

## CASE REPORT

**A Case of Ewings Sarcoma involving Mandible, an Unusual Site****Rishi Diwan, Manu Mathur, Devendra Mathur**

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**ABSTRACT**

Ewings sarcoma /PNET are defined as round cell sarcomas that shows varying degree of neuroectodermal differentiation. ES/PNET is the second most common primary malignant tumour of bone in childhood and adolescent. The median age for patient with Ewings family of tumour is 15 years and more than 50% of the patients are adolescents. Primary malignant tumours of jaws and ES of the mandible is rare. (though more common than the maxillary involvement. Present case was mistaken as dental inflammation and resulted in delayed diagnosis.

**Key Words:** Ewings Sarcoma; Mandible**INTRODUCTION**

ES/PNET is relatively uncommon accounting for 6-8% of primary malignant bone tumours.<sup>[1]</sup> It is second most common sarcoma in bone & soft tissue in children.<sup>[2]</sup> The median age for patient with Ewings family of tumour is 15 years and more than 50% of the patients are adolescents.<sup>[3]</sup>

ES/PNET tends to arise in diaphysis, or metaphyseal-diaphyseal portion of long bone, pelvis and ribs are common locations. The skull, vertebrae, scapula & short tubular bone of hands & feet are rarely involved. <sup>[1]</sup> Less than 3% of all ES originates in the maxillofacial region usually involving the mandible.<sup>[4,5]</sup> Because of the rarity of tumour involving the mandible and misdiagnosed for long time as dental inflammation /cyst the present case is being reported because of the unusual site.

**CASE REPORT**

A 15 year old girl visited the department of Dentistry with complaints of pain & swelling in

the left side of face. The case was previously attended outside the institute and was treated for long time for dental infection. There was history of swelling & pain in left jaw for 3-4 months. Intraoral examination reveals swelling involving the left mandible.

OPG finding –mandible shows irregular lytic bone destruction with ill-defined margins. The routine hematological investigations (CBC) were within normal limits except for raised ESR. On histological examination –H & E stained sections showed sheets of uniform small round cells arranged in diffuse pattern with scanty indistinct outline, little cytoplasm & round to oval nuclei. Diagnosis of small round cell tumour with possibility of Ewings sarcoma/PNET was given. The blocks were sent for Immunohistochemical studies showed PAS positivity and CD99 positive. Other markers were also done to rule out other different round cell tumour like malignant lymphoma, metastatic neuroblastoma, small cell osteosarcoma and embryonal rhabdomyosarcoma.

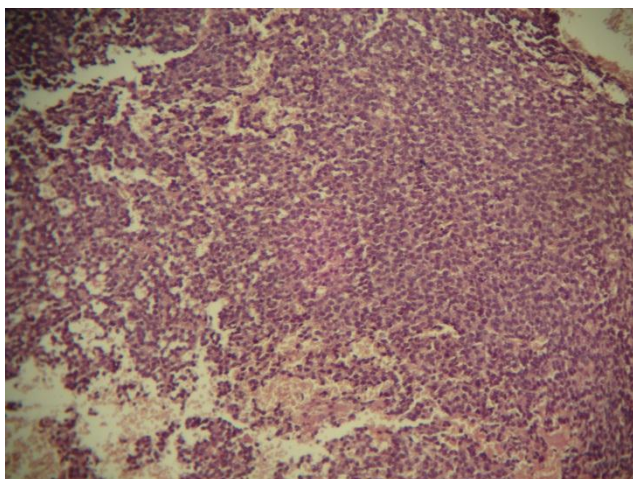
After marker studies the diagnosis of Ewings sarcoma was confirmed. A picture of small

round cell tumour with PAS positivity and CD 99 was helpful in finalizing the diagnosis.

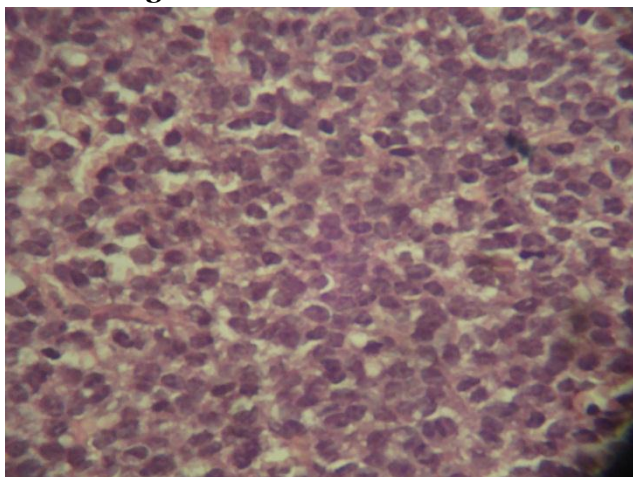
**Figure-1: OPG: Right Mandible shows Irregular Lytic Bone Destruction with Ill-defined Margins**



**Figure-2: H & E Section shows Sheets of Small Round Cells in Low Power**



**Figure-3: H& E Section shows Small Round Cells in High Power**



## DISCUSSION

Ewings sarcoma is rare in the head and neck region more so in the mandibular area. This case presented with swelling and pain of the left sided jaw and was mistaken and treated for dental inflammation. X ray finding with biopsy helped in diagnosis of the case. This case is presented to emphasize the importance of biopsy in case of jaw swelling as Ewings sarcoma which are very aggressive tumour can rarely present like this so histopathological evaluation should be mandatory.

## CONCLUSION

After marker studies the diagnosis of Ewing's Sarcoma was confirmed. A picture of small round cell with PAS positivity and CD99 was helpful in finalizing the diagnosis. The present case emphasizes the importance of OPG and Biopsy in any persistent jaw swelling as ES a rare aggressive tumour can present like this clinical picture.

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