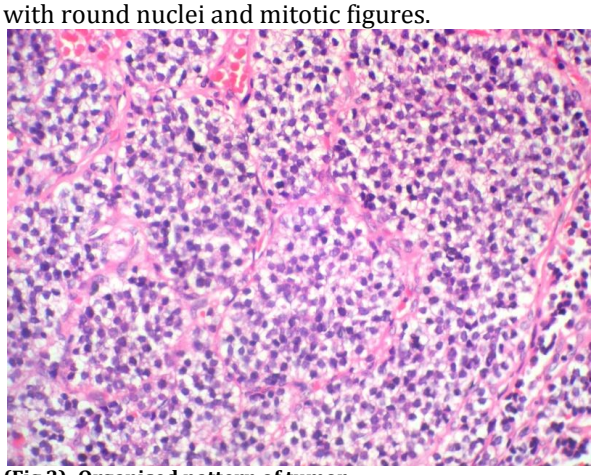
(Fig 1): Hyperdense soft tissue mass in the left orbit.

So, a combined craniofascial resection with enucleation of the left eye ball was performed. The specimen was sent for histopathology.

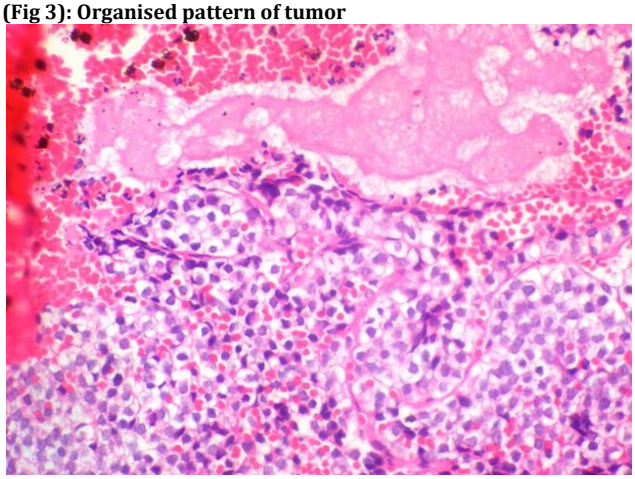


(Fig 2):Gross

Histopathology examination of both the specimens showed an organized pattern characterized by bicellular strands of tissue separated by a vascular stroma in a filigree pattern. Other areas showed solid sheets of cells divided into irregular masses by fibrous septa with pseudorossettes. Individual cells were small and uniform with round nuclei and mitotic figures.

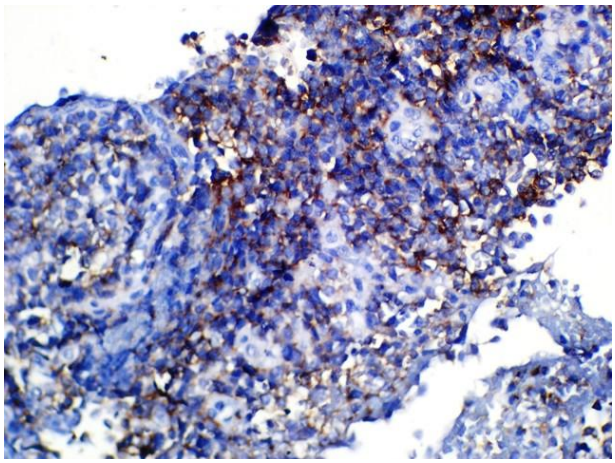


(Fig 3): Organised pattern of tumor



(Fig 4): Filigree pattern of tumor cells

PAS stain shows tumor cells positive for intracytolasmic glycogen. Immunohistochemistry shows positivity for CD99.



(Fig 5): CD 99 positivity

With the typical filigree pattern on histology, PAS positivity and CD99 positivity, a confirmed diagnosis of Ewing's Sarcoma involving the ethmoidal and nasal bone (left side) and Ewing's sarcoma of the soft tissue – orbit was given.

DISCUSSION

Ewing's sarcoma is a highly malignant, small round cell neoplasm of the bones and belongs to a family of primitive neuroectodermal tumors. 90% of the tumor occur in the first and third decades of life and males are more often affected than females (male : female = 3:2). [3] It is thought to arise from immature reticulum cells or premature mesenchymal cells of the bone marrow.[5]

The occurrence of this tumor in the paranasal sinuses with soft tissue involvement is a rare presentation. The infiltration of the tumor from the bone into the surrounding soft tissues carries a poor prognosis in keeping up with its reccurrence in the soft tissue (orbit) two years later with exostosis, headache and visual loss. Reported cases of primary Ewing's sarcoma arising in the orbital region have presented with a visible exostosis or a massive intracranial tumour or both.[6]

The common metastatic sites are lungs, pleura, bones (particularly skull), central nervous system and regional lymph nodes. [7] An extra skeletal form of this tumor has been described by Angerwall and Enzinger, which is termed as Ewing's Sarcoma of soft tissues. The ultra structural characteristics of these cells of these tumors studied by Gilipsie and his associates proved that these cells are identical to those typical of Ewing's sarcoma cells. [8] The prognosis of Ewing's Sarcoma of soft tissue is poor at the time of diagnosis. It is hard to define whether the present case took origin in the bone or soft tissue, though the likelihood of arising from the bone is more plausible.

Radiographically, the neoplasm is most often seen as a destructive, expansile, mottled radiolucent lesion which may produce a laminated periosteal reaction.[10] Some authors have stated that this pattern denoting an "onion skinning" is exceedingly rare in head and neck lesions. [7]

Both ES/PNET are no longer considered a distinct entity because of histological and molecular similarities. More than 90% show a clear translocation t (11;22) (q24;q12) resulting from the fusion of EWS and FLI gene. The gene rearrangement causes a fusion product which function as an oncogene aberrant transcription factor with structural variability and potential prognostic impact.[11] The neuroepithelium differentiation is varied and Ewing's sarcoma shows a minimum degree of differentiation and PNET shows obvious degree of differentiation. [7]

A filigree microscopic presentation is said to represent an unfavourable prognostic indicator.[7] Histopathologically the tumor must be differentiated from small round cell tumor, indicating small cell osteosarcoma and malignant neoplasms like mesenchymal chondrosarcoma, small cell osteosarcoma, rhabdomyosarcoma, malignant lymphoma, eosinophilic granuloma, neuro-endocrine tumors and metastatic neuroblastoma. The presence of CD 99 positivity confirmed the diagnosis of Ewing's sarcoma.

CONCLUSION

Ewing's Sarcoma is a rare malignancy that may affect facial bones and soft tissues of orbit of young individuals. Even after multiple therapy modalities, the present case showed a reccurrence after two years. Hence, extensive treatment and follow up is mandatory. An evaluation of the lesion should be carried out via radiography, CT scan, biopsy, special stains and immunohistochemistry.

This case is reported here for its rarity of its presentation in the facial bones.

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