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Massive Pediatric Mandibular Ewing's Sarcoma Presenting with Life-Threatening Airway Obstruction: A Case Report

Bayad Jaza Mahmood *

Department of Oral and Maxillofacial Surgery, College of Dentistry, University of Sulaimani, Madam Mitterrand Street, Old Campus, Sulaymaniyah, Iraq.

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**Corresponding author:*
Bayad Jaza Mahmood

Department of Oral and Maxillofacial Surgery, College of Dentistry, University of Sulaimani, Madam Mitterrand Street, Old Campus, Sulaymaniyah, Iraq.

Tel: +964771102 0000
Email: bayad.mahmood@univsul.edu.iq

ABSTRACT

Ewing's sarcoma is a rare and aggressive malignant small round-cell tumor that typically arises in the long bones and pelvis, with only 1–2% of cases involving the maxillofacial skeleton. Mandibular presentation is exceptionally uncommon and presents diagnostic and therapeutic challenges. This study reports a clinical case and includes a focused literature review. A 9-year-old girl presented with a large mandibular mass associated with airway compromise and recurrent intraoral bleeding. Clinical, radiological, and histopathological examinations were performed. The patient underwent urgent tracheostomy, near-total mandibulectomy with a lip-split incision and 2 cm margins, followed by immediate titanium plate reconstruction. Postoperative care included adjuvant multiagent chemotherapy. A literature review of cases reported between 1988 and 2025 was conducted for comparison. Surgery achieved complete tumor resection with immediate stabilization of the airway and mandibular continuity. Postoperative histopathology confirmed the diagnosis of Ewing's sarcoma with clear margins. The patient recovered with stable airway function and initiated adjuvant chemotherapy. Review of published cases demonstrated variability in presentation, management, and survival outcomes. A summary of comparative mandibular cases is presented in Table 1. Pediatric mandibular Ewing's sarcoma is rare but may present with airway emergencies. Early recognition, multidisciplinary planning, and prompt surgical intervention with airway protection are critical for favorable outcomes.

Keywords: Airway management; Ewing sarcoma; Mandibular neoplasms; Maxillofacial surgery; Pediatric oncology; Reconstructive surgical procedures.

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Introduction

Ewing's sarcoma (ES) is a highly aggressive malignant neoplasm of neuroectodermal origin, first described by James Ewing in 1921. It most commonly arises in long bones, pelvis, and ribs, with only 1–2% of cases reported in the jawbones.1 Mandibular involvement is particularly rare and may mimic odontogenic tumors, osteomyelitis, or inflammatory lesions [2]. Ewing's sarcoma primarily affects children and adolescents between 10–20 years, with a slight male predominance (1.5:1) [3,4]. Clinical features include pain, swelling, loosening of teeth, paresthesia, and intraoral bleeding. Systemic findings, such as fever and leukocytosis, can mislead clinicians toward a diagnosis of infection.5 Radiographically, mandibular ES typically presents with ill-defined mixed lesions, cortical erosion, and occasionally a "sun-ray" periosteal reaction [6,7]. CT and MRI are crucial for delineating the extent of bone destruction and soft tissue spread [8], while PET-CT provides accurate staging and treatment monitoring [9]. Histopathology reveals small round blue cells arranged in sheets. Immunohistochemistry shows strong CD99 positivity, with vimentin and BCL-2 frequently co-expressed [10]. Over 90% of cases exhibit the EWSR1-FLI1 translocation, detectable via RT-PCR or FISH [11]. Treatment of Ewing's sarcoma is multidisciplinary, combining systemic chemotherapy with local control through surgery and/or radiotherapy. Multi-agent chemotherapy regimens, including vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide, have significantly improved survival outcomes, with 5-year survival rates exceeding 70% in localized disease [12,13]. Surgical resection with negative margins remains the cornerstone of local control, particularly in accessible sites such as the mandible, while radiotherapy is reserved for unresectable or residual tumors [14]. Advances in targeted therapy and immunotherapy are under investigation and may further improve prognosis [15].

