IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 24, Issue 2 Ser. 2 (February. 2025), PP 32-35
www.iosrjournals.org

Extra-Skeletal Ewing Sarcoma Mimicking A Vascular Tumor: A Case Report

R.Ezzahraoui; O. Elfilali; O. Almaghraoui; M.Alaoui
Department Of Peripheral Vascular Surgery Hma

Abstract
Introduction :
Extra-skeletal Ewing sarcoma (EES) is a malignant tumor with a male predominance, accounting for less than 1% of malignant tumors in the head and neck. The definitive diagnosis is histopathological. Treatment consists of surgical excision, as well as postoperative radiotherapy and chemotherapy.
Case Report :
A 13-year-old girl was admitted for severe anemia following prolonged active bleeding from an intraoral mass. She underwent embolization combined with surgical excision, which successfully removed the entire tumor. The histopathological examination confirmed the diagnosis of extra-skeletal Ewing sarcoma.
Conclusion :
Extra-skeletal Ewing sarcoma is a rare condition, and its diagnosis is often challenging. Treatment is based on complete excision of the tumor followed by appropriate radiotherapy and chemotherapy.

Date of Submission: 27-01-2025 Date of Acceptance: 07-02-2025

I. Introduction

In 1921, a new family of tumors was described by James Ewing (1866-1943): Ewing's sarcoma, which was distinguished from lymphomas and other types of cancers known at that time. The term 'sarcoma' is actually inappropriate, as it is a primitive neuroectodermal tumor (PNET). [1]

It is a malignant, locally aggressive tumor observed more frequently in males than in females during the first three decades of life. [2]

Head and neck sarcomas represent approximately 2% to 15% of all sarcomas. [3] Tumors located in the orbit, the retro-pharynx, and the nose have been reported. [4-5]

The mechanism behind the development of this type of sarcoma remains unknown due to its relative rarity, making it difficult to determine the oncogenic factors. [7] Sarcomas are classified based on their tissue of origin (bone or soft tissue). [8]

The diagnostic approach is based on clinical evaluation and radiological investigations: soft tissue ultrasound and MRI. However, the definitive diagnosis is histopathological. The development of immunohistochemistry and tumor markers has strengthened our ability to subclassify sarcomas.

Surgery remains the cornerstone of curative treatment for soft tissue sarcomas and the central element of the therapeutic arsenal. [9,10] Adjuvant radiotherapy should be considered for patients with locally recurrent lesions and intermediate to high-grade tumors.

Given the poor prognosis of Ewing's sarcoma, management should involve multidisciplinary consultation committees composed of radiologists, surgeons, radiotherapists, pathologists, and medical oncologists. [10,11]

We report here an extremely rare case of extra-skeletal Ewing sarcoma of the face affecting the left internal jugular vein.

II. Case Report

The patient, R.B., a 13-year-old female with no significant medical history, presented with a left lateral cervical swelling that had been evolving for 6 months. The swelling progressively increased in size until it protruded intraorally, preventing mouth closure, speech, and eating, with intermittent bleeding and associated respiratory difficulties.

The patient then developed severe anemia with a hemoglobin level of 3.5 g/L due to prolonged active bleeding, which required her admission to the emergency department for management.

Embolization of the tumor was performed, along with a transfusion of 7 blood units, which successfully stopped the bleeding (Figure 1)

