Radiology Case Reports

Volume 5, Issue 1, 2010

Intraosseous meningioma of the sphenoid bone

Syed Mansoor Hussaini, MD, MPH; Kristine Dziurzynski, MD, MSPH; Jonathan D. Fratkin, MD; J. Randall Jordan, MD: Syed Afzal Hussain, MD; and Majid Khan, MD

A 50-year-old female presented to the Neurosurgery clinic with dimness of vision and proptosis of her right eye. Maxillofacial CT showed a hyperostotic mass involving the right sphenoid ridge, anterior clinoid process, orbital roof, and lateral wall with mass effect on the intraorbital contents and lateral wall of the sphenoid sinus. MRI of the brain and orbit showed a heterogeneous enhancement of underlying dura and right orbital apex extending into the cavernous sinus. The patient underwent a staged resection in which pathological analysis showed an intraosseous meningioma. When a hyperostotic mass of the skull is encountered, meningioma should be considered in the differential diagnosis. Although primary intraosseous meningiomas are rare benign tumors, they can be associated with morbidity secondary to mass effect.

Case report

A 50-year-old female with history of hypertension and right eye injury three years ago presented with a several months' history of "dimming" of her vision in the injured eye. On examination, there was no relative afferent pupillary defect, and visual acuity was 20/20 OS and 20/25 OD, but she could only achieve a result of 1/14 on color chart tests. A fundus exam showed temporal disc pallor, on the right more than the left. There was no motor, sensory, coordination, or gait deficit, or any asymmetry in reflexes. Lab results showed her to be slightly anemic but were otherwise unremarkable.

An MRI brain scan with orbit protocol, with and without gadolinium, was obtained. It demonstrated heterogeneous enhancement involving the right orbital apex, extending to the right cavernous sinus, with linear thick nodular enhancement involving the dura overlying the right temporoparietal lobes. Precontrast T-weighted images demonstrated bony expansion with predominantly low marrow signal involving the right sphenoid wing and right lateral orbital wall. On T2-weighted images, these demonstrated slightly hyperintense signal (Fig. 1).

There was a moderate mass effect on the right orbital wall with thickening of the right lateral rectus. No optic nerve enhancement was seen. Noncontrast maxillofacial CT images were obtained for pre-operative planning and showed a 38.5-mm hyperostotic calvarial mass, primarily involving the right sphenoid wing, with involvement of the right lateral orbital wall, the anterior clinoid process, and the lateral wall of right sphenoid hemisinus (Fig. 2).

There was also demonstrated linear adjacent dural calcification/ossification with mass effect on the right lateral rectus, which was bowed medially. The optic canal and the superior and inferior orbital fissures were unremarkable, with a normal-appearing globe and its contents. Following the surgery, a maxillofacial CT image without contrast showed postoperative changes with partial resection of the hyperostotic bony mass.

The patient underwent a staged resection of the mass, optic nerve decompression, and tumor debulking with orbital reconstruction, respectively (Fig. 3). More than 90% of the tumor was removed. All of the grossly visible tumor was resected except the optic strut, the lateral wall of the sphenoid sinus, a portion of the greater sphenoid wing comprising the inferior lateral wall of the superior orbital fissure and posterior lateral wall of the inferior orbital fissure, and an eggshell remnant of the anterior clinoid process overly-

Citation: Hussaini SM, Dziurzynski K, Fratkin JD, Jordan JR, Hussain SA, Khan M. Intraosseous meningioma of the sphenoid bone. *Radiology Case Reports*. [Online] 2010;5:357.

Dr. Hussaini is a Neurology Resident in the Department of Neurosurgery, Dr. Dziurznski is in the Department of Neurosurgery, Dr. Fratkin is a Professor of Pathology, Dr. Jordan is a Professor of Otolaryngology, Dr. Hussain is a research assistant in the Department of Psychiatry, and Dr. Khan is an Assistant Professor of Radiology, all at the University of Mississippi Medical Center, Jackson MS.

Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rcr.v5i1.357

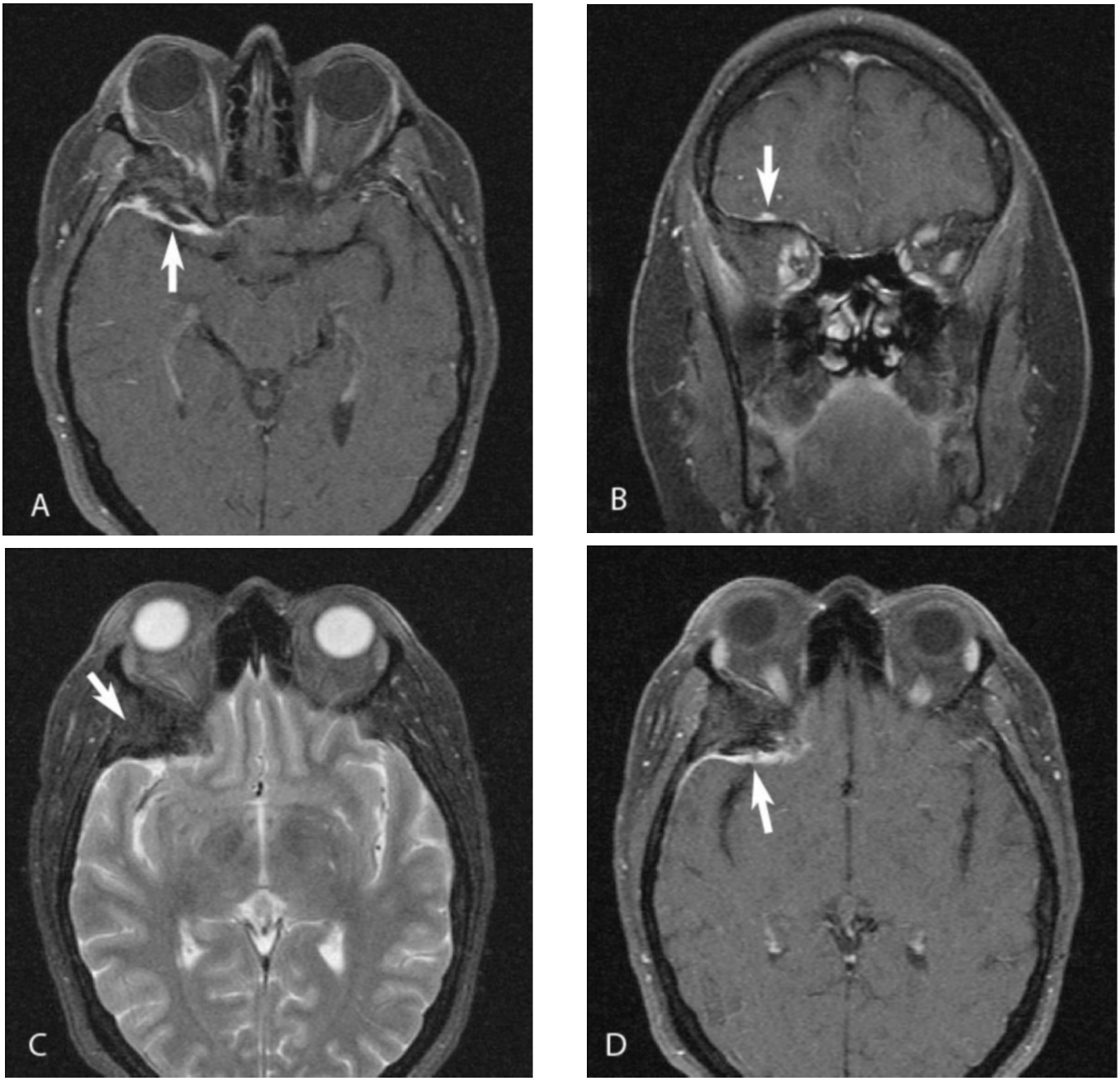

Figure 1. 50-year-old female with intraosseous meningioma. A (axial), B (coronal), and C (axial) T1-weighted fat saturation postgadolinium with fat saturation; D, axial T2-weighted image. These images demonstrate calvarial thickening involving right sphenoid wing and lateral orbital wall (arrows), with adjacent dural enhancement overlying anterior temporal lobe encroaching on the planum sphenoidale. There is a mass effect on the right lateral rectus muscle.

ing the carotid artery. Postoperatively, the patient reported improvement in her vision, claiming that vision in her right eye was far less dark. Formal postoperative evaluation is pending at the time of this publication.

Histopathological examination of the sphenoid bone specimen with hematoxylin and eosin staining showed a meningothelial mass within the bony trabeculae, with a few scattered psammoma bodies without nuclear atypia, confirming an intraosseous meningiomas (WHO grade I) (Fig. 4).

