Journal of Population Therapeutics & Clinical Pharmacology

RESEARCH ARTICLE
DOI: 10.53555/jptcp.v29i03.6333

EWINGS SARCOMA OF MAXILLA : FEW TO FAR

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Abstract :
Ewing's sarcoma is a rare but aggressive form of bone cancer that primarily affects children and young adults. It typically develops in the long bones of the body, such as the arms and legs, but it can also occur in other bones, including the skull. When Ewing's sarcoma develops in the maxilla, which is the upper jawbone, it poses unique challenges for patients and healthcare professionals alike. In this article, we will delve into the intricacies of Ewing's sarcoma in the maxilla, exploring its causes, symptoms, diagnosis, treatment, and prognosis.

Introduction :
Ewing's sarcoma is a highly malignant bone cancer primarily affecting Caucasian children and young adults, with rare occurrences in the maxilla (1) . It involves genetic rearrangements leading to the EWS-FLI1 fusion protein. (2) Symptoms in maxillary cases include facial swelling, pain, and

difficulty in oral functions. Diagnosis involves imaging and biopsy, revealing characteristic cellular features. Treatment includes surgery, chemotherapy, and radiotherapy, but the maxilla's complexity makes complete removal challenging. Prognosis is generally poorer due to these challenges.

Case report

A 23-year-old male presented to the oral and maxillofacial pathology department with a chief complaint of swelling on the upper aspect of his face persisting for five months. Initially mild, the swelling responded to medication. He also reported a history of nasal bleeding three months prior and mobility of the upper left posterior tooth.

Upon examination, a diffuse, firm swelling measuring approximately 3x3 cm was noted. It extended from the infraorbital margin to the maxillary alveolar region and from the ala of the nose to the preauricular region, involving the upper lip.(fig 1) Additionally, it extended superiorly to 1 cm above the angle of the mouth and laterally to 4 cm in front of the tragus. The overlying skin appeared mildly erythematous, and the swelling encroached upon the lower lid. Intraorally, obliteration of the buccal vestibule was observed, with tooth #26 absent. (fig 2)

Radiographically, an OPG reveals left maxillary sinus radiopacity with nasal septum deviation, while the maxillary sinus floor and posterior border remain intact. A CT scan shows right-sided nasal septum deviation with a bone spur and homogeneous thickening in the left maxillary region.(fig 3 , 4)

Microscopically : H&E stained section shows, background of uniformly spreaded monomorphic round, ovoid and few elongated cells with eosinophilic cytoplasm and prominent nuclei. The cellular aggregates are interspersed with muscle tissues, eosinophilic sinusoidal spaces, vascular cells and neural elements.Histopathologically were suggestive of Round cell tumor (fig 5 a,b, c)

Immunohistochemistry : Nuclear staining is seen in sheets of monomorphic cells with eosinophilic cytoplasm and prominent nucleoli, positive for CD99, WT-1, CK (focal), FLI-1, ERG, and weak scattered CD31. They are negative for Desmin, CD45, P40, SATB2, TLE-1, cyclind1, BCOR, NKX2.2, HHV8, D240, MYOD1. KI67 is at 50%.(fig 6 a , b)

Discussions :

Etiologpathogenesis :

Ewing's sarcoma in the oral cavity involving multiple factors such as it features a chromosomal translocation between the EWSR1 gene on chromosome 22 and the FLI1 gene on chromosome 11. Potential contributing factors include exposure to ionizing radiation, environmental chemicals, toxins, and developmental abnormalities, but the exact triggers are unknown. (4,5)

Clinical feature :

Patients with Ewing's sarcoma in the maxilla may experience persistent jaw pain, swelling, facial asymmetry, changes in tooth alignment, and a firm lump or ulcerated lesion in the mouth. The tumor can also cause difficulty speaking or breathing and may lead to neurological symptoms like numbness, tingling, or weakness in the face or mouth.(6,7)

Radiological features :

Several radiological modalities may be used to evaluate oral lesions suspected to be Ewing's sarcoma, including plain radiography including OPG , computed tomography (CT) .(8,9)

Histopathological features :