Case Report

A 9-year-old girl presented to the Sulaymaniyah Emergency Hospital, Iraq, with a 2-month history of severe pain, rapidly enlarging mandibular swelling, recurrent intraoral bleeding, and multiple suffocation episodes.

Clinical findings

On admission, the child was cachectic, febrile (38 °C), cyanotic, and dyspneic, with fluctuating oxygen saturation (75–95%). The mandibular mass extended from the ala–tragus line to the supraclavicular region, causing gross asymmetry. The mass was firm, with dilated surface veins, tooth mobility, spontaneous bleeding, and drooling. No lymphadenopathy was evident (Figure. 1a, b). Figure 1. Preoperative clinical presentation of mandibular Ewing's sarcoma. (A) Frontal view demonstrating diffuse swelling with facial asymmetry with mucosal stretching and obliteration of the oral cavity. (B) Lateral view showing lateral expansion of the tumor.

Imaging

Computed tomography showed a diffuse, ill-defined lytic lesion from the left condyle to the right first premolar, destroying the hemimandible with moth-eaten radiolucencies and cortical discontinuity. A sun-ray periosteal reaction was noted (Figure. 2).

Histopathology

Incisional biopsy revealed small, round hyperchromatic cells. Immunohistochemistry confirmed CD99, CD117, vimentin, BCL-2, and EMA positivity, consistent with Ewing's sarcoma (Figure. 3).

Management

Airway Management

Given the extent of tumor bulk and the resulting airway compromise, the procedure was initiated with fiberoptic-guided nasal intubation, followed by a tracheostomy to establish a secure airway and reduce the risk of intraoperative respiratory obstruction. This step ensured safe anesthetic management throughout the procedure.

Surgical Procedure

A lip-split incision was employed to achieve optimal exposure of the mandible and associated soft tissues. A near-total mandibulectomy was then performed with oncologically safe margins of approximately 2 cm. The resection included the left condyle, ramus, and mandibular body, and was extended contralaterally beyond the right first molar to ensure complete removal of the lesion. Figure 4 (a, b).

Reconstruction

Immediate reconstruction was carried out using a titanium reconstruction plate to restore mandibular continuity and function. One end of the plate was anchored to the glenoid fossa using secure wiring, while the opposite end was fixed to the remaining mandibular stump. To maintain airway safety and prevent postoperative tongue base collapse, the digastric and mylohy-

oid muscles, as well as the tongue base, were anchored to the reconstruction plate. This provided both structural stability and dynamic support for swallowing and airway protection. Figure 4 (c, d).

Postoperative Care and Course

Following surgery, the patient was admitted to the intensive care unit for close observation. Management focused on maintaining airway patency, effective analgesia, hemodynamic stability, and nutritional support. Figure 5. Histopathological analysis confirmed that the resection margins were free of tumor involvement. The patient subsequently underwent adjuvant multi-agent chemotherapy with vincristine, dactinomycin, cyclophosphamide, and doxorubicin, in accordance with established protocols for Ewing's sarcoma. Future reconstructive planning considered a vascularized free flap or vascularized bone graft once the patient achieved systemic stabilization, to restore long-term functional and esthetic outcomes.

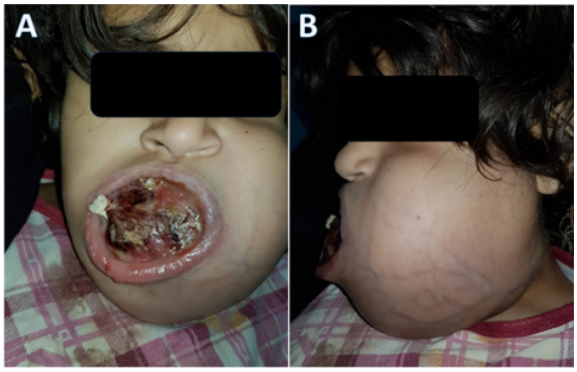

Figure 1. Preoperative clinical presentation of mandibular Ewing's sarcoma. (A) Frontal view demonstrating diffuse swelling with facial asymmetry with mucosal stretching and obliteration of the oral cavity. (B) Lateral view showing lateral expansion of the tumor.

