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Primary Ewing's Sarcoma of the Temporal Bone: An Unusual Case with Metastasis at a Distance

Primary Ewing's Sarcoma of the Temporal Bone: An Unusual Case with Distant Metastasis

Primary Ewing sarcoma of the temporal hue: an unusual case with metastasis to distance

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Summary

Introduction: Ewing's sarcoma is a rare and aggressive type of cancer that affects...
bone or soft tissues, originating from precursor cells of bone tissue or
peripheral nervous system, with the peak incidence occurring in the second decade of life.
They are most commonly found in long bones, but can rarely affect the
Head and neck region. **Report:** The case presented is that of a teenage girl.
A 13-year-old female who initially presented with left-sided ear pain and
He subsequently developed ipsilateral peripheral facial paralysis. The diagnosis
Subsequent examination revealed a primary Ewing's sarcoma of the temporal bone. **Discussion:** The
The topics discussed include the evolution of the disease, its clinical and pathological characteristics, as well as...
including the diagnostic and therapeutic methods employed in this case. **Conclusions:**
This is an extremely rare case of Ewing's sarcoma of the temporal bone with
Distant metastases. The evolution of this case reinforces the need for constant monitoring.
monitoring and considering the possibility of remote dissemination, guiding the
establishing personalized treatment protocols that seek to optimize the
clinical outcomes in such unique and challenging situations.
Keywords: Ewing's sarcoma. Facial paralysis. Head and neck neoplasms.

Abstract

Introduction: Ewing's sarcoma is a rare and aggressive type of cancer that affects bone
or soft tissues. It originates from precursor cells of bone tissue or the peripheral nervous
system, with its incidence peaking during the second decade of life. It is most commonly
found in long bones but can, rarely, affect the head and neck region. **Case Report:** This
case involves a 13-year-old female adolescent who initially presented with left-sided
otalgia and developed subsequently ipsilateral peripheral facial paralysis. Subsequent
diagnosis revealed a primary Ewing's sarcoma of the temporal bone. **Discussion:** The
discussion covers the disease progression, its clinical and pathological characteristics, as
as well as the diagnostic and therapeutic methods applied in this case. **Conclusions:** This is
an extremely rare case of Ewing's sarcoma of the temporal bone with distant metastases.
The progression of this case highlights the need for constant vigilance and the
consideration of the potential for distant spread, guiding the establishment of

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personalized treatment protocols aimed at optimizing clinical outcomes in such unique
and challenging situations.

Keywords: Sarcoma, Ewing. Facial Paralysis. Head and Neck Neoplasms.

1. Introduction

Ewing's sarcoma, a histopathological subtype of sarcoma composed of small
Round cell carcinoma, which frequently affects the long bones of the limbs and trunk, was first
described by James Ewing in 1921. It is much more commonly observed in
Caucasian children and adolescents, and somewhat more common in individuals of
It is more common in males and is generally diagnosed in the second decade of life. Among the
which are located in the skull, an extremely small percentage may be
develop in the temporal bone, usually involving the frontal, parietal or
ethmoid 2.

A complex case of primary Ewing's sarcoma of the temporal bone is presented, with
Intracranial extension, affecting a 13-year-old female adolescent.
age, focusing on clinical manifestations, diagnostic challenges faced, and
a multidisciplinary approach, which was essential for the formulation of a plan.
appropriate therapy. The finding of distant metastasis shows that
This is an atypical presentation of the disease, as there is only one case described in the literature with
a similar evolution. The study was approved by the Ethics Committee and
Research (CEP) with CAEE no. 71314223.9.3001.5042 and is linked to the Hospital
Albert Sabin Children's Hospital – CE.

2. Case Report

A 13-year-old female adolescent presented with a history of left-sided otalgia.
associated with pain in the ipsilateral mandibular region, initially with partial relief after
use of nonsteroidal anti-inflammatory drugs, in addition to sporadic episodes of vertigo
Rotational pain and nausea. Over three months, it progressed to left-sided peripheral facial paralysis.
ipsilateral progressive hearing loss, mild dysarthria, intermittent liquid dysphagia,
She reported bloody otorrhea and a significant worsening of pain intensity, which led her to...

