Intraosseous meningioma, a rare presentation of a common brain tumor: illustrative case

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BACKGROUND The subset of extradural meningiomas arising from bone is called primary intraosseous meningioma. The present article outlines the clinical presentation, investigation, surgical management, and clinical outcomes of a challenging case of intraosseous meningioma.

OBSERVATIONS A 27-year-old male presented with new-onset seizure of 3 years’ duration and growing painless hard swelling involving the left frontoparietal bone, the medial aspect of the left orbit, the nasal bone, and the temporal bone. Brain computed tomography showed a diffuse intradiploic space osseous lesion with a ground-glass appearance. The patient underwent bifrontal craniotomy and tumor excision. The histopathological sections showed the presence of classic whorls of meningothelial cells around reactive bony tissue suggestive of intraosseous meningioma. Postoperatively, the patient was neurologically intact, and he was discharged in a stable condition. The outcome of this case was satisfactory.

LESSONS The treatment of primary intraosseous meningioma is wide surgical excision together with the involved dura and any intradural tumor. The location of the tumor determines the approach and the amount of resection, keeping in mind preservation of function. The use of adjuvant therapy is reserved for cases of tumor recurrence or an atypical type.

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Meningioma, also known as meningéal tumor, is a benign tumor that is typically slow growing.1,2 It originates from the arachnoid cap cells from a highly metabolic active subtype that is involved in cerebrospinal fluid (CSF) resorption.1,2 It is estimated to account for between 13% and 26% of all intracranial tumors, whereas extradural meningiomas constitute from 1% to 2% of all meningiomas.2

Intraosseous meningioma is a subgroup of meningioma that arises in the skull. It can occur at any location of the skull, with the orbital cavity and frontoparietal skull being the most common locations.3 The clinical presentation depends on the site of the lesion, generally in the form of slowly growing painless hard swelling of the skull not causing neurological signs or symptoms until it reaches a huge size and causes mass effect.3,4 The incidence in both sexes is the same.3 The tumors are solitary and can be seen on computed tomography (CT) scans as hyperostosis of the bones, and biopsy can confirm the diagnosis.2–4

Here, we present a case of a 27-year-old male with a diagnosis of intraosseous meningioma. The clinical presentation, investigation, surgical management, and clinical outcomes are described. The challenges in our case are that it involves the whole of the frontal bones and extends to the facial bones, which are very challenging in reconstruction and cosmetic outcome. The case also involved the frontal sinus and skull base, as well as the long segment of the superior sagittal sinus (SSS).

Illustrative Case

Clinical Presentation

A 27-year-old male presented with new-onset seizure of 3 years’ duration associated with slowly growing painless forehead swelling.
The swelling started gradually from the parietal bone, then extended to the left frontal bone and around the left eye. In the past 3 years, the patient had developed recurrent attacks of seizures (1–2 times per month) with upward rolling eyes, lasting approximately 10 minutes. Clinical examination showed a 10 × 4 × 3-cm forehead swelling that involved the parietal bone and extended to the left frontal bone and around the left eye. The swelling was hard and not mobile with no changes in the overlying skin. The findings of the patient’s neurological and eye examinations were unremarkable.

Radiological Features

Brain CT showed a diffuse osseous lesion within the intradiploic space. It involved mainly the left frontal bone, the medial aspect of the right frontal bone, the nasal bone and glabella, and the anterior aspect of the left parietal bone as well as the left temporal bone with a ground-glass appearance (Fig. 1A–D). A CT venogram showed a patent SSS (Fig. 1E). A brain magnetic resonance imaging (MRI) scan showed a left frontal bony nonenhancing lesion with multiple intradural variably sized dural-based lesions demonstrating low T1 and intermediate T2 signals with avid enhancement, involving the right frontal parafalcine and left frontal convexity (Fig. 1F and G).