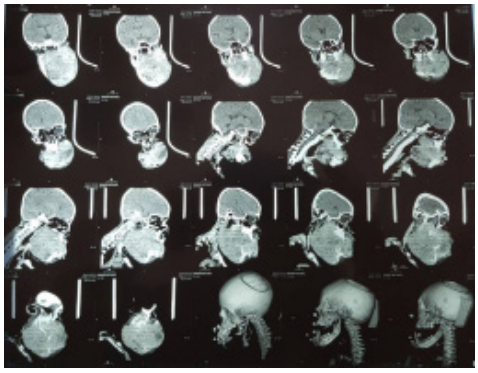

Figure 2. Preoperative CT imaging demonstrating a large expansile mandibular lesion with cortical erosion and extensive soft tissue involvement. Three-dimensional reconstruction shows the extent of mandibular destruction and spatial relation to adjacent craniofacial structures.

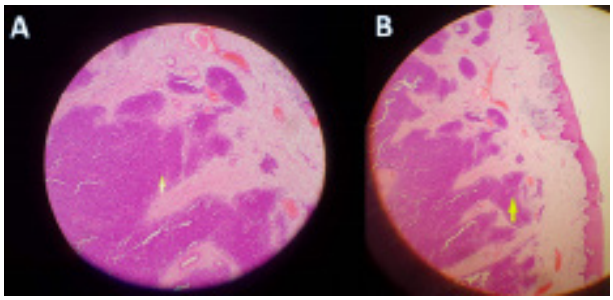

Figure 3. Histopathological features of Ewing's sarcoma (H&E stain). (A) Low-power view showing sheets of small round blue cells (arrow). (B) Higher magnification demonstrating tumor infiltration with preserved surface epithelium (arrow).

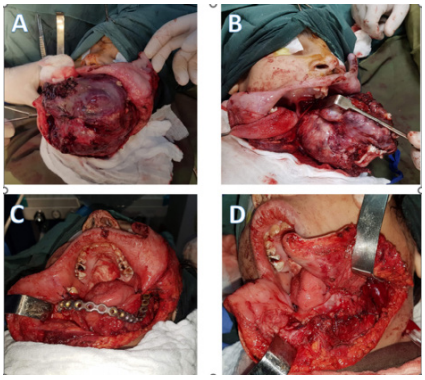

Figure 4. Intraoperative steps of surgical management. (A) Exposure of the tumor mass via lip-split incision. (B) Near-total mandibulectomy with tumor removal. (C) Immediate reconstruction with titanium reconstruction plate. (D) Anchoring of soft tissues including tongue base and floor of mouth musculature to the plate to maintain airway patency.

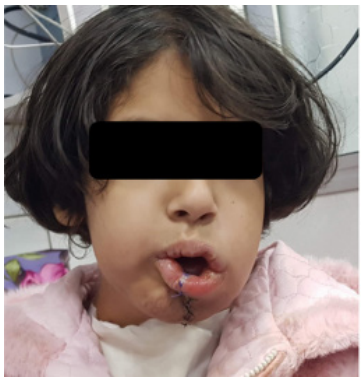

Figure 5. Early postoperative extraoral photograph showing satisfactory healing of lip incision with sutures in place and improvement in facial contour.

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Table 1. Representation of reported mandibular Ewing's sarcoma cases (1988–2025).

Author (Year)	Gender	Age (years)	Location	Clinical features
Van Den Bergh et al. (1988)[21]	M	12	Mandible	Swelling, pain, and fever
Wang et al. (1991)[22]	F	12	Mandible	Pain and paresthesia
Fonseca et al. (1992)[23]	M	4	Mandible	Painless nodule
Bessède et al. (1993)[24]	M	8	Mandible	Painless mass
Berk et al. (1995)[25]	F	5	Mandible	Swelling around impacted tooth
Vaccani et al. (1999)[26]	M	11	Mandible	Painful swelling
Fonseca et al. (2000)[27]	F	35	Mandible	Pain and paresthesia
Gorospe et al. (2001)[7]	F	12	Mandible	Fever and swelling
Talesh et al. (2003)[28]	F	17	Mandible	Painful swelling
Schultze-Mosgau et al. (2005)[8]	M	7	Mandible	Painful swelling
Lopes et al. (2007)[29]	M	14	Mandible	Painful swelling
Gosau et al. (2008)[30]	M	24	Mandible	Pain and sensory loss
Saleh et al. (2018)[31]	F	4	Mandible	Painless swelling
Sinha (2020)[32]	M	8	Mandible	Pain and swelling
Reddy et al. (2022)[10]	M	16	Mandible	Painful rapidly growing mass
Ram et al. (2023)[33]	F	10	Mandible	Swelling and fever
Bellut et al. (2024)[34]	M	12	Mandible	Tender swelling
Present case (2025)	F	9	Mandible	Severe airway compromise

