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Altered Consciousness as the Presenting Symptom of Temporal Bone Ewing's Sarcoma: A Rare Case Report

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ABSTRACT

Ewing's sarcoma is a rare, aggressive small round cell malignancy of neuroectodermal origin that can involve both bone and soft tissues, with cranial involvement being uncommon. We report a case of a 22-year-old male presenting with altered consciousness, left parotid swelling, and purulent otorrhea. Imaging revealed an aggressive parotido-temporo-occipital mass with intracranial extension causing triventricular hydrocephalus. Histopathology and immunohistochemistry (CD99 positivity) confirmed stage III Ewing's sarcoma centered on the temporal bone. The patient received neoadjuvant chemotherapy using the VAC protocol, achieving minimal tumor regression. Cranial Ewing's sarcoma poses diagnostic and therapeutic challenges due to its rarity and proximity to neurovascular structures. Early recognition, accurate diagnosis through imaging and immunohistochemistry, and multidisciplinary management—including chemotherapy, surgery, and radiotherapy—are essential to optimize outcomes. Prognosis depends on tumor size, patient age, presence of metastases, and response to treatment. Clinicians should consider Ewing's sarcoma in young patients presenting with cranial bone masses and neurological symptoms.

KEYWORDS

Ewing sarcoma, temporal bone neoplasms, altered consciousness, young adult, case report

MAIN ARTICLE

INTRODUCTION

Ewing sarcoma is a highly aggressive malignant small round cell tumor of neuroectodermal origin, which can involve both osseous and extra-osseous structures [1]. Osseous forms are more frequent and typically involve the pelvis, axial skeleton, and long bones, whereas extra-osseous forms (1.1% of cases) primarily affect the soft tissues of the lower extremities, paravertebral region, thoracic wall, and retroperitoneum [1,2,3,4]. Cervicofacial involvement represents only 1–4% of Ewing sarcomas [1,3,4]. Primary cranial Ewing sarcoma is rare [4,5] and can involve multiple cranial bones, with a predilection for the temporal bone, as seen in our case. Diagnosis relies primarily on histopathological and immunohistochemical evaluation. Management is multidisciplinary, combining surgery, chemotherapy, and radiotherapy [4]. We report a case of a 22-year-old patient with a primary temporal bone Ewing sarcoma, revealed by a consciousness disorder.

PATIENTS AND OBSERVATION

Patient Information

A 22-year-old male with a history of chronic cannabis use presented to the emergency department for altered consciousness and progressive general health deterioration. He had no known comorbidities and no significant family medical history. Physical examination revealed a painless, gradually enlarging left parotid mass associated with purulent otorrhea. Neurological evaluation showed impaired consciousness but no focal motor deficits. There were no signs of fever or meningeal irritation.

Diagnostic Assessment

Initial cerebral CT demonstrated a large, aggressive left parotido-temporo-occipital mass extending to the ipsilateral occipital and cerebellar lobes, causing active triventricular hydrocephalus with tonsillar herniation (Figure 1).

A biopsy revealed hemorrhagic fragments of an undifferentiated malignant tumor. Immunohistochemistry confirmed the diagnosis of **stage III Ewing sarcoma centered on the petrous portion of the temporal bone**.

Pre-therapeutic cervical MRI showed a large heterogeneous tumor containing calcifications,

causing bone lysis and encasing the left internal carotid artery, internal jugular vein, and facial nerve, with intracranial, meningeal, and endocanal extension leading to cerebellar and tonsillar

DISCUSSION

Ewing sarcoma is the second most frequent malignant bone tumor in children and adolescents, accounting for only 1% of pediatric cancers [4]. It is a small round cell tumor of neuroectodermal origin, with both osseous and extra-osseous forms. Osseous Ewing sarcoma primarily affects long and flat bones, the pubis, ribs, and vertebrae, with a male-to-female ratio of 1.6:1 [1,4]. Extra-osseous Ewing sarcoma primarily affects males aged 15–30, with high aggressiveness and recurrence rates [1,3,4,5]. Cranial involvement is rare, representing 1–9% of cases [4,6]. The temporal bone is the most common site, followed by frontal, occipital, parietal bones, and less frequently the ethmoid and sphenoid bones [4,5].