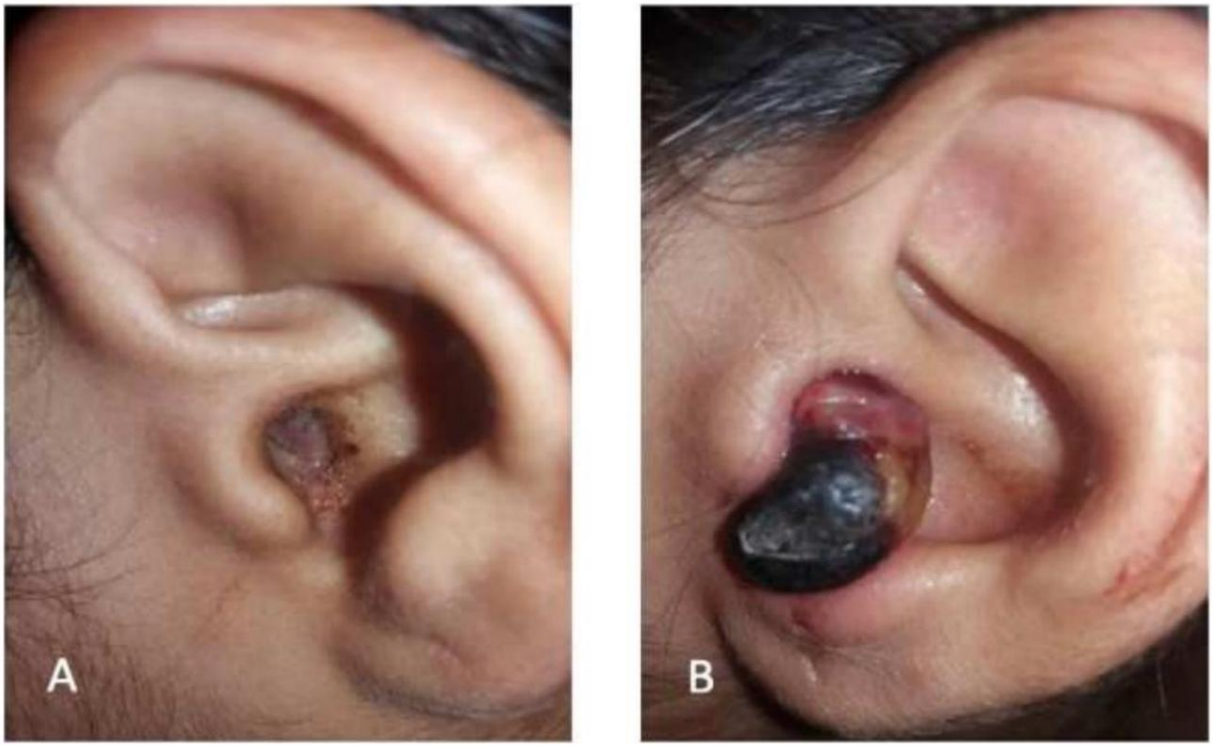
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seeking medical attention, she was referred to the General Hospital of
Strength.

On physical examination, the patient presented with peripheral facial paralysis on the left, House-
Brackmann 4, as well as a polypoid lesion protruding from the auditory canal.
left (Figure 1).

Figure 1: Polypoid lesion protruding from the left auditory canal. (A) Onset.
(B) After 3 months



