Primary Orbital Angiomatous Meningioma

The first tangible description of an intraorbital meningioma can be credited to Antonio Scarpa in 1816. Since then, our knowledge of meningiomas and especially intraorbital meningiomas has greatly expanded. In 1938, Cushing and Eisenhardt described 4 main subcategories of meningiomas, of which the vascular or angiomatous tumor was a minor subtype. We describe what we believe to be the first reported case of an angiomatous meningioma arising primarily from within the orbit.

Report of a Case. A 17-year-old adolescent girl had a 3-month history of progressive painless proptosis of the right eye (Figure 1). The proptosis appeared to worsen in the morning with progressive improvement throughout the day. An initial examination revealed evidence of mild resistance to retropulsion. Her ocular history was otherwise significant only for mild refractive error. A systematic medical record review was noncontributory. Her best-corrected near and distance visual acuity was 20/20 OU. Exophthalmometry revealed a 4-mm proptosis of the right eye. No afferent papillary defect was noted, and extraocular motility was full. A slit-lamp examination was unremarkable in both eyes. A dilated fundus examination revealed normal fundi as well as normal and symmetric optic nerves. Humphrey visual field tests revealed a possible nasal defect, which was not subsequently confirmed with Goldmann perimetry. A magnetic resonance imaging scan was performed. On the coronal T1-weighted, fat-suppressed, contrast-enhanced sequence, a lobulated mass was seen in the superomedial orbit; it partially encased the optic nerve, which was displaced inferolaterally (Figure 2). On the axial proton density series, the mass was seen to cause proptosis without any globe deformity (Figure 3). Within the mass, there was at least 1 vascular flow void.

The presumptive diagnosis was that of an orbital cavernous hemangioma. After informed consent was obtained, the patient underwent a combined medial and lateral orbitotomy with excision of the right orbital tumor. Surgical exploration revealed a poorly defined vascular tumor encasing the optic nerve, but it was not readily separable from the nerve with blunt dissection. The base of the tumor was therefore ligated, and the remaining portion of the mass was excised. Numerous sections at multiple levels were obtained from the specimen for review. A histopathologic examination revealed large intravascular spaces lined by endothelial cells and containing erythrocytes. Multiple septa were noted showing intense focal basophilia that did not stain for calcium. The pupils were found to be equal and reactive to light. The fi-
nal diagnosis was that of a cavernous hemangioma of the right orbit.

Postoperatively the right eye proptosis resolved, but the patient was found to have no light perception with the appearance of an infarcted nerve and retina on a fundus examination. Following the initial procedure, the patient received numerous routine follow-up visits without evidence of any alteration in her ocular examination results. Five years later on a routine visit, the patient complained of mild swelling of her right eyelid as well as general irritation and tearing of the right eye for approximately 2 months. An examination revealed a right afferent papillary defect that had been present since her initial operation. Exophthalmometry revealed a 4-mm proptosis of the right eye. There was some mild ptosis of the right upper eyelid. On a slitlamp examination, moderate engorgement of the episcleral vessels was noted in the right eye. Goldmann applanation tonometry revealed an intraocular pressure of 18 mm Hg OD and 13 mm Hg OS. Examination results for the left eye were unremarkable. A fundus examination of the right eye showed a total retinal detachment with traction bands. A magnetic resonance imaging scan was performed. On the coronal (Figure 4A) and axial (Figure 4B) T1-weighted, fat-suppressed, contrast-enhanced sequences, a large lobulated mass was seen occupying most of the orbit. The optic nerve could no longer be identified and was presumably completely encased by the mass. Inflammatory mucosal disease was incidentally noted in the right ethmoid bone and both maxillary sinuses.

The possibility of a recurrent vascular tumor was considered, and multiple consultations were conducted to evaluate the best treatment modalities. These included surgical exploration and vascular embolization.

