Intraosseous orbitosphenoidal cavernous angioma

Case report

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Primary orbital intraosseous angiomas are rare. The authors report the case of a 55-year-old man who harbored a multifocal cavernous angioma in an unusual sphenoorbital location. The lesion was responsible for unilateral exophthalmos and blindness. Characteristic imaging findings, which included a honeycomb pattern on plain x-ray films and computerized tomography scans, a heterogeneous high signal intensity on T2-weighted magnetic resonance images, and slowly flowing venous lakes on power Doppler ultrasonograms and angiograms, are presented and discussed.

Key Words • angioma • bone tumor • orbital tumor • benign • skull base

Osseous angiomas are most commonly found in the calvaria and the vertebral bodies. Primary osseous angiomas of the cranium are benign tumors comprising 0.2% of all osseous neoplasms. Those originating in the skull base are usually located in the temporal bone and rarely in the orbit. We present clinical and imaging findings in a patient who had an unusual primary intraosseous angioma of the sphenoid bone.

Case Report

History. This 55-year-old man had complained of proptosis with moderate exophthalmos 3 years before the current admission, but had been lost to follow-up examination until 4 months before admission, when he presented with total blindness in the left eye and impaired vision in the right eye. The patient had no history of trauma, and his medical records were unremarkable except for the presence of ankylosing spondylitis.

Examination. Plain skull x-ray films revealed a well-defined lytic lesion of the sphenoid bone on the left side. The lesion had a sunburst pattern with fine reticulated lines radiating from its center (Fig. 1). Computerized tomography (CT) scans delineated the expansive bone lesion, which involved the lesser and greater wings of the sphenoid bone as well as the orbital roof (Fig. 2). Magnetic resonance (MR) T2-weighted images revealed the lesion to be inhomogeneous. It measured 7.8 cm in its greater axis and projected intracranially, with foci of both increased and decreased signal intensity scattered throughout the lesion (Fig. 3 left). The tumor obliterated the sphenoid sinus and displaced the left cavernous sinus and the carotid artery. On T2-weighted MR images there was a markedly inhomogeneous signal intensity, with areas of increased signal intensity that corresponded to pooling of blood (Fig. 3 right). Contrast agent was not administered because of patient discomfort and movement. Transcranial color-coded Doppler ultrasonography, performed using a temporal window, did not reveal abnormal vessels. However, the increased sensitivity of the power Doppler recording allowed us to observe slowly flowing venous lakes interspersed with bony compartments. Angiography demonstrated increased vascularity in the area of the tumor, which had feeder vessels extending from the left ophthalmic and middle meningeal arteries (Fig. 4). No arteriovenous shunt was noted. The intracavernous segment of the carotid artery was encased and the middle and anterior cerebral arteries were stretched and deviated. We decided to embolize the external carotid artery by placing polyvinyl alcohol particles (300–600 μ) before surgery.

Operation. A near-total excision of the lesion was achieved via a bicoronal approach and a frontotemporal craniotomy. Moderate bleeding was controlled using bone wax.

Postoperative Course and Histopathological Findings. At the time of hospital discharge, there was dramatic cosmetic improvement in the patient as well as complete recovery in his right eye. Histopathological investigation confirmed the diagnosis of a cavernous angioma.

Discussion

Osseous angiomas are benign tumors that usually involve the flat bones and the spine and typically appear during the fourth or fifth decade of life. Parietal and frontal bones are the most common sites of involvement in calvarial lesions. Angiomas of the skull base are rare and the petrous portion of the temporal bone is the most
Orbitosphenoidal angioma

![Fig. 1. Plain x-ray film, oblique view, revealing a lytic lesion in the sphenoid bone with a sunburst pattern (asterisks).](image)

Frequent site. Less than 5% are primary intraosseous orbital angiomas (PIOAs), which most commonly involve the frontal bone.

Since the first PIOA was described by Rowbotham in 1942, few reports of this lesion have been published. In an article in which they reviewed reports of PIOAs in the literature prior to 1994 along with a case of their own, Banerji and colleagues stated that the sphenoid bone was involved in only two of 26 cases. Although in most instances the PIOA was small, measuring 1 to 3 cm, the lesion in their case involved three orbital bones (frontal, ethmoidal, and sphenoidal) and was larger than 4 cm in all dimensions. The only similar large PIOA involving the frontal and sphenoid bones was reported by Mortada in 1964. The PIOA in our case, also multifocal, is the largest ever described.

