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Uncommon territory: Primary intracranial Ewing's sarcoma in an adult patient

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Keywords
Ewing's sarcoma, primary intracranial tumour, intra-axial, extra-axial, adult brain tumour, rare neoplasm

Abstract

Introduction: Ewing's sarcoma is a rare malignant tumour predominantly affecting children and adolescents, typically originating in bone or soft tissue. Primary intracranial involvement is extremely rare, especially in adults. Most reported intracranial cases are dural-based and extra-axial, while intra-axial or mixed presentations are exceptionally rare.

Case report: We report a unique case of a 49-year-old male who presented with a generalised seizure and postictal dysphasia. Imaging revealed a large cystic (intra-axial) tumour with a mural solid component (extra-axial part) in the left temporoparietal region. Gross total resection was performed without complications. Histopathological and molecular analyses confirmed the diagnosis of primary intracranial Ewing's sarcoma. Despite recommendations, the patient refused further oncological treatment and was lost to follow-up after three months, during which no recurrence was detected on imaging.

Conclusion: This case illustrates a rare adult presentation of primary intracranial Ewing's sarcoma with mixed intra- and extra-axial features, highlighting the importance of considering rare entities in the differential diagnosis of atypical brain lesions. It also raises the possibility of post-surgical chronic inflammation as a potential factor in tumour pathogenesis, warranting further investigation.

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Introduction

Ewing's sarcoma is a rare malignant tumor that typically arises in the bones or soft tissues of children and adolescents, with primary intracranial involvement being extremely uncommon. Intracranial Ewing's sarcomas are usually extra-axial and dural-based, while intra-axial presentations are exceedingly rare, especially in adult patients. Due to their rarity and nonspecific clinical presentation, these tumors

often pose a diagnostic challenge and may be mistaken for more common intracranial pathologies (1).

We present a rare case of primary intracranial Ewing's sarcoma in a 49-year-old adult male, with unusual mixed intra-axial and extra-axial features.

CASE REPORT

A 49-year-old male presented to our emergency department following a generalized epileptic seizure, accompanied by postictal motor dysphasia. Initial non-contrast CT of the brain revealed a cystic expansive lesion located in the left temporoparietal region, surrounded by significant perifocal edema and signs of impending brain herniation due to displacement of midline structures (Figure 1). The patient was admitted for further diagnostic workup and treatment.

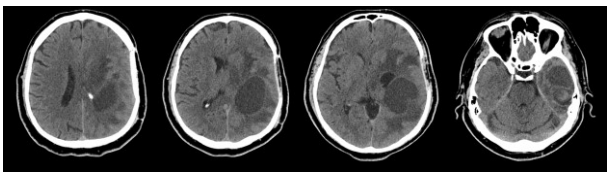

Figure 1. Initial non-contrast brain CT demonstrating a large cystic lesion in the left temporoparietal region surrounded by significant perifocal edema and signs of impending brain herniation.

Upon detailed history-taking, it was revealed that the patient had experienced a similar seizure episode approximately four months earlier, which resulted in hospitalization at another medical facility. A brain CT scan performed during that admission showed postoperative changes consistent with a prior left-sided temporoparietal craniotomy, along with encephalomalacic changes in the same region. No mass lesion or tumor was noted at that time (Figure 2). The patient reported having undergone three neurosurgical procedures in rapid succession around the age of 25, allegedly due to intracranial hemorrhage. However, he was unable to provide medical documentation or clear details regarding those events.

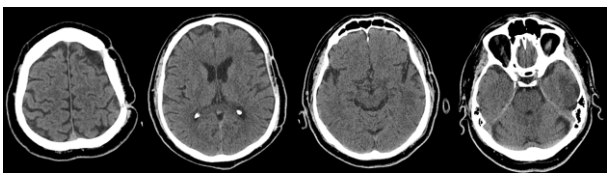

Figure 2. Non-contrast brain CT performed four months prior to admission to our hospital, following a seizure episode. The scan shows postoperative changes consistent with a prior left temporoparietal craniotomy, including encephalomalacic alterations in the affected region. No clear space-occupying lesion or midline shift was identified at that time.

Further on, an brain MRI performed in our institution confirmed the presence of a cystic intra-axial tumor in the left temporoparietal region with a solid mural component adherent to the dura at the base of the left temporal lobe (Figure 3A and 3B). The patient underwent preoperative preparation and was subsequently operated on (Figure 3C).

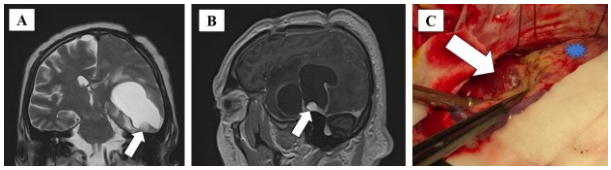

Figure 3. Preoperative brain MRI in T2-weighted coronal section (A) and T1-weighted sagittal section (B) demonstrates a cystic tumor in the left temporal region with a mural solid component, which is also visible in the intraoperative image (C). Arrows indicate the mural component.