Symptoms in cranial Ewing sarcoma are diverse, including headaches, localized swelling, signs of intracranial hypertension, and sometimes vertigo. Temporal bone involvement may present with facial paralysis, hearing loss, and otorrhea, occasionally accompanied by nausea and vomiting [4,5]. Our patient had vertigo, signs of intracranial hypertension, and purulent otorrhea. Early diagnosis is crucial due to the high aggressiveness and metastatic potential [4,5]. Imaging is essential for tumor assessment and monitoring. CT typically shows a lytic, isodense mass with heterogeneous enhancement. Cranial lesions often exhibit a “honeycomb” pattern, whereas periosteal reactions are rare in cervicofacial locations [4]. MRI shows heterogeneous intermediate T1 and T2 signals with heterogeneous enhancement, sometimes with calcifications, as in our case. Definitive diagnosis relies on histopathological evaluation of biopsy samples and immunohistochemistry. Classic histology shows small round cells in solid sheets, scant cytoplasm, prominent nuclei, mitoses, and bone formation [1,4,5,6]. Immunohistochemistry highlights CD99/MIC2, a sensitive and specific marker for Ewing sarcoma [1,4,5]. Cytogenetically, Ewing sarcoma is characterized by t(11;22)(q24;q12) translocation [4,5]. Differential diagnoses include meningioma, neuroblastoma, rhabdomyosarcoma, and lymphoma, distinguished by the absence of markers such as desmin, synaptophysin, and leukocyte antigen expression [1,3,4,5].

Management is multidisciplinary, combining surgery, chemotherapy, and radiotherapy [1,3,4,5]. Radical excision is critical. Neoadjuvant chemotherapy reduces tumor volume and

micrometastases, while adjuvant chemotherapy improves survival, with rapid responses to vincristine, cyclophosphamide, actinomycin D, and doxorubicin [1,3,4,5]. Our patient received VAC chemotherapy. Radiotherapy is indicated for inaccessible or inoperable tumors, with a total dose of 40–50 Gy, especially in patients with poor chemotherapy response [3,4,5,6]. For cranial tumors, surgical procedures may aim to relieve complications, such as ventriculoperitoneal shunting to treat hydrocephalus, as in our patient [5,7]. Despite multidisciplinary management, prognosis remains challenging due to aggressiveness and metastatic potential. Temporal bone Ewing sarcoma metastases are rare [4,8]. Poor prognostic factors include age >15 years at diagnosis, tumor size >10 cm, and poor chemotherapy response, as observed in our patient. Favorable factors include absence of metastases and complete surgical excision with clear margins. Early diagnosis and treatment before metastasis are essential for long-term survival [1,9].

CONCLUSION

This case illustrates a rare and aggressive presentation of temporal bone Ewing sarcoma with extensive intracranial extension and poor response to neoadjuvant chemotherapy. It underscores the need for clinicians to promptly investigate atypical skull base lesions, especially when accompanied by neurological deterioration and otologic symptoms. Early suspicion, timely imaging, and rapid histopathological confirmation are essential to guide appropriate multidisciplinary management. This case reinforces the crucial message that even subtle or nonspecific cranial symptoms may conceal highly aggressive pathology, and that early recognition remains key to improving patient outcomes.