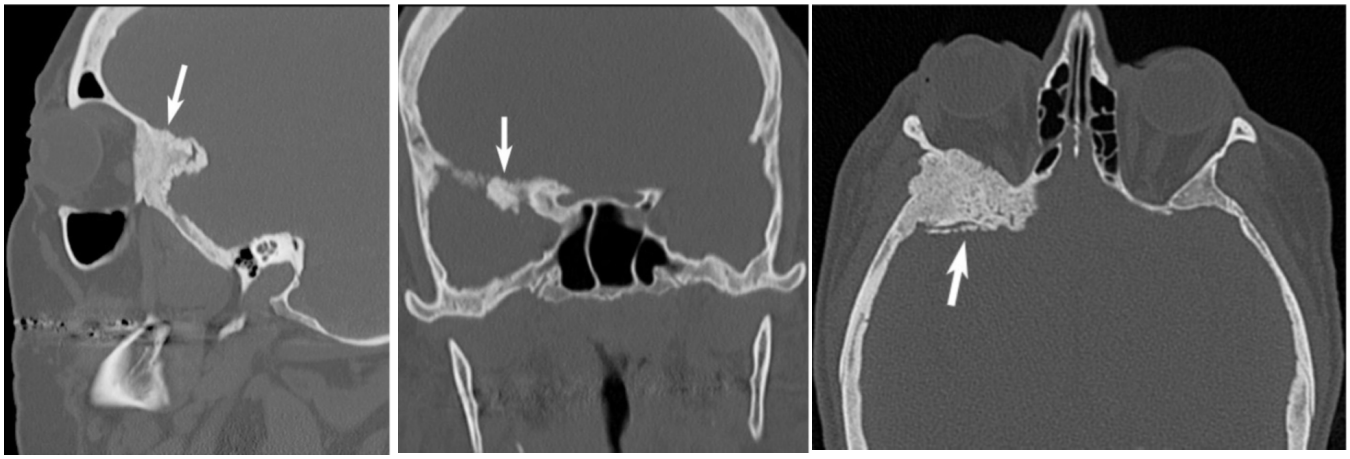

Figure 2. 50-year-old female with intraosseous meningioma. Sagittal (left), coronal (middle), and axial (right) CT images of the orbit in bone algorithm demonstrate an infiltrative osseous mass (arrows) involving the right orbital wall, orbital apex, and adjacent sphenoid wing with extension to involve anterior clinoid process on the right. (Note: normal appearance of left clinoid process). Linear dural reaction also noted particularly on the right axial image (arrow).

Discussion

Meningiomas are the second most frequent primary brain tumors (1). They arise from arachnoid cap cells of arachnoid villi and represent 15 % of all intracranial tumors (1, 2). 90% are benign (WHO Grade I) and are encountered commonly from age 40 to 70, with a male-to-female ratio of 1:32 (3). Meningiomas can be dural-based or extradural. The extradural subgroup has been referred to as ectopic, calvarial, cutaneous, extracranial, extraneuraxial, or intra-osseous (4, 5).

The differential diagnosis of a primary calvarial lytic lesion includes epidermoid tumor, multiple myeloma, eosinophilic granuloma, fibrous dysplasia, brown tumor, skull dermoid, lytic intraosseous meningioma, giant-cell tumor, Paget's disease, hemangiopericytoma, hemangioma, osteogenic sarcoma, and aneurysmal bone cyst (6).

Primary intraosseous meningiomas (PIM) are rare, and few cases have been reported in the literature. Fewer than 2% of meningiomas are extradural, out of which 14% are intra-osseous. Several explanations are proposed in the literature as to the probable origin of PIM. Azar-Kia suggested they arise from ectopic meningocytes or arachnoid cap cells trapped in the cranial sutures during molding of the head at birth. A second explanation is that PIM arises from dura and arachnoid entrapped by previous trauma. A third explanation is that extradural meningiomas arise from the multipotent mesenchymal cells. The latter may explain the mass located far from head and neck (7). Three different types of PIM exist: osteoblastic, mixed osteoblastic-osteolytic (most common), and purely lytic (least common). Frontoparietal and orbital regions are the most common locations for intra-osseous meningiomas (6).

The symptomatology of intra-osseous meningiomas is due mostly to the compression of the surrounding structures. If there is dural involvement, it can cause pain. Our patient had PIM of the right sphenoid bone involving both greater and lesser wings. There was a mild mass effect on the right orbital apex, with compression of the adjacent optic nerve, accounting for the patient's visual symptoms. The patient had a history of injury to the right eye which, as proposed by etiological theories in literature, could have contributed to development of intra-osseous meningioma.

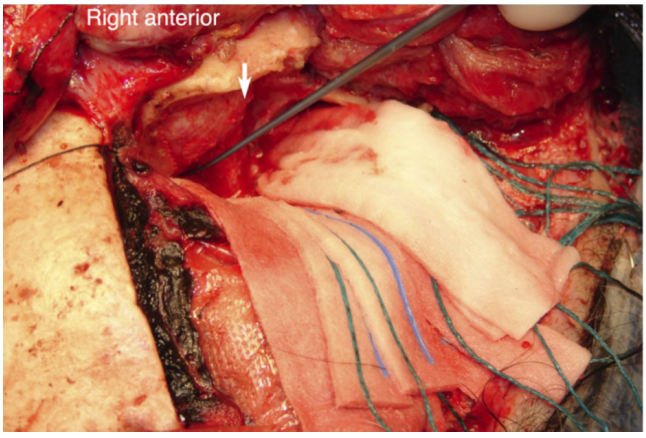

Figure 3. 50-year-old female with intraosseous meningioma. Intraoperative images, showing periorbita after the lesion was drilled away. The superior orbital and a portion of the lateral orbital rim (arrow) are seen. The surgical probe directly crosses the periorbita. Surgical cotton pads directly overlie the frontal and temporal surface of the brain.