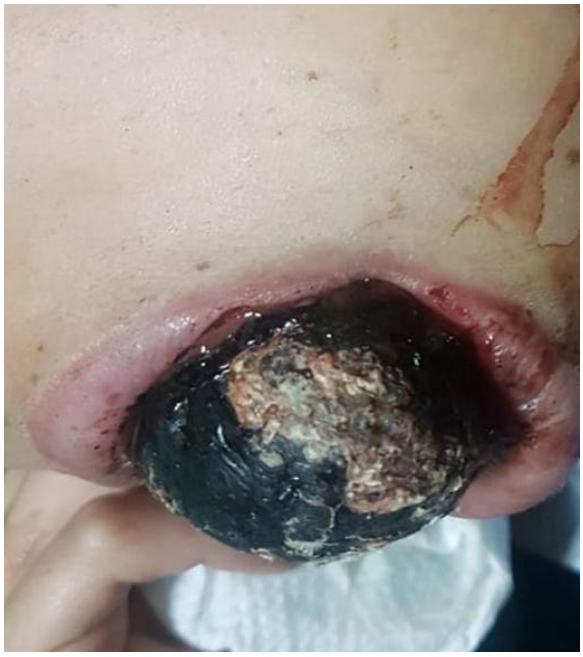
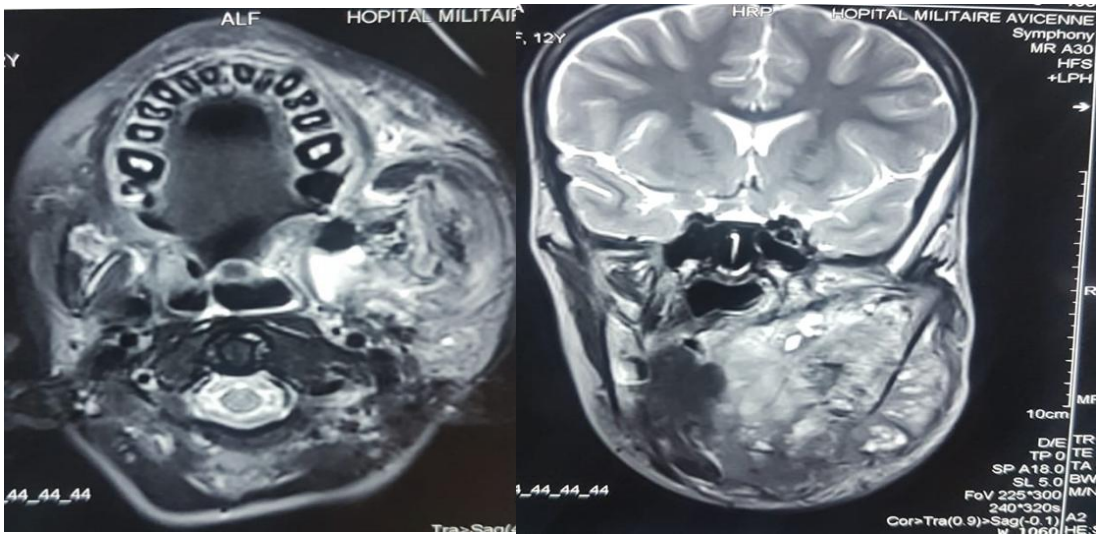


Figure 1: The intraoral tumor after embolization.

The cervical-facial CT scan reveals a large, highly aggressive tumor process in the left mandibular cavity, centered on the mandibular angle, lytic to the cortical bone with significant invasion of the deep facial spaces, particularly the masticator space, the floor of the mouth, and the intraoral soft tissues, which significantly narrow the oropharynx.

The cervical-facial MRI highlights a left jugal process centered on the mandibular angle, poorly defined and heterogeneous, measuring 130 mm in the antero-posterior axis, 50 mm in width, and 58 mm in height. It lysis the mandible and infiltrates the parotid gland and the external masticatory muscles, while displacing the pharynx, yet sparing the signal of the tongue (Figures 2 and 3).



Figures 2 and 3: MRI images showing the heterogeneous and poorly defined mass

A complementary surgery was performed by a double team of vascular and maxillofacial surgeons under general anesthesia with the patient in the dorsal decubitus position. After a left cervical incision, the first step involved a vascular procedure with ligation of the left internal jugular vein, ensuring preservation of the carotid artery. In the second step, the inferior pole was resected, followed by intraoral excision of the mass and the infiltrated alveolar bone, which allowed complete isolation of the process at the buccal and infratemporal levels. Finally, the cervical access was sutured with an intraoral plastic procedure to close the left jugal and retro-trigonal substance loss, resulting in a good postoperative outcome (Figure 4)