Surgical Intervention

The patient underwent bifrontal craniotomy and tumor excision. The steps of surgery were almost similar to those described in a previous report. A bicoronal skin incision was made almost 4 cm behind the palpable margin of the bony lesion, and a donut-shaped craniotomy was performed around the lesion. The dura and the anterior third of the SSS were very adherent to the inner table of the frontal bone and came out with bone (Fig. 2A–E). A small convexity meningioma in the left temporal lobe was excised. Most of the posterior bony lesion was removed and dissected from the posterior half of the SSS, which was kept intact. The thick forehead bone above the eyebrows and nasal bridge was thinned out and shaved carefully (to avoid opening the frontal air sinus and nasal cavity) using an oscillating saw and drill to contour with the outer table of the skull. The bone was very hard and marble-like. The dural defect was covered with dural substitute, and the bone defect was reconstructed with titanium mesh, contoured and fixed with screws to the inner table of the bone flap, and covered with hydroxyapatite bone cement. The redundant skin was not excised and was left to shrink with time (Fig. 2F–H). The patient tolerated the surgery well with no complications.
Histopathological Features

The histopathological sections of the lesion were composed of reactive cortical and trabecular bone with a focus of meningioma exhibiting classic whorls of meningothelial cells (Fig. 3).

Outcome and Follow-Up

Postoperatively, the patient was doing well and neurologically intact. His postoperative brain CT scan showed complete resection of the tumor (Fig. 4). The patient was discharged in a stable condition (hospital stay 13 days).

Discussion

Observations

Accounting for 36% of primary brain tumors, meningiomas are considered the most common primary central nervous system tumor. Most meningiomas are located in the subdural space. Only 1%–2% of meningiomas are considered extradural, arising from locations other than the dura mater. The subset of extradural meningioma arising from bone is called primary intraosseous meningioma.

Primary intraosseous meningiomas represent two-thirds of extradural meningiomas. The presentation of these tumors usually is less aggressive than intradural meningiomas. Furthermore, the location and size of the tumor are very important factors in this regard. Painless scalp mass is the most common presentation in cases of convexity intraosseous meningioma. In cases of skull base intraosseous meningioma, more neurological symptoms are expected due to cranial nerve defects being very common. Headache is reported to be the second most common symptom in both types. In the present case, the patient presented with a painless scalp mass. Our case presented with disfigurement (Fig. 5) and epilepsy.

Primary intraosseous meningiomas can exhibit either osteoblastic or osteoclastic behavior. On a CT scan, the osteoblastic type shows focal hyperdense lesions with prominent expansion of the skull, whereas the osteolytic type differs in that thinning of the bone is predominant with similar hyperdense lesions. Both types have a similar appearance on brain MRI, being hypointense on T1-weighted images and hyperintense on T2-weighted images. In the present case, a brain CT scan showed a ground-glass appearance, and brain MRI showed bony nonenhancing lesions.

The importance of the location of the tumor is not only in the presentation but also for guiding the surgical approach and the extent of resection. The location of the tumor in our case was surgically challenging regarding the functional (nose, paranasal sinuses, and eye) and cosmetic (forehead) outcomes.

The most common location of this type of tumor is the frontal bone followed by the parietal bone. In the present case, the tumor was in the frontoparietal region. At the University Medical Center Hamburg-Eppendorf, Butscheidt et al. reported that a majority of the cases were located in the sphenoid bone with predominant visual symptoms in their study.

The best treatment modality in cases of intraosseous meningiomas is wide surgical excision. Our surgical plane was created to dissect the SSS from bony tumor and excise the tumor completely, then reconstruct the dura and bone defect. Intraoperatively, we could not identify or dissect the dura and anterior one-third of the SSS from the bony tumor, because it was en bloc as one piece. To preserve the dura of the skull base and to avoid opening the paranasal sinuses with its sequelae of CSF leak and meningitis, we decided to shave the bony tumor and preserve the inner table of the forehead bone with the adherent dura of the skull base. This, of course, made the patient’s postoperative recovery smooth, short, and uneventful, and he was discharged home after 12 days. The need for excision of the dura in every case of intraosseous meningiomas is still not clear, given that Bassiouni et al. reported that 11 of 11 cases showed microscopic evidence of dural invasion even with no dural enhancement on MRI.

The follow-up plan was to administer radiotherapy if the tumor regrew or the histopathology showed atypical or anaplastic meningioma; the use of adjuvant therapy, including radiotherapy, chemotherapy, or bisphosphonate therapy, could be added if complete surgical excision could not be attained.

The rate of recurrence depends on multiple factors, including tumor grade; tumor location; radiological type, with osteolytic being worse; and extent of surgical excision.
Lessons
The treatment of primary intraosseous meningiomas is wide surgical excision together with the involved dura and any intradural tumor. The location of the tumor determines the approach and the amount of resection, keeping in mind preservation of function. The use of adjuvant therapy is reserved for cases of tumor recurrence after incomplete resection or cases in which histology reveals atypical meningioma.

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Disclosures
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