The hallmark histological feature of Ewing's sarcoma, is the presence of small round blue cells. These cells are uniform in size and shape, with scant cytoplasm and hyperchromatic nuclei. The densely packed arrangement of these cells gives the tumor a characteristic "blue appearance.Homer-Wright rosettes, characterized by a central core of tumor cells surrounded by radiating neuropil-like fibrils, may be observed . It exhibits high mitotic activity, with frequent mitotic figures .Areas of necrosis

may be present within the tumor,Infiltration of tumor cells into blood vessels may be observed histologically. This feature is associated with the potential for hematogenous spread and metastasis to distant organs. It This can manifest as periosteal elevation, new bone formation, or a characteristic "onion skin" appearance.(10,11,12)

Observations :
In the span of 10 years records of institution shows total of 7 cases of Ewings Sarcoma were reported , which evidentally shows the rare occurrence of this neoplasm . (13 , 14, 15)

Immunohistochemistry :
Immunohistochemical Staining: Immunohistochemical staining may be performed to aid in the diagnosis of oral Ewing's sarcoma. Tumor cells typically express markers such as CD99 (MIC2), FLI1, and neuron-specific enolase (NSE), which can help differentiate Ewing's sarcoma from other small round blue cell tumors. (17)

Genetic testing :
Molecular techniques, such as fluorescence in situ hybridization (FISH) or reverse transcription-polymerase chain reaction (RT-PCR), may be used to detect the characteristic EWSR1-ETS gene fusion, providing additional support for the diagnosis of Ewing's sarcoma.(18)

Prognosis :
Oral Ewing's sarcoma prognosis depends on stage: early-stage tumors in the oral cavity have better outcomes than advanced ones with local invasion or distant spread. Distant metastases, especially to the lungs, worsen prognosis significantly.(19)

Treatment :
The treatment of oral Ewing's sarcoma typically involves a multidisciplinary approach, incorporating surgery, chemotherapy, and radiation therapy. (20)

Conclusion :
In conclusion, Ewing's sarcoma in the maxilla is a rare and aggressive form of cancer that presents unique challenges for diagnosis and treatment. Prompt recognition and appropriate management are crucial for improving patient outcomes. Multidisciplinary collaboration between oncologists, oral and maxillofacial surgeons, radiologists, and pathologists is essential in delivering optimal care to individuals with this devastating disease. multidisciplinary approach are vital for improving outcomes and enhancing the quality of life for patients affected by this condition. Ongoing research and advancements in medical science continue to provide hope for better treatment options and improved long-term survival rates for individuals battling Ewing's sarcoma in the maxilla.

Fig 1 . Diffuse firm swelling on left side of the cheek and infra orbital

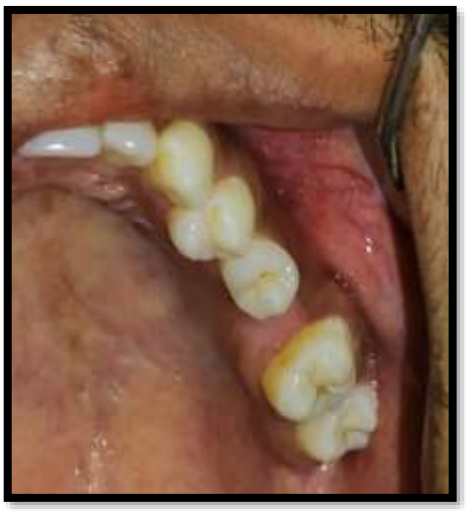

Fig 2 intraorally shows obliteration of the buccal vestibule with missing 26

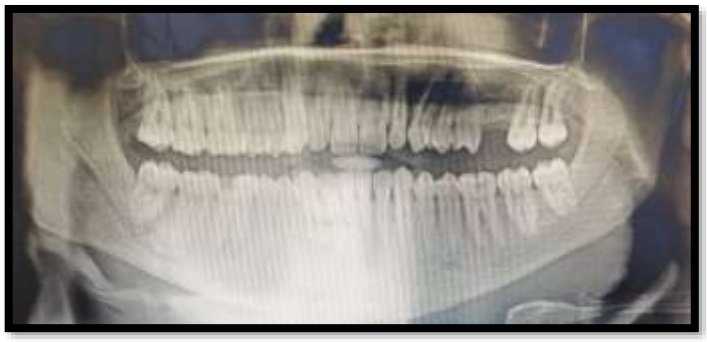

Fig 3 . OPG reveals haziness on the left side of the maxillary sinus with deviation of the nasal septum , maxillary sinus floor and the posterior border appears to be intact.