FIGURES:

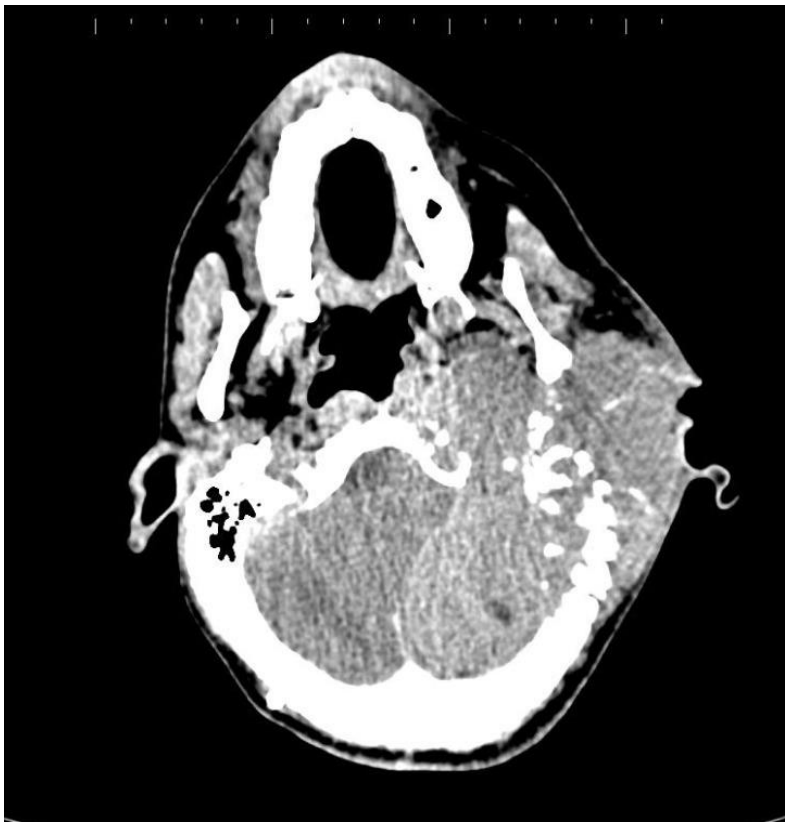

Figure 1: *Axial CT scan demonstrating a lytic tumor lesion of the left mastoid.*

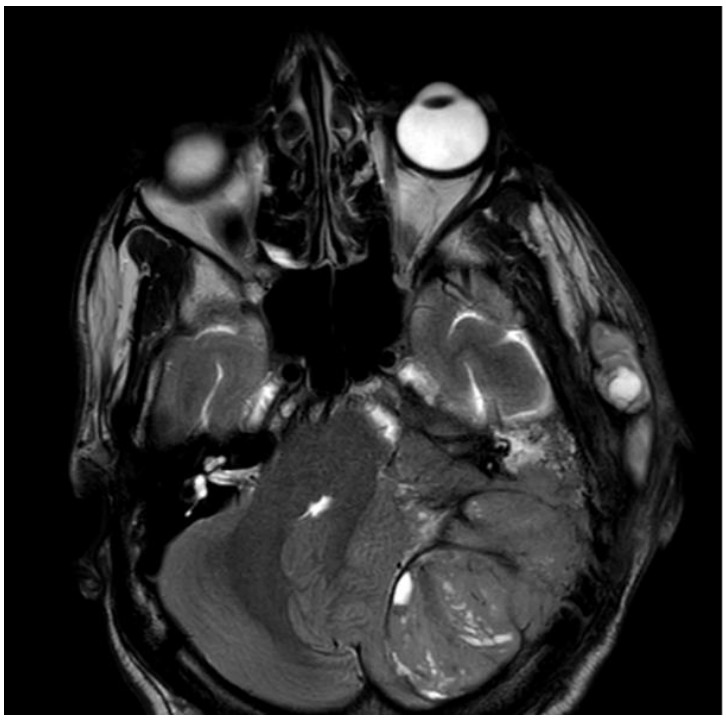

Figure 2: *Axial T2-weighted MRI showing a tumor process centered on the left mastoid, invading the internal auditory canal.*