In our review of literature, one of the close differential diagnoses that is important to separate out from PIM is fibrous dysplasia, as the treatment options may be different. Fibrous dysplasia is a developmental disease that is encountered at a younger age and ceases to grow at bone maturation (8). Radiologically, both of them expand bone and

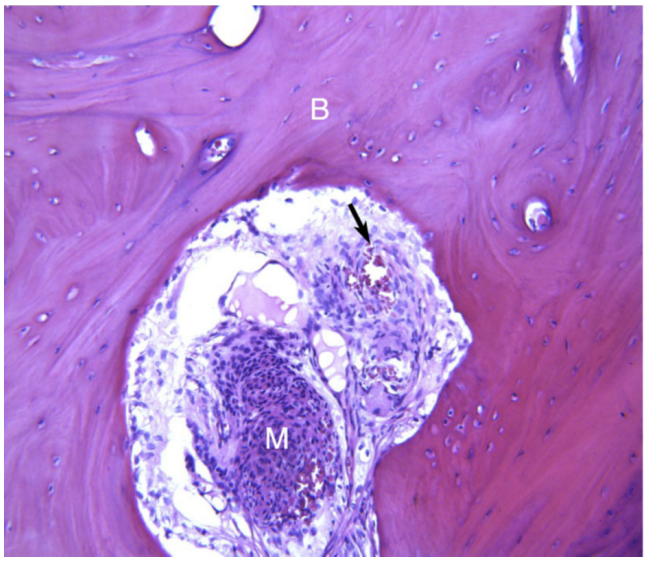

Figure 4. 50-year-old female with intraosseous meningioma. Medium-power microscopic section with hematoxylin and eosin stain. Meningioma (M) has invaded trabecular bone (B), forming a dense nest of meningothelial cells. Loose, spindled meningothelial cells enwrap nearby blood vessels (arrow).

have a ground glass appearance. However, in fibrous dysplasia, the inner table of the skull is typically smooth. In PIM, there is irregularity of the inner table, particularly at the site of origin, almost always with associated dural reaction. This irregularity is the key to the diagnosis on imaging (9).

References

1. Radhakrishnan K, Mokri B, Parisi JE, O'Fallon WM, Sunku J, Kurland LT. The trends in incidence of primary brain tumors in the population of Rochester, Minnesota. *Ann Neurol* 1995; 37:67-73. [PubMed]
2. Longstreth WT, Jr., Dennis LK, McGuire VM, Drangsholt MT, Koepsell TD. Epidemiology of intracranial meningioma. *Cancer* 1993; 72:639-648. [PubMed]
3. Tabernero MD, Espinosa AB, Maillo A, et al. Patient gender is associated with distinct patterns of chromosomal abnormalities and sex chromosome linked gene-expression profiles in meningiomas. *Oncologist* 2007; 12:1225-1236. [PubMed]
4. Tokgoz N, Oner YA, Kaymaz M, Ucar M, Yilmaz G, Tali TE. Primary intraosseous meningioma: CT and MRI appearance. *AJNR Am J Neuroradiol* 2005; 26:2053-2056. [PubMed]
5. Lang FF, Macdonald OK, Fuller GN, DeMonte F. Primary extradural meningiomas: a report on nine cases and review of the literature from the era of computerized tomography scanning. *J Neurosurg* 2000; 93:940-950. [PubMed]
6. Agrawal V, Ludwig N, Agrawal A, Bulsara KR. Intraosseous intracranial meningioma. *AJNR Am J Neuroradiol* 2007; 28:314-315. [PubMed]
7. Cho W, Im S, Oh CW. Cho-2005. J Korean Neurosurg Soc 2005; 37:238-240.
8. Harris WH, Dudley HR, Barry RJ. The natural history of fibrous dysplasia. An orthopaedic, pathological, and roentgenographic study. *J Bone Joint Surg Am* 1962; 44-A:207-233. [PubMed]
9. Daffner RH, Yakulis R, Maroon JC. Intraosseous meningioma. *Skeletal radiology* 1998; 27:108-111. [PubMed]