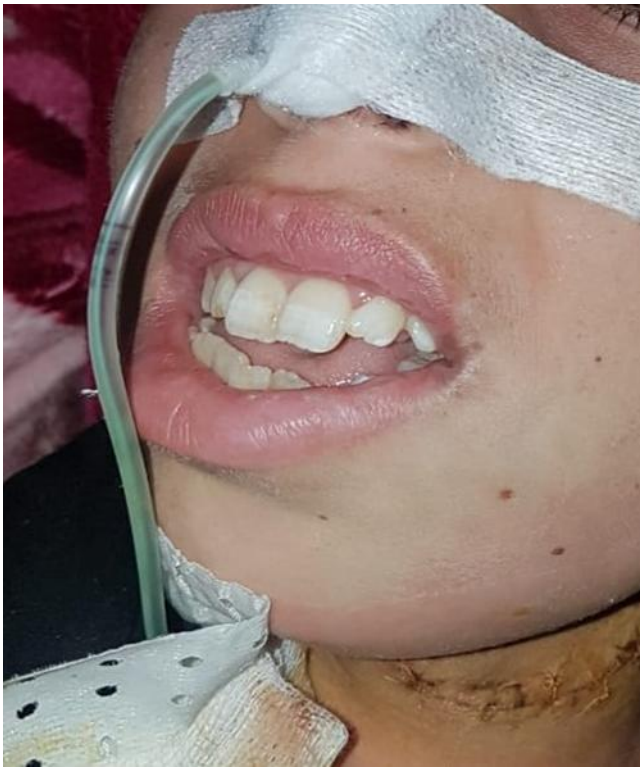


Figure 4: Postoperative result after complete removal of the mass

The histopathological analysis of the surgical specimen revealed a malignant mesenchymal tumor with small round cells, showing a CD99-positive immunohistochemical profile, which pointed towards the PNET/Ewing sarcoma group.
Cytogenetic examination confirmed the diagnosis of Ewing sarcoma with a t(11;22) translocation.

III. Discussion :

Soft tissue sarcomas are rare [10,12], accounting for 0.5 to 1% of malignant tumors in adults, with an annual incidence estimated at 3 to 8 per 100,000 [13]. They are less frequent than benign soft tissue tumors, according to the study by Enzinger and Weiss [14].

Extra-skeletal Ewing sarcomas (EES) in the head and neck region are extremely rare, representing only 1% to 4% of cases [2,6]. They rarely occur after the third decade of life, with a male predominance [2,6]. The head and neck region is a rare primary site for extra-skeletal Ewing sarcoma (EES). Chao et al. described only 5 cases out of 118 EES cases located in the head and neck region [15].

There are no specific symptoms for extra-skeletal Ewing sarcoma. Most patients present with a painless mass, indicating rapid growth. One-third of cases have distant metastases at the time of diagnosis [16].

While ultrasound is the first-line examination for any soft tissue swelling [17], MRI remains the method of choice for diagnosing soft tissue sarcomas, showing signal and contrast abnormalities that vary according to histological types [9,18]. Angiography is very useful for studying vascular structures near the tumor.

Radiological assessment does not allow for confirmation or exclusion of the malignancy of the tumor or its histological type. However, it is essential for guiding biopsy, selecting the approach, and performing a disease extension workup.

The definitive diagnosis is based on histopathology, supplemented by immunohistochemical studies in cases of diagnostic difficulty [7, 9, 12]. The histological appearance of EES shows round cells with sparse cytoplasm.

Angervall and Enzinger first described the characteristics of EES [19]. Ewing sarcomas express CD99 on their cell membranes, and thus the staining with anti-CD99 antibodies confirms the diagnosis.

In our case, physical examination and radiological exploration suggested a vascular tumor. Indeed, fibrovascular septa are present in most lesions and can mimic a vascular tumor.

The treatment of extra-skeletal Ewing sarcoma is based on complete surgical excision of the tumor followed by radiotherapy and chemotherapy.

Exclusive radiotherapy has a high chance of local recurrence; therefore, radical surgical removal is necessary to reduce the risk of recurrence.

Tumors of the neck have a better survival rate compared to other anatomical locations [6]. The 5-year survival rates for EES are reported between 38% and 67%.

EES has a poor prognosis; however, surgical excision with chemotherapy and radiotherapy improves survival chances [20,21].

IV. Conclusion

Despite its rarity and high risk of recurrence, the management of extra-skeletal Ewing sarcomas has improved through scientific and technological advancements that have standardized their complex treatment approach.

The diagnosis is based on radiological investigations as well as histopathological data.

