Title: Ewing Sarcoma of the Mandible in a Pediatric Patient: A Rare Case Report and Review of Management

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Introduction: Ewing sarcoma is a highly aggressive malignant tumor of bone and soft tissue, most frequently affecting the pelvis, femur, and ribs. Mandibular involvement is uncommon, accounting for only 3–5% of cases. Because of its clinical similarity to odontogenic infections, diagnosis is often delayed, which can compromise outcomes. Early biopsy and timely initiation of multimodal therapy are critical. Here, we present a rare case of mandibular Ewing sarcoma in an 11.5-year-old boy, highlighting diagnostic challenges, therapeutic strategies, and the emerging role of immunotherapy.

Case Report: An 11.5-year-old boy presented with a progressively enlarging, firm, and painful swelling of the left mandible for one month. The lesion was initially misdiagnosed as a dental abscess and treated with antibiotics, without improvement. On admission, physical examination showed a large mandibular mass without erythema, inflammatory changes, or systemic symptoms. Laboratory tests revealed WBC 9,300/µL, hemoglobin 10.7 g/dL, and platelets 255,000/µL.Histopathology demonstrated a small round blue cell tumor. Immunohistochemistry confirmed Ewing sarcoma/PNET, showing NKX2.2 positivity, weak CD99 positivity in a subset of tumor cells, and negativity for LCA, CD34, myogenin, desmin, synaptophysin, and SOX10.Radiologic evaluation included CT scan of the neck (6×7×8 cm mandibular lesion with periosteal sunburst reaction), PET/CT (mandible, sphenoid sinus, and tibia involvement), and MRI (heterogeneous mass infiltrating adjacent muscles). Chest CT and abdominal ultrasound were negative for metastasis, and bone marrow examination was normal. Treatment and Outcome:The patient received neoadjuvant chemotherapy (Ifosfamide, VP-16, Vincristine, Doxorubicin, Cyclophosphamide), achieving partial response after six cycles. Following 16 cycles, relapse occurred with new mandibular and sphenoid lesions. Salvage therapy with irinotecan plus temozolomide was started, combined with nivolumab. He subsequently underwent 33 fractions of radiotherapy, resulting in near-complete response, and surgical mandibular resection was performed in 2025.

Discussion: Mandibular Ewing sarcoma is rare and easily mistaken for odontogenic infections, leading to diagnostic delays. The differential diagnosis includes rhabdomyosarcoma, osteosarcoma, and lymphoma. Early biopsy of any persistent mandibular swelling in children is crucial.The standard treatment approach consists of multi-agent chemotherapy and local control via surgery and/or radiotherapy. Despite aggressive multimodality therapy, relapse rates remain high. Immunotherapy and targeted agents represent emerging options in the relapsed/refractory setting. In our case, the addition of nivolumab to salvage chemotherapy contributed to a meaningful radiologic response, suggesting potential benefit in selected patients.

Conclusion: This case highlights the importance of early recognition of mandibular Ewing sarcoma, the role of multidisciplinary management, and the potential application of immunotherapy in relapsed disease. Any persistent mandibular mass in a child should raise suspicion for malignancy. Prompt biopsy is essential to avoid diagnostic delay. Standard care remains multi-agent chemotherapy with local surgical and/or radiotherapy control.

Immunotherapy may offer benefit in refractory or relapsed cases. Close follow-up is vital due to the high risk of recurrence.

Keywords: Ewing sarcoma, Mandible, Pediatric oncology, Immunotherapy, Relapse, Case report