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CASE REPORT

Primary intracranial extraosseous Ewing's sarcoma

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Abstract

Introduction Common sites of occurrence of extraosseous Ewing's sarcoma include the soft tissues and bones of the lower extremity, 12 paravertebral, and retroperitoneal regions. Primary intracranial Ewing's sarcoma/pPNET is usually intraparenchymal located 13 when supratentorially, and an extraaxial epidural tumor radiographically mimicking a meningioma is extremely rare.

Case Presentation A 20-year14 old male presented to the emergency department with a 1-day history of drowsiness, headache, and fever. Neurological exam15 ination revealed decreased muscle strength (4/5) in the left lower limb. Head computed tomography scan showed an epidural 16 space-occupying lesion in the right temporoparietal region, which was assumed to be a meningioma by radiographic criteria. However, the surgical specimen was diagnosed as Ewing's sarcoma.

Conclusion Primary intracranial extraosseous Ewing's sarcoma is a rare condition that may mimic a meningioma on imaging. Physicians must be cognizant of this possibility, particularly in any young individual with a solitary contrast-enhancing dural-based lesion.

Keywords Central nervous system · Brain neoplasm · Ewing's sarcoma · Epidural · Extraosseous · Molecular analysis

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Introduction

Ewing's sarcoma (ES)/peripheral primitive neuroectodermal tumor (pPNET) is a malignant small, round cell tumor that is the second most common bone tumor in children [1, 2]. The chromosomal translocation t(11;22)(q24;q12) is pathognomonic for Ewing's sarcoma family of tumors and is present in around 85% of cases [2, 3].

Ewing's sarcoma commonly arises in the cortex of long bones, ribs, and the vertebrae [3]. Central nervous system (CNS) ES mostly results from metastases from extracranial sites [2]. Primary intracranial ES is usually intraparenchymal; rarely, it may exist extradurally, mimicking a meningioma on radiology [2]. Due to the small number of reported cases of intracranial extraosseous ES, there is a lack of knowledge regarding its clinical presentation, prognostic factors, and disease course. This can lead to misdiagnosis by imaging (as central PNET, meningioma, epidural hematoma, etc.) [1, 3]. We report the case of a 20-year-old male with an intracranial epidural extraosseous ES mimicking a meningioma on radiology.

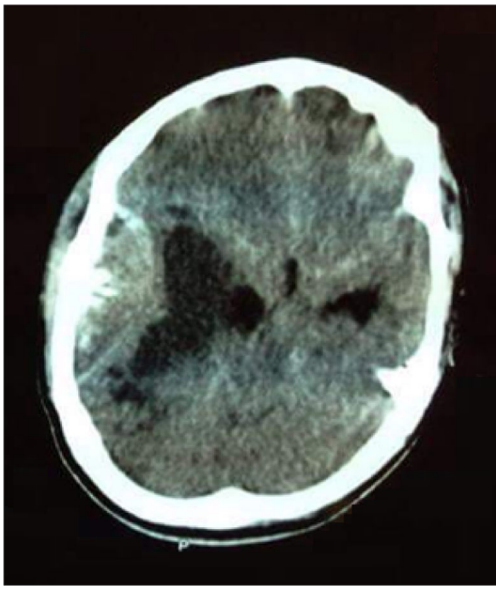

Fig. 1 Axial CT scan of the brain showing a large, well-defined lesion in the right temporoparietal region involving the squamous part of temporal bone, suggesting a meningioma. Significant mass effect and midline shift are also seen

Case presentation

A 20-year-old male presented to the emergency department with a 1-day history of drowsiness, headache, and fever. Past medical, drug, and family histories were unremarkable. General physical examination was unremarkable. Neurological examination showed motor weakness (4/5) in the left lower limb. Rest of examination and laboratory work-up were non-contributory. On head computed tomography (CT), an epidural space-occupying lesion was identified in the right temporoparietal region (Fig. 1), which was suspected to be a meningioma.

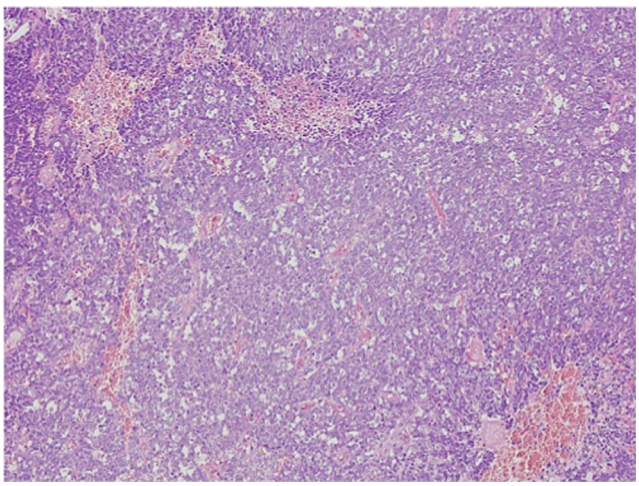

Fig. 2 Tumor composed of sheets of small cells with scant basophilic cytoplasm, and hyperchromatic nuclei with clumped chromatin. (H&E, × 40)

The patient underwent craniotomy and complete tumor excision. Intraoperatively, the tumor was arising from the dura and extended to the bone, with evidence of bone invasion. Post-operative recovery was unremarkable.