Primary intraosseous orbital angiomas are equally distributed between both genders of patients and usually present with ophthalmic symptoms. They cause a slowly progressive painless proptosis without pulsation or bruit and tend to expand downward into the orbits, displacing the globes. Our patient’s lesion, however, had both orbital and intracranial expansion.

On histopathological examination, angiomas appear to be composed of mature thin-walled blood vessels lined by a single layer of flattened endothelial cells. The most striking difference between these lesions and their soft-tissue counterparts is the abundant trabecular bone located between the thick connective tissue stroma that surrounds the vascular spaces. These trabeculae are not part of the primary hamartomatous pattern, but are secondary to reactive bone destruction and formation. Angiomas can be subdivided into two types. Although most lesions of the skull and, particularly, those of the orbital bones are of the cavernous type, those found in the vertebrae are more often capillary. Some are mixed types.

Primary intraosseous orbital angiomas have to be differentiated from other similar slow-growing bone tumors, such as fibrous dysplasia, aneurysmal bone cyst, dermoid cyst, meningioma, osteoma, or, rarely, osteogenic sarcoma. Careful radiological evaluation coupled with clinical findings allows differentiation among these lesions. However, diagnosis is frequently made only on the basis of findings from a pathological examination. In fact, the radiographic appearance of PIOA is highly diagnostic on plain x-ray films, with a characteristic roentgenographic presentation first described by Bucy and Capp. It shows an expansive, well-circumscribed intradiploic area of rarefaction with a honeycomb configuration on axial views and a classic sunburst pattern of trabeculation on tangential views. There is usually no reactive sclerosis at the margins. The use of CT scanning for diagnosis of intraosseous angioma can provide additional useful information concerning intracranial extension, effect on orbital contents, or vascular displacement. However, findings of an of capillaryvenous malformations in accordance with the Mulliken–Merland classification.

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expansive lytic process with a high-density, enhancing, amorphous texture can suggest fibrous dysplasia, known to cause cranial nerve compression. Furthermore, in the series published by Leeds and Seaman, the sphenoid bone was the second most commonly involved bone in fibrous dysplasia of the skull. Nevertheless, the uniformly reticulated structure is not seen in fibrous dysplasia, but recalls the classic appearance of osseous angioma. Magnetic resonance imaging can demonstrate a mottled increased signal on T₁- and T₂-weighted images. Chemical-shift images and histopathological studies reveal that fatty tissue, a major component, causes the increase in signal intensity on T₁-weighted images, whereas the high signal intensity observed on T₂-weighted images is caused by the slow flow or pooling of blood. After a bolus injection of contrast agent during rapid serial MR imaging, enhancement is observed in the focal area during an early phase and spreads into the entire lesion in a later phase. It has been suggested that these features are characteristic of osseous angiomas. Recently, the use of Doppler power imaging, as advocated by Slaba in cases of venous malformations (unpublished communication, 1996), can be helpful in detecting these slowly flowing venous lakes that otherwise fail to be identified on conventional color Doppler ultrasonography. Although not necessary in small tumors, angiography plays an important role in preoperative surgical planning and embolization in large ones. It typically shows increased vascularity in the area of the lesion, with feeder vessels but no large draining veins. In rare instances the lesion may be supplied by dural arteries, as in our case.

Treatment of osseous angiomas of the orbit by surgical resection of tumor with a normal bone rim has been advocated by all authors. The surgical approach and excision become difficult for extensive cranial base tumors. Prognosis after complete excision is uniformly good. Radiation therapy is reserved for cases in which only subtotal resection has been achieved or for unresectable lesions in symptomatic patients.

Conclusions

Intraosseous angioma of the skull should be included in the differential diagnosis of expansive osseous orbital lesions. Classic radiological features can occasionally differentiate these tumors. If a honeycomb pattern is visualized on CT scanning and an increased mottled signal is demonstrated on both T₁- and T₂-weighted images, a diagnosis of osseous angioma should be considered. Although angiography and embolization are helpful in treating large lesions, complete excision is the treatment of choice. Our case is original in that the lesion was multifocal and the largest ever described in the sphenoorbital region, which remains an exceptional location. Imaging in this case was complete, including plain x-ray films, CT scans, MR images, Doppler ultrasonograms, and angiograms.

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