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Case report
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A rare manifestation of Ewing sarcoma: Primary intracranial Ewing sarcoma

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Abstract

Primary intracranial Ewing sarcoma (ES) is an extremely rare condition, frequently presenting as supratentorial and intraparenchymal. This case report presents a 22-year-old male diagnosed with primary intracranial ES exhibiting dura, muscle, and bone invasion. The patient initially presented with swelling in the preauricular region and was operated on with a preliminary diagnosis of meningioma, but pathology confirmed primary intracranial ES. This case highlights the rarity of primary intracranial ES and provides significant contributions to the diagnostic process.

Keywords: Ewing sarcoma, primary intracranial Ewing sarcoma, dura invasion, muscle invasion, bone invasion

Introduction

Ewing sarcoma (ES) is a highly aggressive small round cell tumor that typically originates from bone and soft tissue, predominantly affecting children and adolescents [1]. The most common sites are the pelvis and extremities, with about 30% of cases involving soft tissues [2]. Primary intracranial ES is exceedingly rare, often presenting as supratentorial and intraparenchymal [3,4]. Clinically and radiologically, primary intracranial ES can mimic various other tumor types. This report describes a case initially operated on with a provisional diagnosis of meningioma, but later confirmed as primary intracranial ES through pathology. The purpose of this article is to present this rare case of primary intracranial ES, as reports in the literature are limited.

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Informed Consent
The authors stated that the written consent was obtained from the patient presented with images in the study.

Conflict of Interest
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Case presentation

A 22-year-old male with no known chronic illnesses presented to the otolaryngology clinic with swelling in the preauricular region following blunt head trauma two months prior. The patient was referred to our neurosurgery clinic after a brain computed tomography (CT) scan and magnetic resonance imaging (MRI). The patient's history revealed that the headache started post-trauma and the swelling in the preauricular region appeared a week later. A physical examination showed a firm, immobile swelling of approximately 4x4 cm in the left temporal muscles. The neurological examination was normal.

The brain CT and MRI revealed a parenchymal lesion and vasogenic edema, with mass involvement in the dura, temporal muscle, and bone (Figure 1, 2a, 2b). Considering these findings, meningioma was preliminarily diagnosed, and a Simpson grade 1 surgical excision was planned. The patient underwent gross total excision of the lesion, the invaded dura, bone, and temporal muscle. The dura was repaired with synthetic dura, and the bone defect was covered with a titanium mesh. Postoperative examination was consistent with the preoperative state, with no early or late surgical complications. The patient was discharged after the removal of sutures in the first postoperative week.

Pathology results indicated "surgical specimen CD99 diffuse positive, CD56 focal positive, synaptophysin focal positive; PAX5, CD20, CD45, TTF1, chromogranin negative". Diagnosis was Ewing sarcoma, central nervous system (CNS) Grade 4 [5]. A positron emission tomography (PET) was performed to investigate additional foci, with no other foci detected. The patient was diagnosed with primary central nervous system Ewing sarcoma based on the current pathology results.

The patient received focal radiotherapy from radiation oncology, and chemotherapy planning was done by medical oncology.

Figure 1: Brain CT shows a hyperdense nodular lesion in the left temporal pole with minimal surrounding edema.

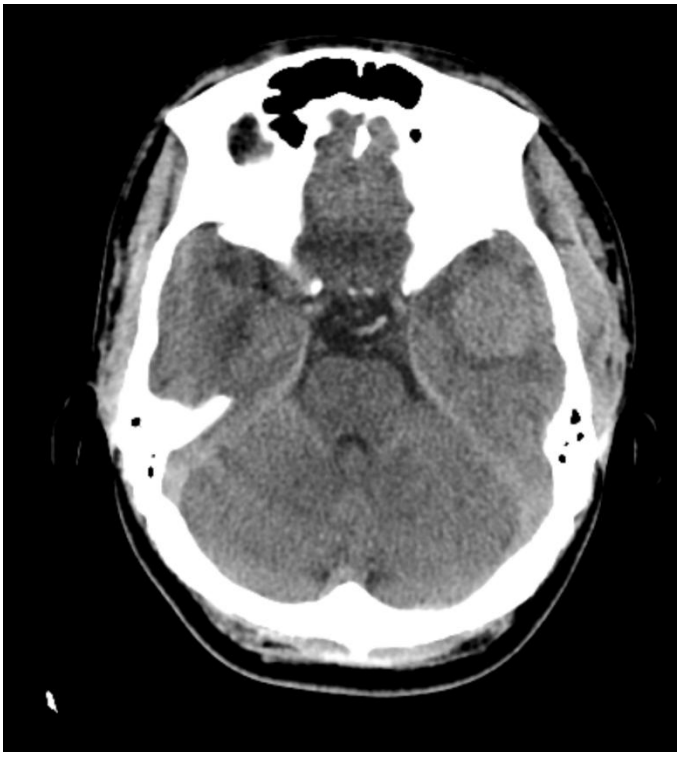


Figure 2a: MRI with contrast shows a parenchymal lesion with homogenous contrast enhancement and vasogenic edema, with mass involvement in the dura, temporal muscle, and bone.

