Endoscopic resection of a venous hemangioma of the optic nerve sheath

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Abstract
We describe a case of a venous hemangioma that arose in the optic nerve sheath of the orbital apex in a 28-year-old man who presented with progressive vision loss. To the best of our knowledge, this is the first reported case of a venous hemangioma occurring at this location. A definitive diagnosis and partial excision was achieved via an endoscopic transsphenoethmoid approach with interactive, computer-assisted, frameless stereotactic surgical navigation. The intervention resulted in minimal morbidity, demonstrating yet again that this surgical approach is a safe and effective way to treat lesions of the orbital apex.

Introduction
Venous hemangiomas are characterized by the proliferation of vascular channels whose walls contain layers of smooth muscle. These lesions were originally called atri venous hemangiomas because it was believed that they contained both arterial and venous elements. However, most studies have since failed to detect the presence of elastic lamina, leading some authors to conclude that the thick-walled vessels in these lesions represent veins and not arteries. With little evidence to support the presence of arterial components, the term venous hemangioma has been proposed to more accurately reflect the histology of these lesions.

Hemangiomas, including the more common capillary and cavernous forms, are hamartomatous lesions. Although the lesions themselves are benign, they can cause symptoms when they grow and compress neighboring structures. They are believed to enlarge via canalization of hyperplastic solid masses of endothelial cells in a way that mimics the process that occurs during the embryogenesis of the vascular system.

Venous hemangiomas have been reported at sites throughout the body. They are rarely associated with significant morbidity, although some that have arisen in the brain were complicated by infarction, which resulted in various degrees of neurologic deficits.

In this article, we describe a case of venous hemangioma that arose in the optic nerve sheath of the orbital apex. To the best of our knowledge, this is the first report of such a lesion occurring at this location.

Case report
A 28-year-old man was referred to the Department of Otorhinolaryngology—Head and Neck Surgery with a history of a slowly progressive loss of vision in his left eye over a period of 15 months. The degeneration of his vision had accelerated during the preceding 5 months. The patient reported the presence of an expanding white patch in the center of his left visual field, as well as a deterioration of both his peripheral vision and color perception. He reported no pain, diplopia, sinus symptoms, or other neurologic complaints. His medical, surgical, and family history was noncontributory.

A recent vision test had determined that the patient’s visual acuity was 20/400 in the left eye and 20/20 in the right eye. The left eye also exhibited a large central scotoma, dyschromatopsia, and a left afferent pupillary defect. A fundal examination revealed that the left optic nerve was pale and cupped; the right optic nerve was normal. No proptosis or ophthalmoplegia was evident.

Computed tomography (CT) detected soft-tissue density in the left orbital apex and a widening of the left bony optic canal (figure 1). Magnetic resonance imaging (MRI) identified a 1.4 × 0.5-cm lesion within the left optic apex with isointensity on T1-weighted imaging and homog-
enous hyperintensity on T2-weighted imaging (figure 2). The left optic nerve was severely atrophic, with changes extending posteriorly toward the chiasm. Endoscopic orbital decompression with possible biopsy or excision of the lesion was planned to preserve the remaining vision, provide a histopathologic diagnosis, and plan for future management.

All appropriate preoperative assessments and planning were conducted, and the patient returned for an extradural tumor biopsy and partial resection via an endonasal transthamoid, transphenoid approach. An interactive, computer-assisted, frameless stereotactic surgical navigation system with accuracy to 1.7 mm was used to confirm the anatomic location. Following the application of topical decongestants, mucosal and transnasal sphenopalatine injections of 1% lidocaine with epinephrine were administered to achieve hemostasis. Following an uncinectomy and resection of the ethmoid bullae, the medial orbital wall was identified and traced posteriorly to the basal lamella. Perforation of the lamella exposed the superior turbinate, and the inferior one-third was resected. The resection exposed the sphenoid os, which was then enlarged. The optic nerve was seen in the superolateral area of the sphenoid

Figure 1. Preoperative CT demonstrates the soft-tissue density in the left orbital apex (arrow) and the widening of the left bony optic canal.