Histopathologic examination of the resected tissue showed the presence of bone and fibrocollagenous tissue exhibiting an infiltrating neoplastic lesion arranged in sheets, nests, and rosettes (Fig. 2). Neoplastic cells had pleomorphic hyperchromatic to vesicular nuclei, along with moderate to scant cytoplasm (Fig. 2). Prominent mitotic activity, necrosis, and vascular proliferation were also seen. Glycogen was positive on special stain (PAS+/−D). Immunohistochemistry reactivity pattern for the biopsy sample showed positive results for the markers CD99 (Mic-2), synaptophysin, and BCL-2 while the results for markers desmin, EMA, CD34, CD3 (Pan-T), CD20 (Pan-B), CD10, and TdT were negative.

A diagnosis of small round blue cell tumor was made with the morphological and immunohistochemical features pointing towards Ewing's sarcoma/pPNET. On fluorescent in situ hybridization (FISH), t(11;22)(q24;q12) was not detected. Detailed workup for metastasis did not yield any metastatic lesions.

Three months later, the patient presented again, with a headache and bilateral complete loss of vision. A right temporoparietal mass was seen on CT scan, and decompressive craniectomy and debulking of the tumor were

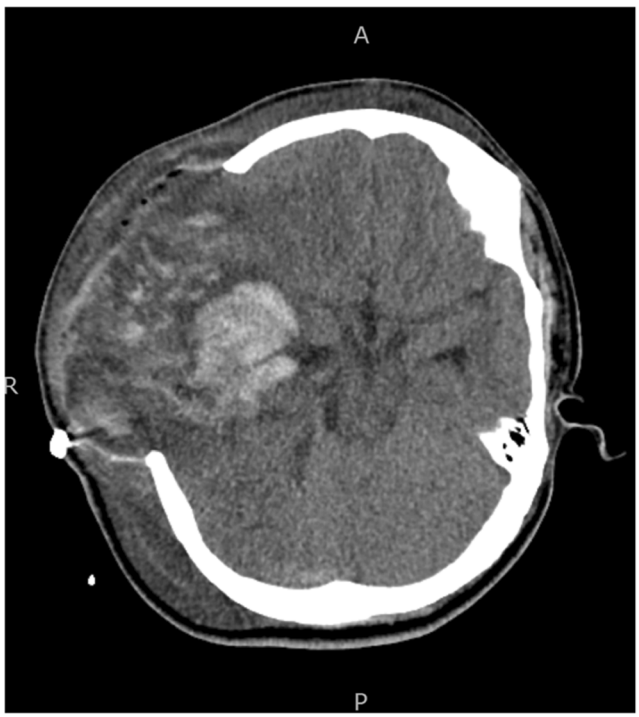

Fig. 3 CT axial section showing right-sided craniectomy defect with brain herniation through it and parenchymal bleed in right temporal lobe. Multiple contusion hemorrhages with cerebral edema, midline shift, and soft tissue scalp swelling right frontotemporal region, representing post-surgical changes are also seen

Table 1 Cases of intracranial extraosseous Ewing's sarcoma identified in literature review and the present case

Author (year of publication)	Age at presentation (years)	Sex	Radiological findings	Management	Prognosis
Present case	20	Male	Large, contrast-enhancing, meningioma-like mass in right temporoparietal region	Craniotomy and complete tumor excision	Local recurrence at 3 months. Craniotomy and debulking performed; radiochemotherapy recommended
Kumar et al. [2]	22	Male	Left frontal mass, contrast-enhancing, dural-based mimicking a meningioma	Surgical excision with chemotherapy and radiotherapy	Disease free
Tanboon et al. [15]	22	Female	Right frontal, dural-based mass	Craniotomy with gross total excision	Multiple extracranial metastases 2 months later; died 6 months after surgery
Gupta et al. [8]	11	Male	Well-defined lytic lesion in frontoparietal bone with subgaleal and epidural component	Excision of tumor with skull reconstruction	3-month follow-up, symptom free
Choudhry et al. [1]	11	Female	Left temporoparietal contrast-enhancing mass	Surgical excision, chemotherapy with vincristine, doxorubicin and cyclophosphamide, alternating with etoposide and ifosfamide, and radiotherapy	Disease free at 9-month follow-up
Mellai et al. [10]	56	Female	Right temporal cystic mass adhering to dura	Surgical resection	Disease free at 18 months
Mobley et al. [11]	21	Male	Dural-based mass in right occipital lobe mimicking a meningioma	Partial resection with unspecified "adjuvant therapy"	Recurrence treated with cyclophosamide, topotecan, and radiotherapy
Pekala et al. [12]	8	Female	Solid, contrast-enhancing mass with tentorial attachment	Complete gross surgical excision	Unknown
D'Antonio et al. [6]	50	Male	Right temporal dural-based mass	Surgical resection	Disease free at 1-year follow-up
Dedeurwaerdere et al. [7]	17	Male	Right frontal contrast-enhancing dural-based mass	Surgical resection, chemotherapy with intrathecal methotrexate, carboplatin and VP-16, and radiotherapy	Disease free at 8-year follow-up
Autunes et al. [5]	6	Male	Left frontal, contrast-enhancing, cystic mass	Surgical resection with chemotherapy; radiotherapy was recommended	Unknown
Katayama et al. [9]	5	Male	Large, contrast-enhancing, left tentorial mass in middle and posterior fossa	Surgical resection, and chemotherapy with intrathecal methotrexate	Disease free at 7-year follow-up
Papotti et al. [13]	30	Female	Multiple right frontal dural-based masses mimicking a meningioma	Total gross surgical excision, and chemotherapy with adriamycin, vincristine, and cyclophosphamide	Local and distant recurrence—treated with radiotherapy. Died 10 years after surgery
Stechschulte et al. [14]	25	Male	Mass in temporal fossa, associated with subdural hematoma	Surgical excision, chemotherapy with ifosfamide, adriamycin, cyclophosphamide, vincristine, and dactinomycin, and radiotherapy	

