

## Case Report

# Ewing's sarcoma of the mandibular condyle in a young adult: a rare case report

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**Abstract – Introduction:** Ewing's sarcoma is a malignant, small, round, blue-cell tumour with metastatic potential that usually occurs in older children and young adults. Its occurrence in the jawbones is exceedingly rare and even rarer in the mandibular condyle. This case is being reported due to the extreme unique imaging finding of Ewing's sarcoma arising in the mandible in a young male. **Observation:** A 22-year-old healthy male presented with a complaint of a progressive swelling in his right preauricular region. Magnetic resonance imaging showed a hyperintense lesion with the right ramus with loss of fat planes. True cut biopsy and histopathological examination of the lesion was suggestive of Ewing's Sarcoma. Immunohistochemical analysis confirmed the diagnosis. **Discussion:** The most common presenting symptom of Ewing's sarcoma in the head and neck region is a rapidly growing expansile mass along with pain, paraesthesia, and visual disturbances often mimicking odontogenic infections. Higher imaging modalities play a major role in the evaluation of the extent of the lesion and diagnosis. **Conclusion:** Prompt and pertinent treatment planning is critical for appropriate and timely management.

## Introduction

Ewing's sarcoma (ES) is a lethal round cell sarcoma with unclear etiology and is known to affect long bones. Its occurrence in the jawbones is exceedingly rare and condylar involvement is rarer. It is a malignant neoplasm with an aggressive growth pattern and a tendency for metastasis [1,2]. Here, we report a case of Ewing's sarcoma of the right mandibular condyle with extensive destruction of the ramus in a 22-year old male which adds to the existing body of literature on this rare tumour in the jaws.

## Case report

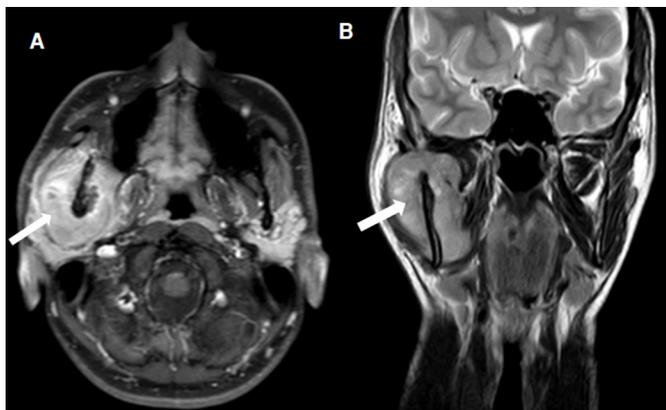
A 22-year-old otherwise healthy male presented to the Department of Oral and Maxillofacial surgery with a complaint of a progressive swelling in his right preauricular region of a

3-week duration. The swelling was non-painful and associated with fever. On extraoral examination, there was 5x1.5cm diffuse swelling on the right side with mild ear lobe elevation.

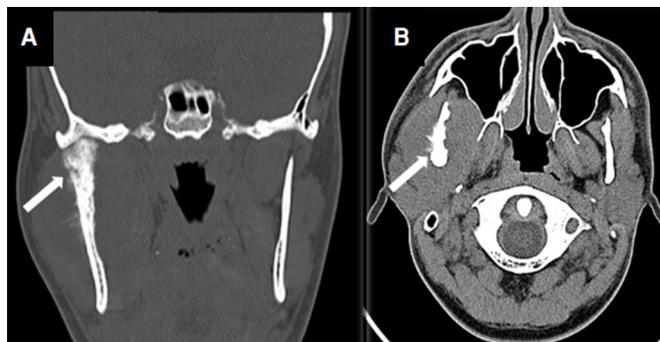
Since the patient presented with soft tissue swelling, ultrasonography was the preferred modality of imaging. Ultrasonography-guided Fine Needle Aspiration Cytology (FNAC) was performed which revealed the presence of a lytic lesion of the mandible suggestive of a neoplastic etiology. Magnetic Resonance Imaging (MRI) showed a hyper-intense lesion with a diffusion restriction with the right ramus and adjacent soft tissue enveloping the condyle ~1.8 cm in maximum thickness on the medial aspect (Fig. 1). The lesion was extending anteriorly to the masseter muscle, medially to the pterygoid muscles, posteriorly abutting the parotid gland with loss of fat planes, and maintained low flow voids.

