

Case Report

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Small Round Cells, Big Diagnostic Challenge: Ewing Sarcoma of the Mandible in a Child

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Abstract: Ewing sarcoma is a malignant small round blue cell tumour which is aggressive in nature and is driven by recurrent chromosomal translocations involving FET–ETS family genes, most frequently the EWSR1–FLI1 fusion. It primarily affects children and adolescents, commonly arises in the diaphysis of long bones. Occurrence in the head and neck region is uncommon, and primary involvement of the mandible is particularly rare. Here we describe the case of a 15-year-old boy who presented with a gradually enlarging painful swelling over the right cheek. On Fine-needle aspiration cytology features were of small round blue cell tumour with occasional rosette-like arrangements. The diagnosis of Ewing sarcoma of the mandible was subsequently established by histopathological examination and immunohistochemistry, which demonstrated diffuse positivity for CD99, NKX2.2, and FLI1. This case underscores the importance of correlating clinical presentation with radiological, cytological, histopathological, and immunohistochemical findings to achieve an accurate diagnosis of this uncommon tumour and facilitate timely management.

Keywords: Small round blue cell tumour; Ewing sarcoma; Mandible; CD99; NKX2.2.

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INTRODUCTION

Ewing sarcoma is a malignant bone tumour composed of uniform small round cells and characterized by FET–ETS fusion genes [1]. It is the second most common malignant bone tumour in children and young adults, with a slight male predilection.

It commonly involves the diaphysis of long bones such as the femur and tibia. Ewing sarcoma affecting the head and neck region is rare, representing approximately 1–4% of all cases [2]. The incidence in the mandible is estimated to be around 0.7% of all Ewing sarcoma sites [3]. Molecular confirmation by demonstration of EWSR1 gene rearrangement using FISH or RT-PCR is considered the diagnostic gold standard.

Because of its rarity in this location, early recognition can be challenging and requires correlation of clinical, radiological, histopathological, and immunohistochemical findings.

CASE PRESENTATION

A 15-year-old boy presented with a painful swelling over the right cheek that had been progressively increasing in size over the preceding three months. The swelling measured approximately 4 × 2 cm. There was

no history of facial trauma, discharge from the swelling, or difficulty in chewing. Clinical examination revealed a firm to hard, fixed swelling involving the right mandibular region. Routine laboratory investigations which includes complete blood count, liver function tests, and renal function tests, were unremarkable.

Magnetic resonance imaging (MRI) demonstrated a well-circumscribed lesion involving the right side of the mandible. The lesion appeared isointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images (Figure 1).

Fine-needle aspiration cytology (FNAC) yielded a cellular smear comprising a monomorphic population of atypical small round cells. These cells exhibited round to oval nuclei with fine stippled chromatin, inconspicuous nucleoli, and scant to moderate cytoplasm. Occasional rosette-like arrangements were identified in a haemorrhagic background, along with scattered mitotic figures (Figure 2). Based on the cytomorphological features, a provisional diagnosis of a small round blue cell tumour was rendered, with lymphoproliferative disorder considered in the differential diagnosis. Histopathological examination was recommended for definitive diagnosis.

The patient subsequently underwent mandibulectomy. Gross examination of the resected specimen revealed a grey-white mass measuring 5×5 cm, with focal areas of haemorrhage and necrosis.

Microscopic examination showed sheets of uniform small round cells with round nuclei, fine stippled chromatin, inconspicuous nucleoli, and scant cytoplasm. Occasional rosette-like arrangements were

also observed. The histomorphological features were consistent with a small round blue cell tumour, favouring Ewing sarcoma (Figures 3 and 4).

Immunohistochemical analysis demonstrated diffuse membranous staining for CD99 and strong nuclear positivity for NKX2.2 and FLI1, confirming the diagnosis of Ewing sarcoma of the mandible.