Discussion

Ewing's sarcoma (ES) of the mandible is a rare and aggressive tumor, comprising only 1–2% of jaw neoplasms [1]. Clinically, it often presents with painful swelling, bleeding, tooth mobility, and paresthesia, which may resemble odontogenic infections or inflammatory lesions [2,3]. Radiographic features include cortical erosion, ill-defined lytic lesions, and occasionally a "sun-ray" periosteal reaction, which can mimic osteomyelitis [4-7]. Advanced imaging with CT and MRI is indispensable for delineating tumor spread and guiding surgical planning [8], while PET-CT plays a vital role in staging and treatment monitoring [9]. Histopathological examination typically shows sheets of small round blue cells, with immunohistochemical positivity for CD99, vimentin, and BCL-2 [10]. More than 90% of cases demonstrate the characteristic EWSR1-FLI1 translocation, detectable by RT-PCR or FISH.11

Advances in therapy (2020–2025)

Management of ES has shifted toward a multidisciplinary model, with surgery, chemotherapy, and radiotherapy integrated into treatment protocols. Surgical resection with clear margins remains the cornerstone of local control, and when combined with chemotherapy, 5-year survival rates for localized disease can exceed 70% [12,13]. Standard chemotherapy regimens employ vincristine, doxorubicin, cyclophosphamide, ifosfamide, and actinomycin-D [14], while radiotherapy is generally reserved for unresectable or residual lesions [15]. In recent years, advances in molecular profiling and targeted therapy have opened new avenues. Specific EWSR1-FLI1 fusion subtypes are associated with prognostic differences, offering opportunities for risk stratification. Novel targeted agents, including IGF-1R inhibitors and PARP inhibitors, are under active clinical investigation, showing encouraging results as adjuncts to conventional regimens [16-18]. Furthermore, immunotherapy and other molecularly driven strategies are being explored, holding promise for cases resistant to standard treatment [19-20].

Reconstructive strategies and airway considerations

Mandibular ES presents unique surgical challenges, especially in children, where both facial growth and function must be preserved. Advances in reconstructive approaches, such as patient-specific implants and staged vascularized bone grafting, have significant-

ly improved outcomes [8,12]. Airway compromise, though rarely emphasized, can become life-threatening in advanced mandibular disease. In our case, severe obstruction necessitated emergency tracheostomy before tumor resection. This highlights the critical need for early diagnosis and airway-focused multidisciplinary planning.

Comparison with the literature

A review of previously reported mandibular ES cases from 1988–2025 is summarized in Table 1. Most patients presented with swelling and pain, with occasional paresthesia or misdiagnosed cases. Treatment typically included wide resection and chemotherapy, with varying reconstructive methods. Our case is distinguished by its delayed presentation, critical airway compromise, and the need for near-total mandibulectomy with immediate titanium plate reconstruction.

Conclusion

Mandibular Ewing's sarcoma remains a rare but serious pediatric malignancy requiring prompt recognition and coordinated care. Surgical resection with negative margins, supported by systemic chemotherapy, remains the foundation of treatment. Airway protection should be prioritized in advanced cases, and reconstruction should balance functional rehabilitation with growth considerations. Recent advances in molecular biology, including targeted therapies and immunotherapy, provide optimism for improving survival and long-term quality of life.

Conflict of Interest

There is no conflict of interest to declare.

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