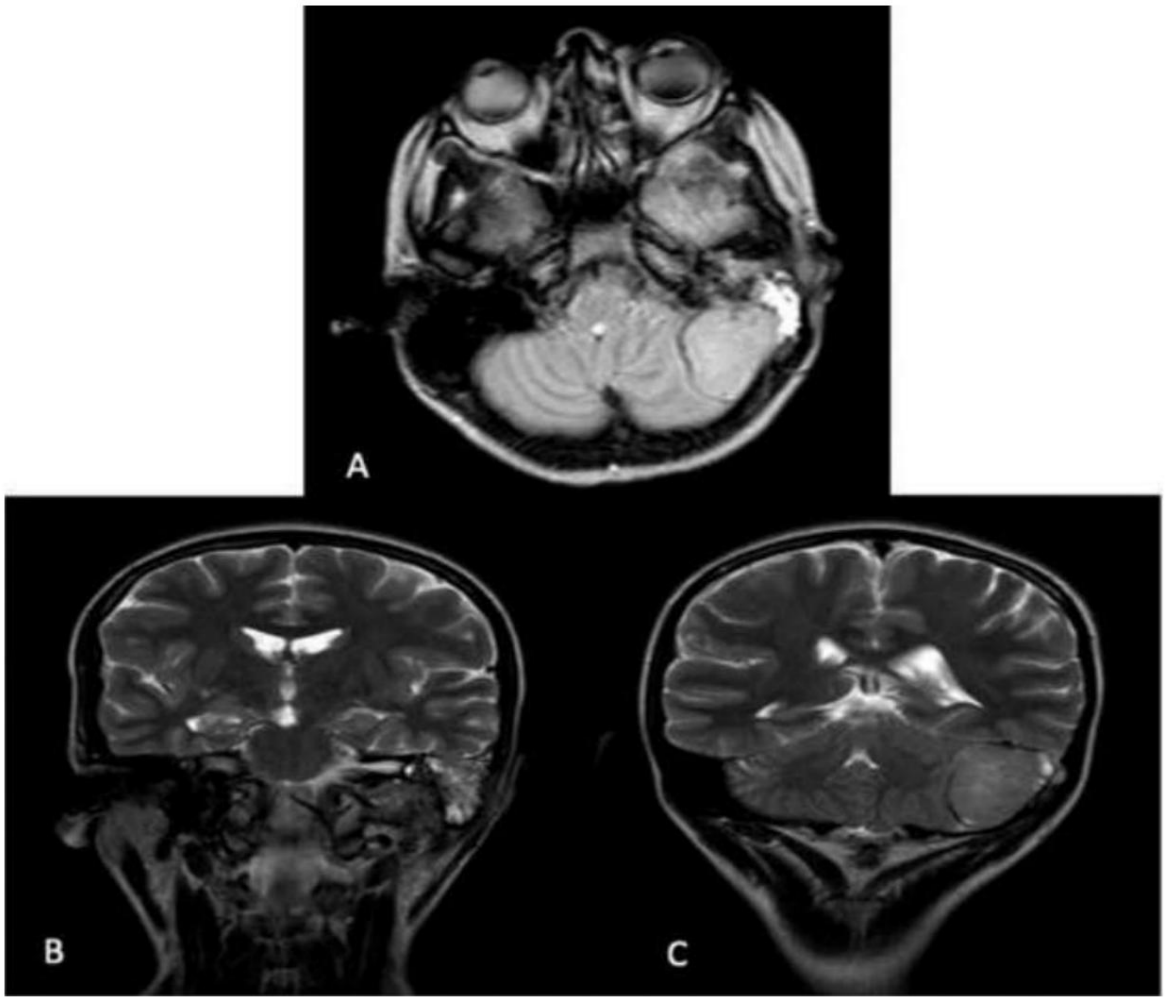
A head and neck computed tomography (CT) scan with contrast revealed
A lesion showing contrast enhancement in the region of the left jugular foramen, with
erosion of the adjacent temporal bone, communication with mastoid cells and ear.
middle ear and involvement of the components of the inner ear, with compression of the
ipsilateral sigmoid sinus and jugular bulb by the posterior component, measuring 3.0 x 1.9
2.3 cm x 2.3 cm. Initially, it was hypothesized to be a low-grade neoplastic lesion.
Aggressiveness (such as meningioma, paraganglioma, or schwannoma) or apical petrositis.
A cranial Magnetic Resonance Imaging (MRI) scan was also performed, which showed that the lesion
It showed predominantly isointense signal on T1 and T2, infiltrating the space.
left parotid gland, with involvement of the deep portion of the parotid gland,

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extending to the left posterior fossa, leading to thrombosis of the sigmoid sinus, bulb jugular and the cranial aspect of the internal jugular vein on the left (Figure 2). In Audiometry revealed profound mixed hearing loss on the left and a hearing threshold Normal on the right.