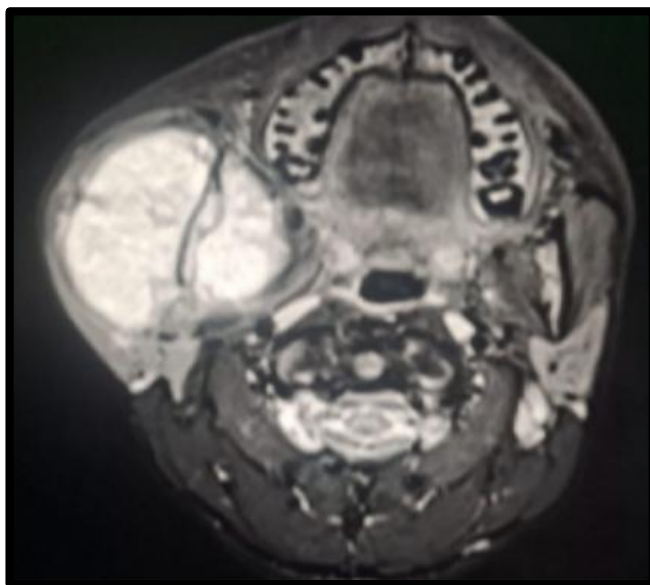


Figure1: Well-defined SOL hyperintense on T2-weighted MRI

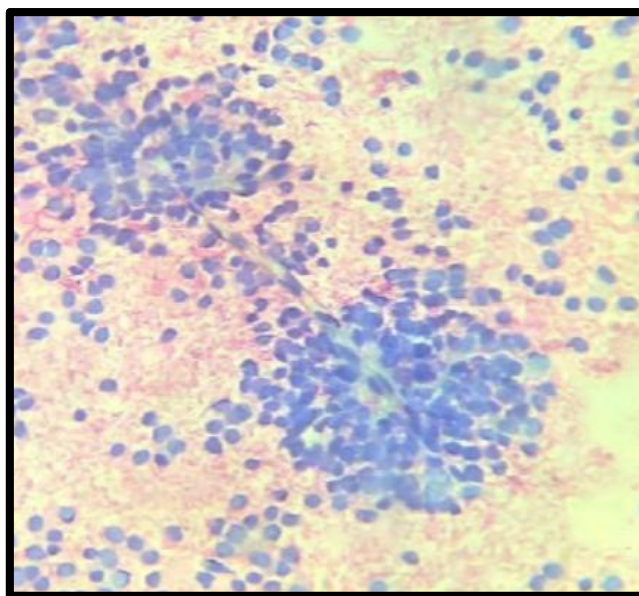


Figure2: FNAC, Geimsa stain, 40x, smear showing rosette formation

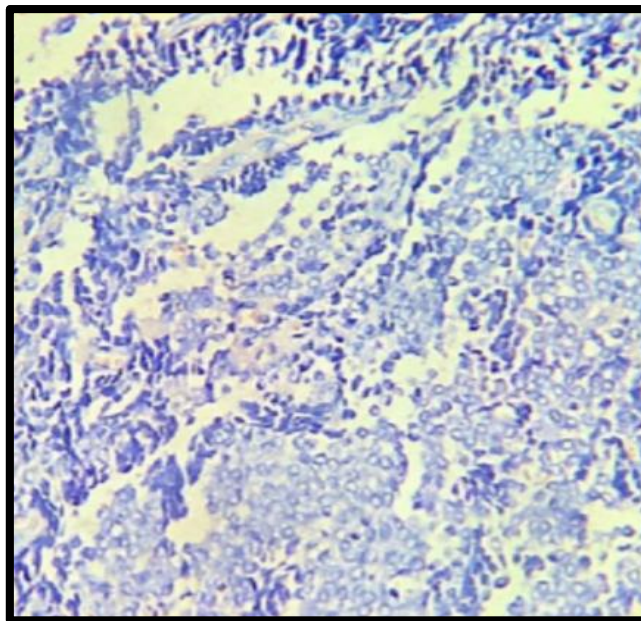


Figure3: H&E, 40x-Sheets of small round blue cells

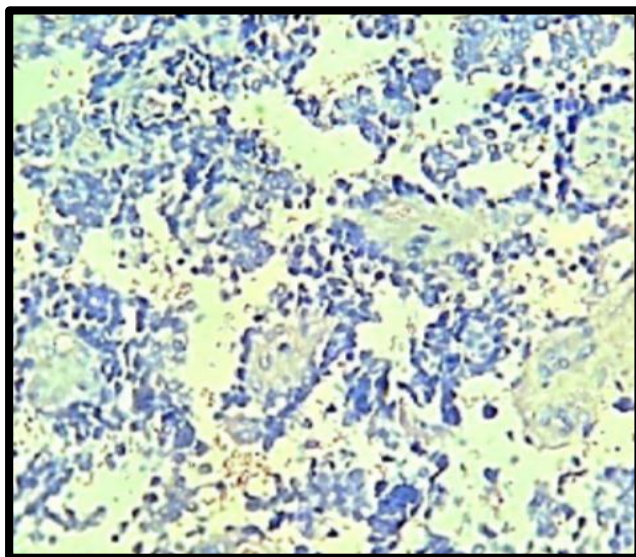


Figure4: H&E 40x, Homer-Wright rosette seen

DISCUSSION

Ewing sarcoma is a rare malignant bone tumour that primarily affects children and adolescents. Although it commonly involves the long bones and pelvis, occurrence in the mandible is uncommon [1-3]. Because of its rarity and nonspecific presentation, mandibular Ewing sarcoma may mimic inflammatory or odontogenic lesions, making early diagnosis difficult.

In the present case, the patient presented with a progressively enlarging painful mandibular swelling. FNAC provided an important initial clue by suggesting a small round blue cell tumour, while histopathological examination and immunohistochemistry confirmed the diagnosis. Similar clinical and pathological findings

have been described in previously reported mandibular cases [2, 3].

The differential diagnosis includes lymphoma, rhabdomyosarcoma, neuroblastoma, and small cell osteosarcoma. Since these tumours share overlapping morphological features, immunohistochemistry is essential for accurate diagnosis. Diffuse membranous positivity for CD99 with nuclear expression of NKX2.2 and FLI1 strongly supported the diagnosis in our case. Although molecular confirmation of EWSR1 gene rearrangement is considered the diagnostic gold standard, it could not be performed because of limited availability.

