

## Case Report

### A Rare Case of Ewing's Sarcoma affecting Mandible of a Child

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#### ABSTRACT :

##### Summary:

Ewing's Sarcoma of the body of the mandible is rare and can be mistaken as inflammation of dental origin. Here we presented a case of Ewing's Sarcoma of a female child of 2 years and 3 months old. After histological and immunohistochemical confirmation, patient was treated by VAC and IE therapy alternatively. The reported case describes the clinical, radiological and histopathological findings of Oral Ewing's Sarcoma with difficulties in management with initial response of treatment.

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#### Introduction:

Ewing's Sarcoma (ES) of jaw bone is rare & member of Primitive Neuro Ectodermal Tumor Group<sup>1</sup>. Usually it affects the long bone & in the maxillofacial region mandible is the longest bone, it involves the mandible mostly. Less than 3% of all ES originate in the maxillofacial region, usually involving the mandible<sup>1,2</sup>. 90% occur in the first three decades of life and males are more often affected than females with the ratio of 3:2<sup>1,3</sup>.

Clinically, patients are usually young adults. Typical symptoms are bone swelling & often pain, progressing over a period of months. Teeth may loosen, tooth germs are absent,

and overlying mucosa ulcerates. Clinical symptoms such as swelling, pain and sensory disturbances are rather non-specific and can sometimes be misleading<sup>3</sup>. Fever, leukocytosis, a raised ESR & anemia may be associated and indicators of poor prognosis<sup>4</sup>.

The case report presents the clinical, radiological & histopathological findings of a case of Ewing's Sarcoma in a 2 years and 3 months old female patient.

#### Case Report:

A 2- year 3-month old female child presented with rapidly progressive swelling of her left lower jaw to the department of Oral & Maxillofacial Surgery, Dhaka Dental College and Hospital. Patient's attendant also complained of occasional bleeding from the swelling in the oral cavity without any history of fever, weakness or change of body weight. Patient was in good general health with 10 kg weight and 0.47 m<sup>2</sup> body surface area and normal body temperature during

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admission. Examination of patient revealed diffuse extra-oral swelling on left side of mandible with shiny, thin skin & ulceration with color change in overlying mucosa. The swelling was ill-circumscribed, measuring 7cmX7cm in diameter with slight tenderness and soft in consistency. Teeth were absent related to the lesion. There was no other swelling or lymphadenopathy were detected in other parts of body. Liver and spleen were normal in clinical examination. X ray skull with mandible P/A and lateral views showed ill-defined osteolysis areas with overlying soft tissue mass shadow in left hemi mandible. The lesion caused displacement of a partially formed tooth in the vicinity. Investigations showed reduced hemoglobin (8.2gm/dl), raised ESR (140mm in first hour), elevated WBC ( $15.2 \times 10^3/\text{ul}$ ) with normal other values.

Incision biopsy revealed presence of round cells arranged in sheets. The cells had hyperchromatic nuclei with scanty cytoplasm. The appearance was that of a malignant round cell tumor, consistent with Ewing's sarcoma/PNET. In immunohistochemical examination, CD99 marker was positive but other panel of markers e.g. CD3, CD20, CHR, MPO, desmin, vimentin were seen considering the expense. The tumor board decided to give alternate VAC (Vincristin, Adriamycin/Doxorubicin, and Cyclophosphamide) and IE (I-Phosphamide, Etoposide) therapy 2 weeks alternatively with total of 14 cycles followed by surgery if needed. Accordingly initial cycle was given with VAC following which patient developed leukopenia. Then she was treated by Granulocyte Stimulating Factor (Filgrastin 30mg subcutaneously daily for consecutive 5 days. After the initial therapy the lesion was disappeared with a small residual swelling in submental region. After 4 months follow up she is feeling well with normal oral function and good appearance. Patient will receive chemotherapy according to the schedule and will be in regular follow up.

### Discussion:

Ewing's sarcoma is a malignant small round cell tumor of primitive neuro ectodermal tumor group and accounts for around 5% all bone

malignancy<sup>2</sup>. It shows a characteristic translocation t(11:22) (q24;q12) resulting in the fusion of the EWS and FLI-1 genes. This gene rearrangement causes a fusion product which functions as an oncogenic aberrant transcription factor with structural variability<sup>5</sup>. Adolescents and young adults are mostly affected and is rarely seen before the age of 5 years but the reported case is 2 years 3 months old. The tumor is aggressive in nature and characterized by rapid growth and high probability of early micrometastasis<sup>6</sup>. ES arising from the bones of the head and neck region is exceedingly uncommon<sup>6</sup>. When it occurs in the jaw, mandible is more frequently affected than the maxilla. Swelling, pain, increased ESR, leukocytosis & increased temperature are some signs of ES, but it also mimick to other odontogenic infections<sup>3</sup>. In this case, bleeding, displacement of teeth from vicinity & absence of tooth germs, bone loss were suspicious for malignant lesion as well as infection. It is usually diagnosed by histology and immunohistochemistry. A series of marker like CD99, CD3, CD20, CHR, MPO, Vimentin, and Desmin are usually tested to diagnose such case and simultaneously to exclude other small round cell tumors<sup>7</sup>. The reported case was positive for CD99, but other markers were not seen considering the financial burden to patient. Ewing's sarcoma is usually sensitive to chemotherapy and radiotherapy<sup>8</sup>. Modern treatments are based on combined modality of treatment with local therapy by surgery and/or radiotherapy to the main tumor followed by systemic therapy with chemotherapy for management of micrometastasis<sup>9</sup>. Our case responded very well with initial dose of VAC chemotherapy, though management was complicated by development of leucopenia which was immediately treated by administration of granulocyte stimulating factor.

### Conclusion:

Oral Ewing's Sarcoma is a rare tumor in oral cavity which can mimic odontogenic inflammatory lesion or infection. For such an aggressive large lesion, early biopsy is recommended. Any surgery may risk the patient if pre-operative chemotherapy is not

administered. Long term follow up is necessary to see the outcome of the lesion.

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