Figure 2: MRI revealing expansive extra-axial lesion in the pre-cerebellar cistern, extending to the tympanic cavity, external auditory canal, and ipsilateral jugular foramen. (A) Axial section – FLAIR (B) Coronal section - T2 (C) Coronal section - T2



Opting for joint surgery with an otolaryngology team, surgery head and neck and neurosurgery. Preoperatively, she underwent angiography. Cerebral and external carotid branch embolization. The surgical procedure It consisted of a partial left temporalectomy with muscle flap rotation.

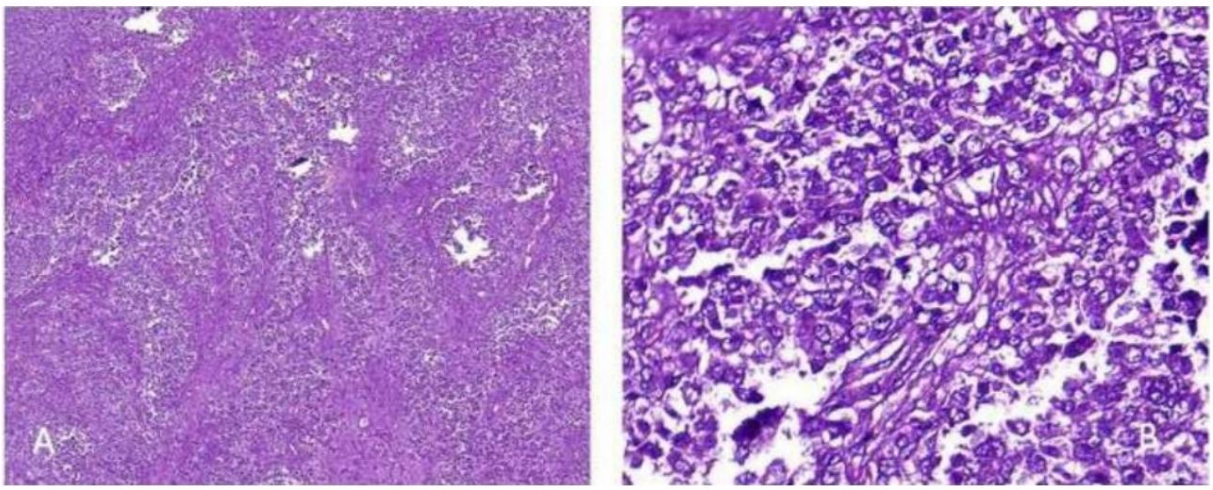
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temporal, and during surgery, it was observed that there was tumor invasion of the dura mater.
mater and posterior fossa.

The histopathological examination showed it to be a poorly differentiated neoplasm with a pattern of
Rounded or slightly elongated cells, with hyperchromasia and nuclear polymorphism.
eosinophilic cytoplasm, in desmoplastic stroma, with a focal area in which such
cells organize themselves into nests of uncohesive cells, with an alveolar pattern, containing
fibrous bands interspersed (Figure 3). Immunological examination suggested.
Histochemical analysis of lesion histogenesis revealed a pattern consistent with
Undifferentiated CD99-positive neoplasm, morphologically suggestive of sarcoma of
Ewing.

Figure 3: Histological sections stained with hematoxylin-eosin (HE). (A) Nests of poverty
cohesive cells with an alveolar pattern. (B) Poorly differentiated neoplasm with round or
scantily elongated cells, eosinophilic cytoplasm, desmoplastic stroma, hyperchromasia,
and nuclear polymorphism



After the histopathological report, the patient was referred to the Children's Hospital.
Albert Sabin (HIAS), aiming at the follow-up of her treatment. At HIAS, the patient
It maintains joint monitoring with pediatric oncology, oncogenetics, nutrition and
psychology. She underwent tests for tumor staging, including a bone marrow aspirate, which did not...
showed evidence of the presence of tumor cells; bone scintigraphy did not demonstrate
other areas with suspected increased osteogenic activity; neck CT scans,
thorax, upper abdomen and pelvis, which allowed visualization, however, of lesions

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osteolytic lesions in the right ilium and in the vertebral body of L1, which constituted lesions metastatic.