Treatment relies on complete surgical removal of the tumor mass, followed by postoperative radiotherapy and chemotherapy.

Conflict Of Interest: The authors declare no competing interests.

References :

[1] Ewing J. Diffuse Endothelioma Of Bone. Proc N Y Pathol Soc. 1921;21:17.
[2] Prindull G, Willert HG, Notter G. Local Therapy Of Rhabdomyosarcoma, Osteosarcoma And Ewing's Sarcoma Of Children And Adolescents. Eur J Pediatr 1985;144(2):120-4.
[3] Brockstein B. Management Of Sarcomas Of The Head And Neck. Curr Oncol Rep 2004;6(4):321–7.
[4] Alvarez A, Schut L, Bruce D. Localized Primary Intracranial Ewing's Sarcoma Of The Orbital Roof. Case Report. J Neurosurg 1979; 50:811- 3.
[5] Hadfield MG, Luo VY, Williams RL, Ward JD, Russo CP. Ewing's Sarcoma Of The Skull In An Infant. Pediatr Neurosurg 1996; 25:100- 4.
[6] Seigal GP, Oliver WR, Reinus WR, Et Al. Primary Ewing's Sarcoma Involving The Bones Of The Head And Neck, Cancer 1987; 60:2829- 40.
[7] F.Collin , M. Gelly-Marty, M. Bui Nguyen Binh, J.M Et Coll . Sarcomes Des Tissus Mous : Données Anatomopathologiques Actuelles. Cancer/Radiothérapie 10 (2006) 7-14.
[8] Sturgis EM, Potter BO. Sarcomas Of The Head And Neck Region. Curr Opin Oncol 2003;15(3):239–52.
[9] Société Canadienne Du Cancer. Vu D'ensemble Du Sarcome Des Tissus Mous. Encyclopedie Canadienne Du Cancer (2012).
[10] J.Fayette , J.Y.Blay, I.Ray-Coquard. Les Sarcomes Des Tissus Mous: Bonnes Pratiques Médicales Pour Unr Prise En Charge Optimale. Cancer/ Radiothérapie 10 (2006) 3-6.
[11] L. Zelek. La Chimiothérapie Des Sarcomes Des Tissus Mous Cancer/ Radiothérapie 10 (2006) 68-71
[12] JM. Coindre, Pourquoi Et Comment Classer Un Sarcome Des Tissus Mous. Elsevier Masson (2006) 98-109
[13] S.Bonvalot, D.Vanel, P.Terrier Et Coll. Principes Du Traitement Des Sarcomes Des Tissus Mous De L'adulte EMC(Elsevier,Paris), Appareil Locomoteur, (2004) :44-099.
[14] Y. Miki, S. Ngan , J.C.M Et All. The Significance Of Size Change Of Soft Tissue Sarcoma During Preoperative. The Journal Of Cancer Surgery 36 (2010) 678-683
[15] Chao TK, Chang YL, Sheen TS. Extraskeletal Ewing's Sarcoma Of The Scalp. J Laryngol Otol 2000. 114:73–75.
[16] Rud NP, Reiman HM, Pritchard DJ, Frassica FJ, Smithson WA.Extraosseous Ewing's Sarcoma: A Study Of 42 Cases. Cancer 1989; 64(7):1548-53.
[17] A-M. De Shepper, J.Gielien, F.Vanhoenacker Et Coll. Imagerie Des Parites Molles. EMC/31-750-A-10 (2010)
[18] Wilson.Re, Antman.Kh, Brodsky.G Et Coll Facteurs Pronostiques Des Sarcomes Des Tissus Mous. EMC Tome II -2004
[19] Angervall L, Enzinger FM. Extraskeletal Neoplasm Resembling Ewing's Sarcoma. Cancer 1975. 36:240–251.
[20] Raney RB, Asmar L, Newton WA Jr, Et Al. Ewing's Sarcoma Soft Tissues In Childhood: A Report From The Intergroup Rhabdomyosarcoma Study 1972 To 1991. J Clin Onc 1997; 15(2):574-82.
[21] Rud NP, Reiman HM, Pritchard DJ, Frassica FJ, Smithson WA.Extraosseous Ewing's Sarcoma: A Study Of 42 Cases. Cancer 1989; 64(7):1548-53.