True cut biopsy was performed and histopathological examination of the lesion revealed the presence of islands and nests of medium-sized cells with round to irregular nuclei, fine chromatin, pale eosinophilic cytoplasm with indistinct cell borders, and occasional mitotic figures with tumour lobules

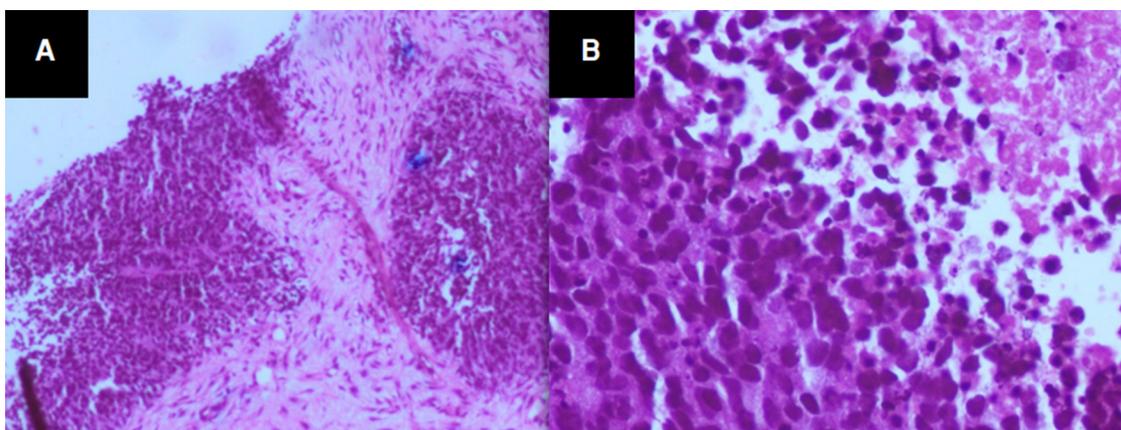
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**Fig. 1.** (A and B) MRI image sections: Post Contrast T1 Fat Saturated Axial sections showing heterogenous post-contrast enhancement; Coronal T2 showing hypertense lesion involving the right condyle and adjacent soft tissue (indicated by white arrows).



**Fig. 3.** (A and B) Axial and coronal sections of the pre-chemotherapy CT scan showing periosteal reaction with loss of retromaxillary fat (indicated with white arrows).



**Fig. 2.** (A and B) Histopathological sections showing tumour cells arranged in sheets with scanty cytoplasm and oval nucleus with coarse chromatin (H&E stain; original magnification,  $\times 400$ ;  $\times 100$ ).

separated by a scanty fibrovascular stroma. The findings were suggestive of Ewing’s Sarcoma or primitive neuroectodermal tumour (PNET) (Fig. 2). Immunohistochemical and immunofluorescence analysis revealed positivity for CD99, synaptophysin, and FLI-1, thus confirming the diagnosis of Ewing’s sarcoma.

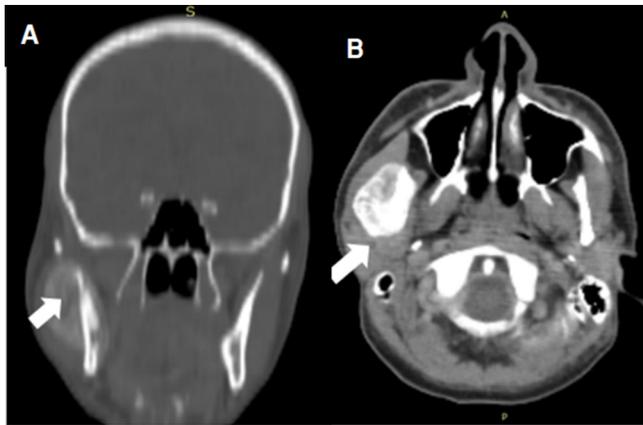
The patient was planned for neoadjuvant chemotherapy followed by surgery and adjuvant therapy. Computed Tomography (CT) imaging prior to chemotherapy showed permeative changes within the ramus of right hemimandible with spiculated periosteal reaction and associated large circumferential soft tissue lesion around the ramus with focal loss of fat planes of the masseter and medial pterygoid muscles. Also, loss of retro maxillary fat was noted (Fig. 3).