At the same time, efforts were made to reconsider the original diagnosis, and the initial pathologic slides were carefully reexamined. In addition to a cavernous hemangioma, the possibilities of a hemangiopericytoma or meningioma were suggested. New slides were prepared from fresh tissue obtained from the original specimen. A careful review of the latter revealed evidence of meningiomatous cells at the base of the tumor, which was otherwise angiomatous in nature (Figure 5). A right frontal craniotomy and orbital zygo-
matic osteotomy were performed to provide an anterior skull-based approach to the orbit. A right orbital exenteration was then performed (Figure 6). A pathologic examination of the specimen revealed a meningothelial meningioma confined to the orbit. Immunohistochemical stains were applied to each surgical specimen. The tumor failed to express desmin, MIB-1, or factor VIII but did express vascular endothelial growth factor (VEGF)-1, VEGF-2, and vimentin, confirming the histopathologic diagnosis.2

Comment. Meningiomas constitute the most common nonglial intracranial tumors in both Western and Eastern countries. The first described case of an intraorbital meningioma was provided by Scarpa in 1816. Various forms of meningiomas are commonly encountered by neurosurgeons, and extensive literature exists regarding their classification and associated diagnostic controversies. When comparing primary orbital meningiomas with primary intracranial meningiomas, Karp et al3 found that they followed similar sex and race predilections, although primary orbital meningiomas tended to be significantly more aggressive in children. Women in their mid thirties appeared to be at the highest risk. The authors found that most of these tumors tended to occur within the right orbit.

Meningiomas have been divided into several histologic subtypes, of which the angioblastic meningioma is a rare form.4 Because they are so uncommon, little is known about their natural history and long-term prognosis. Furthermore, their exact histologic classification has been a source of debate in the past. Sugar et al5 described a case in which they emphasized the difficulty in differentiating a vascular (angioblastic) meningioma from a hemangiopericytoma. They suggested that if the vessels are ensheathed by meningothelial cells, the tumor should be considered an angioblastic meningioma. However, they recognized that morphological analysis alone was not entirely satisfactory to secure a diagnosis. Later in vitro studies fueled this debate by showing that pericytes have pluripotent characteristics and can theoretically develop into a variety of morphologic entities.6

Despite their similarities, it is clear that hemangiopericytomas and angiomatous (or angioblastic) meningiomas are separate histopathologic phenomena. In 1953, Begg and Garrett7 presented a case report followed by a review of the literature on hemangiopericytomas occurring in the meninges and pointed to the fact that in meningiomas, silver staining using the method of Perdrau showed proliferation within the vascular spaces and an absence of reticulation between the cells. More recent studies have focused on immunohistochemical and ultrastructural findings in these tumors in an attempt to establish clear differentiating guidelines. D’Amore et al8 examined various soft tissue and meningeal hemangiopericytomas using light microscopy and a panel of targeted antibodies. Although they found no clear guidelines to separate the ultrastructural features, they found that the immunophenotypical profile of hemangiopericytomas showed a lack of epithelial membrane antigen and cytokeratin. In 1997, Dietzmann et al2

Figure 5. Microscopic sections from the original specimen. A, Extensive vascular channels along with meningothelial cells. B, Arrow indicates a less vascular area of the lesion with a preponderance of meningothelial cells.

Figure 6. Gross specimen from the exenterated orbit revealing a tightly adherent mass posterior to the globe.
performed a comprehensive immunohistochemical comparison of the vascular growth factors and hormonal markers in angiomatous and atypical meningiomas as well as hemangiopericytomas. They demonstrated evidence of hormonal receptors to actin only in angiomatous meningiomas, and to muscle actin only in hemangiopericytomas. In addition, they showed a complete absence of VEGF-1 in the extravasates obtained from hemangiopericytomas. This case further validates their findings by demonstrating the presence of VEGF-1 and VEGF-2 as well as vimentin in the specimen. Therefore, in the absence of clear morphological differences, in certain cases the clinical distinction between vascular tumors and highly vascularized meningiomas can be difficult; one may need to rely on histochemical analysis.