performed. Post-operatively, the patient developed left-sided hemiplegia. Head CT scan showed parenchymal bleeding in the right temporal lobe, with ventricular extension. A craniectomy defect with brain herniation, multiple contusion hemorrhages with perifocal edema, and soft tissue scalp swelling were also seen in the right frontotemporal region (Fig. 3). These findings represented post-surgical changes. Metastatic workup was negative for metastasis once again. The patient was referred for radiochemotherapy.

Discussion

Ewing's sarcoma has a male predominance, usually occurring in the first two decades of life. It has an incidence of one to three per million in the Western hemisphere [3, 4]. Extraosseous Ewing's sarcoma has an equal sex predilection [1].

Primary intracranial ES/pPNET is a recently recognized entity of CNS PNET with only 13 cases reported in the literature (Table 1) [1, 2, 5–15]. Although multifocal intracranial extraosseous ES has been reported in the medical literature, it mostly occurs as a solitary lesion associated with the dura [15]. Most of the reported cases (11 out of 13) occurred in the first 3 decades of life [2].

Intracranial extraosseous ES/pPNET can demonstrate two different patterns of meningeal involvement. One is diffuse involvement of the cranial and spinal leptomeninges while the other is a localized dural-based mass, mimicking a meningioma [2]. Only three cases of primary pPNET radiographically mimicking a meningioma have been reported previously [2, 11, 13]. While meningiomas are mostly benign neoplasms cured by surgical resection, ES/pPNET is an aggressive neoplasm requiring multimodality treatment [2]. Thus, cognizance of this pattern and a high degree of suspicion has immense diagnostic, therapeutic, and prognostic significance, particularly because early diagnosis is an important prognostic factor for intracranial extraosseous ES/pPNET [4].

Interestingly, the literature reveals that epidural extraosseous ES, presenting as a biconvex epidural lesion, may also be mistaken as an epidural hematoma [3, 4]. This is particularly true if the lesion is detected incidentally after trauma. Thus, epidural extraosseous ES is a possible diagnosis for biconvex epidural lesions, particularly if trauma is minor or multiple lesions are detected.

The t(11;22)(q24;q12) translocation, which is pathognomonic for Ewing's sarcoma, is present in over 95% cases of CNS extraosseous ES [4]. Surprisingly, this translocation was absent in our case, but the diagnosis could be reached based upon the morphological characteristics and immunohistochemical features. Thus, the absence of t(11;22)(q24;q12) alone does not preclude a diagnosis of intracranial extraosseous ES [4].

There is no well-defined therapeutic regimen for management of CNS-EES [2]. It is usually treated in a multimodal approach including surgery, chemotherapy, and focal radiotherapy [16]. For epidural extraosseous ES lesions, the treatment involves en bloc surgical excision followed by neoadjuvant systemic chemotherapy, with drugs such as vincristine, actinomycin-d, and cyclophosphamide (VAC) alternated with ifosfamide, cisplatin, and etoposide (ICE), and radiotherapy [2, 16]. A gross total resection, which is important for long-term survival, was possible in most (12 out of 13) reported cases of intracranial extraosseous ES/pPNET [2].

The 5- and 10-year survival rates for extraosseous Ewing's sarcoma are 69.7% and 65.2, respectively [15]. The prognostic factors influencing intracranial extraosseous ES have not been clearly identified yet [2]. For extraosseous Ewing's sarcoma in general, age at diagnosis, complete surgical excision, micrometastasis, and circulating tumor cells are the major prognostic factors [4]. Indicators of poor prognosis are large tumor volume (> 200 ml), atypical histology, metastatic lesions, loss of p16 expression, and gains of chromosomes 1q and 12 [15].

Conclusion

Primary intracranial extraosseous Ewing's sarcoma is a rare condition that may present as an epidural mass mimicking a meningioma. Physicians must be cognizant of the possibility of this rare tumor, particularly in any young individual with a solitary contrast-enhancing dural-based lesion that appears aggressive on imaging.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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