The patient received 4 cycles of chemotherapy with cyclophosphamide, doxorubicin and vincristine under the supervision of the Medical Oncology Department. Post chemotherapy repeat CT scan was performed which showed sclerosis, reduced permeative changes in the ramus with ossification, and reduction in the size of surrounding soft tissue along with

restoration of retromaxillary fat (Fig. 4). Positron Emission Tomography (PET) scan did not reveal any signs of distant metastasis. The patient was then offered surgical treatment and adjuvant therapy; however, he opted to undergo alternate natural therapy and did not consent for surgery. The patient succumbed to the disease two months later.

## Discussion

Ewing’s sarcoma also termed as Primitive Neuroectodermal tumour (PNET) is a malignant, small, round, blue-cell tumour with metastatic potential that usually occurs in older children and young adults. Its occurrence in the jawbones is exceedingly rare (6–7% of all bone malignancies), and shows no specific gender predilection [1,3,4]. The most common presenting symptom in the head and neck is a rapidly growing expansile mass along with pain, paraesthesia, and visual disturbances and associated symptoms vary depending on the anatomic area involved [4].



**Fig. 4.** (A and B) Axial and coronal sections of the post-chemotherapy CT scan showing ossification and reduction in the size of the lesion (indicated with white arrows).

ES of the jaw exhibits varied radiologic appearances. It may show a poorly defined permeative lesion, associated sun-ray spicules of the periosteal bone, localized honeycomb appearance, cortical erosion, a soft tissue mass with adjacent bony destruction, displacement or destruction of unerupted tooth follicles. Histologically, Ewing sarcomas are composed of small, round hyperchromatic cells arranged in broadsheets with scant cytoplasm, well-delineated uniform nuclei, and indistinct cellular borders and fibrovascular septa separating the tumour into lobular domains [1,4,5].

The predictor of negative outcomes of Ewing's sarcoma includes the tumour size (>8cm), metastasis, and poor response to neoadjuvant therapy on histological examination. Immunohistochemistry constitutes a useful tool in the diagnosis of Ewing's sarcoma for differentiating it from other small round cell tumours arising from bone and soft tissue. CD99, human natural killer (HNK-1) and friend leukemia integration 1 (Fli-1) are the traditional antibodies and caveolin 1 (CAV-1) is established as an additional marker in the diagnosis of Ewing's sarcoma family of tumours [6,7].

Early and accurate diagnosis is essential for improved survival for patients with this tumour. However, despite extensive aggressive treatment strategies employed, it presents with a high recurrence rate of 50-80%, and a poor prognosis if not diagnosed in the early stages [5,8].

## Conclusion

Ewing's sarcoma (ES) is a rare round cell sarcoma of unknown etiology with uncommon occurrence in jawbones. Higher imaging modalities play a major role in the evaluation of the extent of the lesion and diagnosis. Prompt and pertinent treatment planning is critical for appropriate and timely management.

**Conflicts of interests:** The authors have no conflict of interests to disclose.

## Authors contributions

A.Kudva: Conceptualization, Methodology, M.Kumar: Conceptualization, Writing original draft. D.Nayak: Investigation. M. Sankaran: Supervision. A.Kudva, M.Kumar: Writing- Reviewing and Editing. A.Kudva, M.Kumar, D.Nayak, M.Sankaran: Reviewing and Editing

## Informed consent

The authors declare that informed consent not required as patient's identity is not disclosed.

## Ethical committee approval

The authors declare that Ethical approval not required.

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