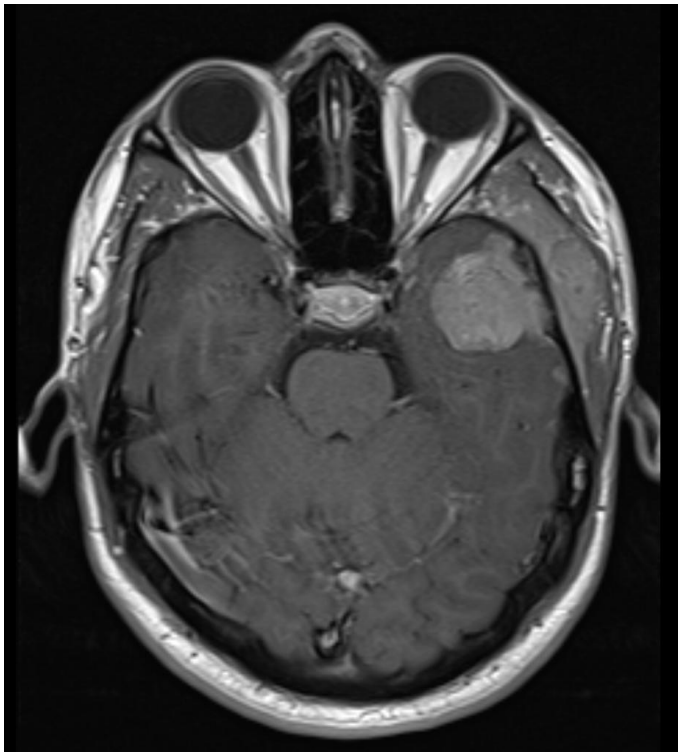
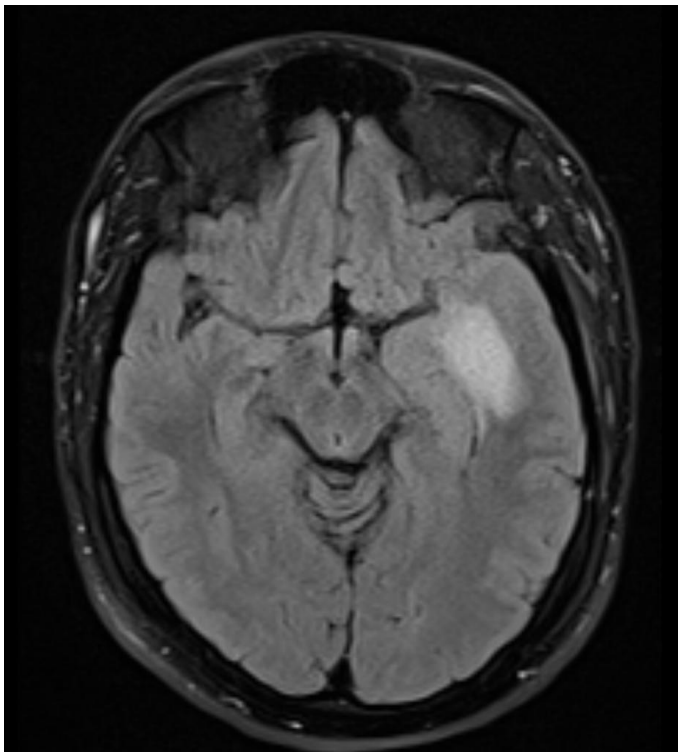


Figure 2b: MRI shows a vasogenic edema, with mass involvement in the dura, temporal muscle, and bone.



Discussion

ES is a malignant bone tumor characterized by round nuclei and primitive small cells [1]. High failure rates (>80%) in patients treated solely with local therapy before the advent of systemic chemotherapy suggest that most patients with ES have micrometastases at diagnosis [3]. Distant organ metastasis mainly occurs hematogenously, commonly affecting the lungs (38%), bones (31%), and bone marrow (11%), with the spine being a frequent site for bone metastasis [3,4,6]. CNS metastases of ES are rare, usually resulting from the growth of the sarcoma into the extradural space and meninges. The incidence of isolated central

nervous system ES ranges from 1.1% to 4.3% [3]. In a study by Paulus et al., only one out of 2500 patients operated on for intracranial mass was reported as ES [7].