The patient has opted to begin treatment with radiotherapy and chemotherapy. She continues to be... Treatment at HIAS, with a good response to the established therapy.

3. Discussion

Although metastases of Ewing's sarcoma (ES) are generally common, there is only one Another case described in the literature involves metastatic lesions originating from a primary SE of skull, making the current report the second to demonstrate the existence of metastases to the skull. distance. In this way, there is a better prognosis when it comes to an injury. primary cranial injury, when compared to primary injuries in other parts of the body 3. Early diagnosis is of utmost importance, as the tumor exhibits growth. Extremely fast. The radiological examination is essential to help determine the To determine the extent of the disease, as well as to define appropriate therapeutic strategies. It is possible to use Computed Tomography (CT), which offers a better evaluation of the cortical bone, in addition to allowing detailed examination of the skull base; of Nuclear Magnetic Resonance (MRI), which provides adequate analysis of soft tissues. and enables the determination of invasion of the spinal cord, orbit, and intracranial region; and of Positron Emission Tomography (PET-CT) to investigate the existence of distant metastases 4.

Despite all the resources available, the diagnosis of SE is difficult to confirm. Histopathological analysis combined with immunohistochemistry is the gold standard . 5. Characteristics Histopathological findings include small, round, monomorphic cells with cytoplasm. Sparse, round nuclei and imperceptible nucleoli. Genetic molecular analysis. highlighting the chromosomal translocation on chromosome 22q12 is pathognomonic. Overexpression of CD99, a transmembrane protein encoded by the MIC-2 gene, is another useful marker detected by immunohistochemistry 6. Advances in multimodal therapy have considerably improved survival rates. For sarcomas in general. The treatment of intracranial SE had a good prognosis. relatively worse, when only surgery and radiotherapy were listed as options. treatment, with a survival rate of approximately 10% at 5 years. However, with the

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Introduction of chemotherapy combined with radiotherapy, the survival rate in
Over the past 5 years, it has increased significantly, reaching 53% 7. It is imperative to highlight the
importance of multidisciplinary team participation in monitoring and
treatment of this type of cancer, involving otolaryngology, head and nose surgery
neck surgery, neurosurgery, speech therapy, pediatrics, oncology, among other professionals.
health professionals, for the efficient management of case 4. Such teamwork allows for
Investigation from different perspectives, aiming for greater accuracy in management.
of the condition.

This is an extremely rare case of Ewing's sarcoma of the temporal bone with
distant metastases. The scarcity of similar reports reinforces the complexity and the
The exceptional nature of this clinical scenario is emphasized. Furthermore, the imperative need for a...
early diagnosis, based on radiological, histopathological and immunological examinations-
Histochemistry, which proves to be crucial for the viability of an effective treatment.
In this context, the use of a multidisciplinary therapeutic approach, encompassing
Surgical intervention, chemotherapy, and radiotherapy are of crucial importance.
The evolution of this case reinforces the need for constant vigilance and consideration of
possibility of remote dissemination, guiding the establishment of protocols
personalized treatment plans that aim to optimize clinical outcomes in various situations.
so unique and challenging.

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Required Declarations (to be filled in)

Ethical approval (CEP/CONEP or equivalent)

(X) Yes ( ) Not applicable

Opinion/Registration Number: CAEE No. 71314223.9.3001.5042

Informed Consent Form: ( ) Yes ( ) Not applicable

Study registration (when applicable): ( ) ReBEC ( ) ICTRP/WHO ( ) ClinicalTrials.gov ( ) Others: _____

Registration number: _____

Reporting guideline (EQUATOR): ( ) CONSORT ( ) STROBE ( ) PRISMA ( ) CARE ( ) COREQ/SRQR ( ) SQUIRE ( ) Other: _____

Data Sharing Statement (ICMJE)

Anonymized data: ( ) will be shared (X) will not be shared

What data: _____

Additional documents (protocol/plan/methods): _____

Availability period: _____

Access conditions: _____