This case is unique because primary orbital meningiomas not arising from the optic nerve are exceedingly rare. Although somewhat controversial, it is believed that a subpopulation of orbital meningiomas arises from within the orbital tissues, unrelated to the walls of the optic nerve. In a review of several cases of free-lying orbital meningiomas, McMichael and Cullen proposed various possibilities as the origin of these tumors: (1) that the tumor is originally attached to the optic sheath but later migrates away from it; (2) that the tumor cells arise from the orbital interstitial tissues; (3) that the tumor cells arise from suture lines between orbital bones that contain herniated meninges; or (4) that the cells originate from the dura mater lining the orbit. Because of their peculiar behavior and location, these orbital meningiomas are often misdiagnosed as hemangiomas or dermoid cysts.

In an attempt to find other diagnostic differentiating clues, Uttley at al reviewed several cases of intracranial atypical meningiomas as compared with hemangiopericytomas. They discovered that on angiography, hemangiopericytomas in particular demonstrated a major vascular supply from the branches of the internal carotid or vertebral artery. However, they were unable to show clear differences on magnetic resonance imaging or computed tomography. Therefore, it is doubtful that radiologic clues alone can truly aid in the diagnosis of this condition.

This case is especially unique in that the histologic examination showed areas completely devoid of tumor and an uninvolved optic nerve sheath. On further review, the histologic findings are consistent with a primary angiomatous meningioma of the orbit, a previously unreported entity. We conclude that in cases of atypical behavior of an otherwise classic vascular tumor of the orbit, one should consider the possibility of an angiomatous meningioma.

Ebrahim Elahi, MD
Murray A. Melzer, MD
Alan H. Friedman, MD
Peter M. Som, MD
New York, NY

Corresponding author and reprints: Ebrahim Elahi, MD, Department of Ophthalmology, One Gustave Levy Place, Annenberg Bldg, 22nd Floor, New York, NY 10029.


Nonarteritic Anterior Ischemic Optic Neuropathy Associated With Acute Glaucoma Secondary to Posner-Schlossman Syndrome

The occurrence of nonarteritic anterior ischemic optic neuropathy (NAION) has been well described following cataract surgery. It has been postulated that a perioperative increase in intraocular pressure leads to a decrease in optic nerve head perfusion pressure, which results in ischemia of the optic disc. Acute rises in pressure secondary to other causes could also result in NAION. The lack of cases in the literature reporting NAION following episodes of acute glaucoma is thus surprising. Although vision loss in acute glaucoma has always been thought to be secondary to optic nerve head ischemia, to our knowledge, NAION following acute glaucoma has only been described in one report. We report a case of NAION occurring during an episode of acute glaucoma secondary to Posner-Schlossman syndrome.

Report of a Case. A 71-year-old white woman with a history of hypertension and hypercholesterolemia sought treatment for a 1-week history of intermittent left eye pain and decreased vision. Results of a right eye examination were normal. Initial visual acuity was 20/200 OS, and a relative afferent pupillary defect was present in the left eye. Conreal edema and trace anterior chamber cell and flare were noted. Intraocular pressure was 69 mm Hg. Gonioscopy revealed grade IV angles with light trabecular meshwork pigmentation and no peripheral anterior synchia.

A dilated fundus examination revealed a mildly edematous left optic nerve and a crowded right optic nerve with a cup-disc ratio of 0.2. Humphrey visual field tests detected an inferior altitudinal defect in the left eye. The erythrocyte sedimentation rate was 16 mm/h. Once the pressure was controlled, the patient had no further complaints of eye pain. The patient had no family history of stroke at a young age, blood clots, unexplained loss of vision, or glaucoma. The patient denied having other neurologic symptoms, including symptoms suggestive of giant cell arteritis. Results of tests for homocysteine, anticardiolipin antibody, lupus anticoagulant, rapid plasma reagin, fluorescent treponemal antibody absorption, and antinuclear antibody were all within reference ranges. Left NAION was diagnosed. The patient was advised to take an aspirin daily. The patient’s intraocular pressure was acutely controlled with eye