Current management involves multimodal therapy combining chemotherapy with surgery, with or without



radiotherapy, depending on the extent of disease. Early diagnosis and prompt treatment are important for improving clinical outcomes [6].

CONCLUSION

Primary Ewing sarcoma of the mandible is a rare entity that should be considered in the differential diagnosis of rapidly enlarging mandibular swellings in children and adolescents. This case highlights the value of a multidisciplinary diagnostic approach, with FNAC providing an important initial clue, followed by histopathological examination and immunohistochemistry for definitive diagnosis. Early recognition and timely multimodal treatment are essential for achieving favourable clinical outcomes.

Patient Consent: Informed consent was obtained from the patient's guardian for publication of clinical details and images.

Conflict of Interest: None declared.

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Ethical Statement: None

REFERENCES

1. Ewing J. Diffuse endothelioma of bone. JAMA. 1950;143(4):313–321.
2. Kumar MN, Deepak SB, Singh DK, et al. Ewing's sarcoma of mandible: A rare case report and review of literature. J Clin Diagn Res. 2023;17(6):ZD16–ZD19.
3. Ewing's sarcoma of the mandible: A rare case report and review of literature. Available from: <https://www.researchgate.net/publication/314561041>
4. Balamuth NJ, Womer RB. Ewing's sarcoma. Lancet Oncol. 2010;11(2):184–192.
5. Rekhi B, Basak R, Desai SB, et al. NKX2.2 as a diagnostic marker in Ewing sarcoma: An immunohistochemical study. Indian J Pathol Microbiol. 2018;61(4):567–572.
6. Gaspar N, Hawkins DS, Dirksen U, et al. Ewing sarcoma: Current management and future approaches. Clin Cancer Res. 2015;21(15):3327–3334.