Figure 2. A: Preoperative T1-weighted coronal MRI demonstrates the isointense enlargement of the optic nerve (arrow) in the area of the left optic canal. B: Preoperative T2-weighted axial MRI demonstrates the hyperintense lesion (arrow) along the left optic nerve.
sinus and confirmed by interactive imaging. A high-speed suction and irrigation diamond bur was used to thin the bone overlying the optic nerve from the posterior ethmoid cavity to the area of the optic chiasm. The optic nerve sheath was then incised parallel to the nerve fibers with an arachnoid knife, revealing a purplish, fibrinous growth that protruded through the incised sheath. Multiple biopsies along the medial aspect of the nerve were obtained, but a complete excision of the lesion was not attempted because of its close circumferential relationship to the optic nerve. A small cerebrospinal fluid leak was noted through the optic nerve sheath, and a mucosal graft from the nasal septum was placed over this area and secured with fibrin glue. Nasal packing was inserted, and the patient awakened from surgery. The patient’s brief postoperative course was notable for an improvement in visual acuity in his left eye from 20/400 to 20/200.

The biopsy specimens were fixed in 10% neutral-buffered formalin and transferred to the Department of Surgical Pathology at the Hospital of the University of Pennsylvania for appropriate histochemical analysis. There a diagnosis of venous hemangioma was made. Histologic sections showed open vascular channels lined with endothelium. In contrast to the typical hemangioma that contains thin walls of fibrous tissue around capillary or cavernous spaces, here the channel walls were thick and made up of smooth muscle consistent with small veins (figure 3).

Discussion

Our search of the English-language literature identified only 1 report of a venous hemangioma occurring at the optic chiasm. In that case, the patient presented with decreased vision over a 3-week period following infarction of the optic nerve. Hemorrhage was determined to be the cause of the patient’s symptoms, and the patient regained some of his vision following surgical removal of thrombi at the site of the lesion.

To the best of our knowledge, the case we describe herein represents the first report of a venous hemangioma occurring within the optic nerve sheath. Because our patient exhibited no evidence of infarction, we believe that his vision loss had been caused by compression of the optic nerve following growth of the lesion. Furthermore, in view of the fact that the patient had been complaining of visual problems for 15 months, and because the optic nerve was atrophic on MRI, we believe that he had experienced chronic irreversible changes in the optic nerve that are unlikely to fully reverse, even if a complete surgical debulking could be achieved.

There have been 2 cases of a similar cavernous hemangioma of the optic nerve in which growth was documented. However, unlike our patient, neither of these 2 patients had experienced a significant vision loss following the growth of their lesions.

Multiple surgical techniques have been described to approach vascular lesions in the orbital apex. These procedures include lateral orbitotomy, transconjunctival cryoextraction, medial orbitotomy, gamma-knife radiosurgery,11 and anterior orbital rim incisions.12 In particular, the transconjunctival and medial orbitotomy approaches to hemangiomas medial to or enveloping the optic nerve have been described.9,10

Endoscopic sphenoethmoidectomy is a less invasive and less morbid approach to removing lesions of the orbital apex than are traditional surgical approaches such as transcranial and transorbital procedures. The sinonasal endoscopic approach has been well described as a means of repairing cerebrospinal fluid leaks13 and orbital decompressions14 and as an approach to sella turcica lesions.15 Only a few reports of the use of a transnasal endoscopic approach to lesions of the orbital apex have been published in the literature.16,17 Sethi and Lau used this technique to obtain tissue biopsies in 6 patients with orbital apex lesions; they reported that this approach was reliable and provided good visualization of the operative field.16 Kingdom and Delgaudio used an endoscopic approach in 5 patients with orbital apex lesions, and it provided a definitive diagnosis in all 5 cases.17

The anatomy of the lateral sphenoid sinus wall is intricate and variable; many vital structures are located in close proximity, including the optic nerve, carotid artery, second branch of the trigeminal nerve, and cavernous sinus. The use of powered endonasal instruments is often necessary, but this increases the risk of significant complications. The use of interactive, computer-assisted, frameless stereotactic surgical navigation has become increasingly common in endoscopic sinus surgery; this modality provides surgeons with the ability to correlate point-to-point anatomy with a
patient’s CT and/or MRI scans. Fine-cut axial CT scans are reformatted into sagittal and coronal planes, which allows the surgeon to intraoperatively localize the anatomy in three dimensions. Although stereotactic navigation adds to the expense of surgery, increases intraoperative time, and in no way substitutes for a thorough understanding of paranasal sinus anatomy and direct visualization of anatomic landmarks, it can be a powerful adjunctive tool for confirming tumor location and for increasing the safety of endoscopic sinus surgery.18,19

References