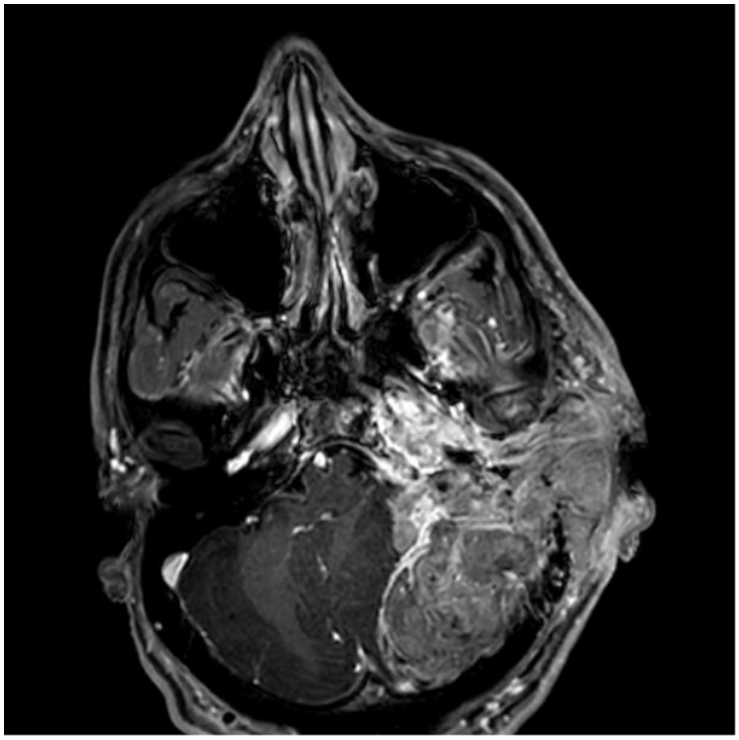

Figure 3: *Enhanced MRI sequence showing a tumor process compatible with Ewing sarcoma.*

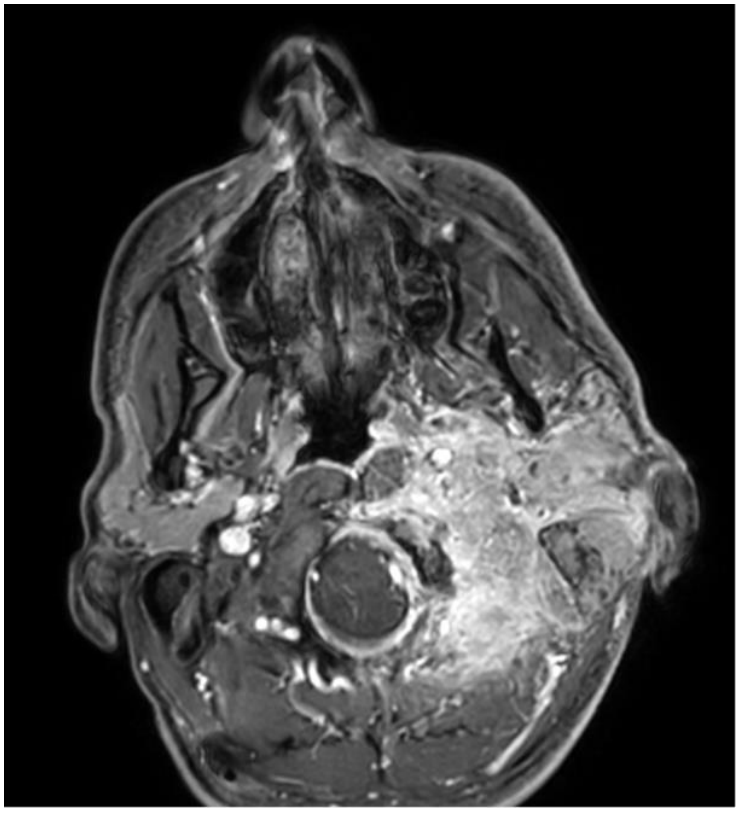

Figure 4: *Enhanced MRI sequence showing a tumor process compatible with Ewing sarcoma.*

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