Primary intracranial ES, constituting 0.03% of all intracranial tumors, is exceedingly rare [8]. It typically arises in bone or surrounding soft tissues, predominantly in supratentorial locations [9]. While ES mainly affects children and adolescents, it can also be seen in the second decade of life and is more common in males [10]. Our patient, consistent with the literature, was a male in his second decade.

All reported cases of primary intracranial ES in the literature originate from the dura, with the majority (70%) located in the cerebral hemispheres and two cases (20%) in the posterior fossa [11]. Kim et al. [11] reported on a rare case that originated from brain parenchyma in a 50-year-old patient with no known history of ES who presented with headaches unresponsive to analgesics. In our case, the absence of a clear boundary between the dura and tumor surface during surgery and the tumor's adherence to brain parenchyma suggested a parenchymal origin.

The most common clinical features of primary intracranial ES are seizures, headaches, vomiting, and other signs of increased intracranial pressure (ICP) [12]. However, reported cases also include symptoms such as hemiplegia, hearing loss, lethargy, fatigue, and ataxia, with an average symptom duration of 5.9 months [8]. Our patient presented with a headache, and the symptom duration was two months, shorter than reported in the literature. The trauma history and subsequent diagnosis were unique features of our case, although the lack of acute phase radiological imaging post-trauma precluded determining the trauma's role in tumor development.

Radiological findings in reported primary intracranial ES cases typically show mixed iso-hypointense signals on T1-weighted MRI and iso-hyperintense signals on T2-weighted MRI [8]. In Cherif et al.'s [9] study of 48 primary intracranial ES cases, contrast-enhanced MRIs showed heterogeneous enhancement in approximately 40% of cases, dense enhancement in 52.5%, and moderate enhancement in 7.5%. Our findings were consistent, showing iso-hypointense signals on T1-weighted MRI, iso-hyperintense signals on T2-weighted MRI, and dense contrast enhancement on contrast-enhanced MRI.

ES is a highly aggressive tumor with focal necrosis, composed primarily of small, round, or oval undifferentiated cells with hyperchromatic nuclei, increased mitotic activity, and slightly basophilic cytoplasm [8]. Tumor cells are also notably fibrotic, highly mitotic, and separated into cell groups by collagen bands. CD99 expression is a highly reliable and sensitive diagnostic biomarker for primary intracranial ES, detected in nearly all reported cases [9]. However, it is not specific to ES, as it can also be found in other small, blue round cell tumors, such as lymphoblastic lymphomas, ependymomas, and rhabdomyosarcomas [8,9]. In our case, CD99 was diffusely positive, CD56 and synaptophysin were focally positive, and PAX5, CD20, CD45, TTF1, and chromogranin were negative, consistent with other cases in the literature.

Primary intracranial ES is predominantly described as a dura-based tumor [11]. Radiological diagnosis is challenging, as it can be confused with other extra-axial tumors, such as meningiomas, hemangiopericytomas, solitary fibrous tumors, and leiomyomas [6]. Primary intracranial ES often mimics meningioma radiologically, showing dura, bone, and muscle invasion and appearing as a well-defined, homogeneously enhancing solid mass [9]. Similar to other cases, our patient was initially diagnosed with meningioma and underwent Simpson grade 1 surgical excision.

The aggressive behavior of locally situated ES and its early onset reduces survival and necessitates combined therapy. Although there is no standard treatment approach for these malignancies, gross total surgical excision remains the cornerstone of treatment [9,10]. In addition to surgery, chemotherapy and radiotherapy are also treatment options [3]. Standard chemotherapeutic agents include vincristine, etoposide, doxorubicin, and ifosfamide [9,10]. Our patient received focal radiotherapy following gross total surgical excision, and chemotherapy planning was done during follow-up by medical oncology.

Conclusion

This report presents a rare primary intracranial ES case diagnosed post-trauma, highlighting its rarity and potential for misdiagnosis with other tumor types. Primary intracranial ES should be considered in the differential diagnosis of various tumors, particularly in patients in their second decade of life. Intraoperative and histopathological findings are critical for confirming the diagnosis. A PET scan should always follow postoperative pathological diagnosis to assess metastasis.

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