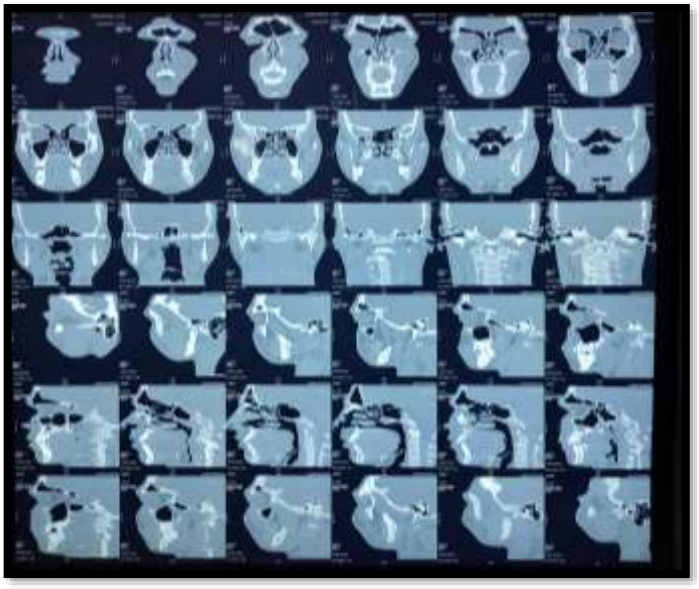

Fig 4. Coronal section – shows mucosal thickening in the maxillary sinus with mild deviation of the nasal septum .

Sagittal section – shows mucosal thickening in the floor of maxillary sinus extending to the sinus cavity with involvement of the ethmoidal sinus .

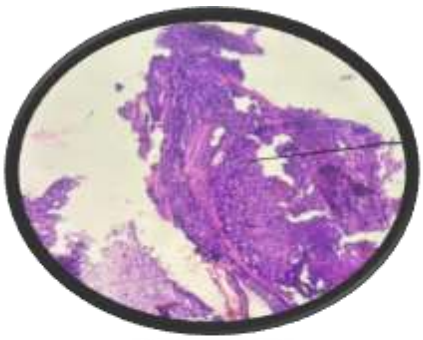

Fig 5 a. shows 4x view with dense connective tissue component incorporated with areas of round cell

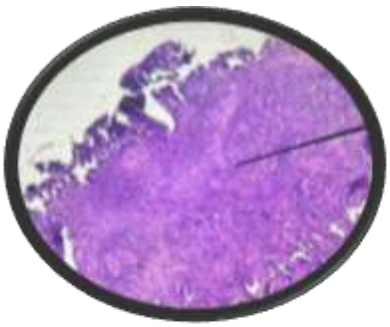

Fig 5 b shows 10x view background of uniformly spreaded monomorphic round, ovoid and few elongated cells with eosinophilic cytoplasm and prominent nuclei. The cellular aggregates are interspersed with muscle tissues, eosinophilic sinusoidal spaces, vascular cells and neural elements.

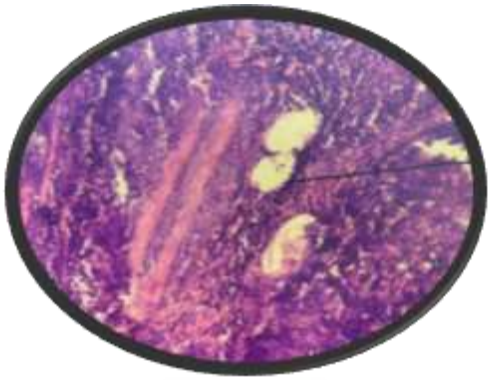

fig 5 c shows round cell morphology

Fig 6 a,bThe tumor cells are positive for CD99, WT-1 , CK (focal), FLI-1, ERG, CD31(Weak scattered)

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