Gross total resection of the lesion was achieved. Postoperative CT revealed no signs of residual tumor with resolution of edema and no signs of midline shift. Despite having undergone a total of four surgical procedures in the left temporoparietal region, there were no complications related to wound healing, and the patient was discharged on postoperative day six with a normal neurological exam and with no complaints.

Histopathological analysis revealed a rare diagnosis of primary intracranial Ewing's sarcoma. Due to the rarity of this entity, the sample was referred for verification to two independent neuropathology centers, both of which confirmed the diagnosis.

The patient was referred to an oncological tumor board for further management, but failed to attend. A follow-up call revealed that he had opted to decline further oncological treatment, instead choosing to pursue self-directed alternative therapy with herbal preparations and scorpion venom. He presented for one follow-up visit three months postoperatively, during which a control CT scan showed no signs of tumor recurrence. (Figure 4). He reiterated his

decision to refuse further treatment, and was subsequently lost to follow-up.

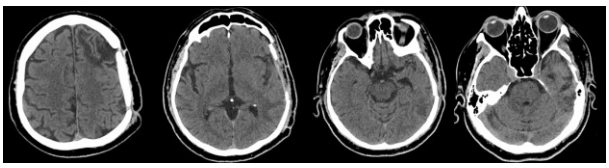

Figure 4. Follow-up brain CT three months after surgery showed no signs of tumor recurrence.

Discsussion

Ewing's sarcoma, also referred to as peripheral primitive neuroectodermal tumor (pPNET), represents the second most common primary bone malignancy in the pediatric and adolescent population, with an incidence of approximately 3 cases per million. It occurs more frequently in males and individuals of European ancestry. The tumor typically arises in the long bones, ribs, vertebrae, or pelvic bones (2). Only a small fraction of Ewing's sarcoma cases involve the cranial bones or brain parenchyma. Histopathologically, these tumors are classified as "small round blue cell" neoplasms of the central nervous system, characterized immunohistochemically by the presence of a distinct genetic alteration, the EWSR1-FLI1 fusion gene (3). In this report, we present a large cystic brain tumor with a mural solid component adherent to the dura mater, representing a rare mixed intra and extra-axial Ewing's sarcoma in an adult patient, thus expanding the spectrum of intracranial presentations of this rare malignancy.

Primary intracranial Ewing's sarcomas are extremely rare, particularly in adults. While the majority of these tumors arise in pediatric populations and typically involve the skull or dura mater, cases with parenchymal (intra-axial) involvement—either partially or exclusively—are exceptionally uncommon. In adults, fewer than 30 cases of intracranial Ewing's sarcoma have been reported in the literature, and only a small subset of these involved patients over the age of 30 (1, 4, 5). Our case is further distinguished by the tumor's dual nature, consisting of a large cystic intra-axial lesion with a mural solid component adherent to the dura (extra-axial part). Such mixed dural and intra-parenchymal presentations are rarely documented, especially in middle-aged patients, making this case a valuable addition to the limited data available on primary CNS Ewing's sarcomas in adults.

The clinical presentation of primary intracranial Ewing's sarcoma in adults is often nonspecific and highly variable, frequently mimicking other, more common intracranial pathologies. Reported symptoms may include seizures, headaches, focal neurological deficits, or signs of increased intracranial pressure—none of which are pathognomonic (6, 7). In our case, the patient presented with a generalized epileptic seizure and transient postictal dysphasia, symptoms that are not uncommon in a broad spectrum of intracranial lesions. What distinguishes this case, however, is the remarkably rapid tumor progression, strongly suggesting a highly malignant biological behavior. A brain CT scan performed approximately four months earlier—during a hospitalization at another facility—showed only subtle changes, which were interpreted as encephalomalacic sequelae from remote neurosurgical interventions in early adulthood. No mass lesion or space-occupying process was identified at that time. The subsequent development of a large cystic tumor with a solid mural component within such a short interval highlights both the aggressiveness and the diagnostic challenge posed by this rare entity in adult patients.

The pathogenesis of primary intracranial intra-axial Ewing's sarcoma remains poorly understood. One speculative but biologically plausible hypothesis in this case is that the tumor may have originated from dural mesenchymal progenitor or ectopic neuroectodermal cells, potentially activated or transformed through repeated surgical interventions and chronic dural irritation (8). The patient had undergone multiple neurosurgical procedures in early adulthood, and imaging prior to the most recent presentation showed encephalomalatic changes and signs of previous craniotomy, without evidence of a tumor at that time. It is conceivable that chronic inflammation, regenerative stimuli, and altered local immune surveillance in the context of prior surgical trauma could have contributed to neoplastic transformation in a susceptible cell population. Although this theory remains unproven, it is scientifically acceptable and compelling, and warrants further investigation in future studies.

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