Cavernous Malformation of the Optic Chiasm – A Diagnostic and Treatment Dilemma

Sundeep Uppal, Randy A. Walker, Edward J. Atkins

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Cavernous malformations affecting the optic chiasm are a rare cause of visual impairment. These low flow vascular lesions are associated with minor recurrent hemorrhage, and if this occurs in the optic chiasm, patients typically present with sudden onset of bitemporal hemianopia, retro-orbital headache, and nausea.[1] The majority of cases have extra-chiasmal involvement extending into the optic nerve or tract.[2] We present a case of chiasmal cavernous malformation, which demonstrates the challenges of clinical and radiologic diagnosis and surgical treatment.

Case Report

A 48-year-old previously healthy male presented with sudden onset severe headache and right nasal visual field loss. An urgent unenhanced computed tomographic (CT) scan report suggested pituitary apoplexy or a distal right internal carotid artery aneurysm (Figure 1a). Visual acuity was 20/40 right eye and 20/20 left eye with a 0.3 log unit left relative afferent pupillary defect (RAPD). Ocular motility was full and there was no diplopia. Goldmann visual field showed an incongruous left homonymous hemianopia (Figure 2). The CT angiogram report suggested a right suprasellar lesion with mass effect on the optic chiasm, and no contrast filling. The radiologic differential diagnosis included a thrombosed aneurysm. Magnetic resonance imaging (MRI) report suggested a hemorrhagic mass lesion arising from the chiasm and the differential included a chiasmal glioma or metastatic focus (Figure 1b). Only after 1) discussion with the radiologist affirming the sudden onset of headache and visual loss, 2) informing the radiologist of previous case reports of cavernous malformations of the optic chiasm with similar radiologic appearance, and 3) MRI reinterpretation, was cavernous malformation of the optic chiasm included in the

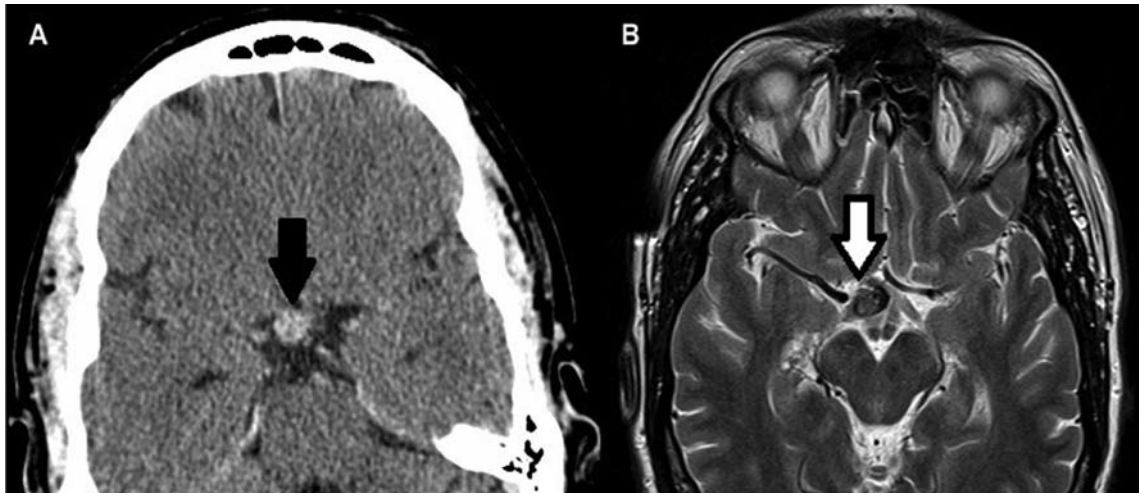

Figure 1: *a) Unenhanced axial CT brain showing a suprasellar hyperdensity reported as suggestive of thrombosed distal internal carotid artery aneurysm or pituitary hemorrhage (black arrow). b) Axial T2 weighted MRI brain and orbits showing an irregular lesion with mixed signal intensity and a "popcorn-like" appearance arising from the suprasellar region within the optic chiasm (white arrow).*

From the College of Medicine (SU); Department of Ophthalmology (RAW, EJA), Saskatoon City Hospital; Division of Neurology (EJA), Department of Medicine, Royal University Hospital, University of Saskatchewan, Saskatoon, Saskatchewan, Canada.

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Correspondence to: Edward J Atkins, Room 1651, Royal University Hospital, 103 Hospital Drive, Saskatoon, Saskatchewan, S7N 0W8, Canada.
Email: edward.atkins@usask.ca

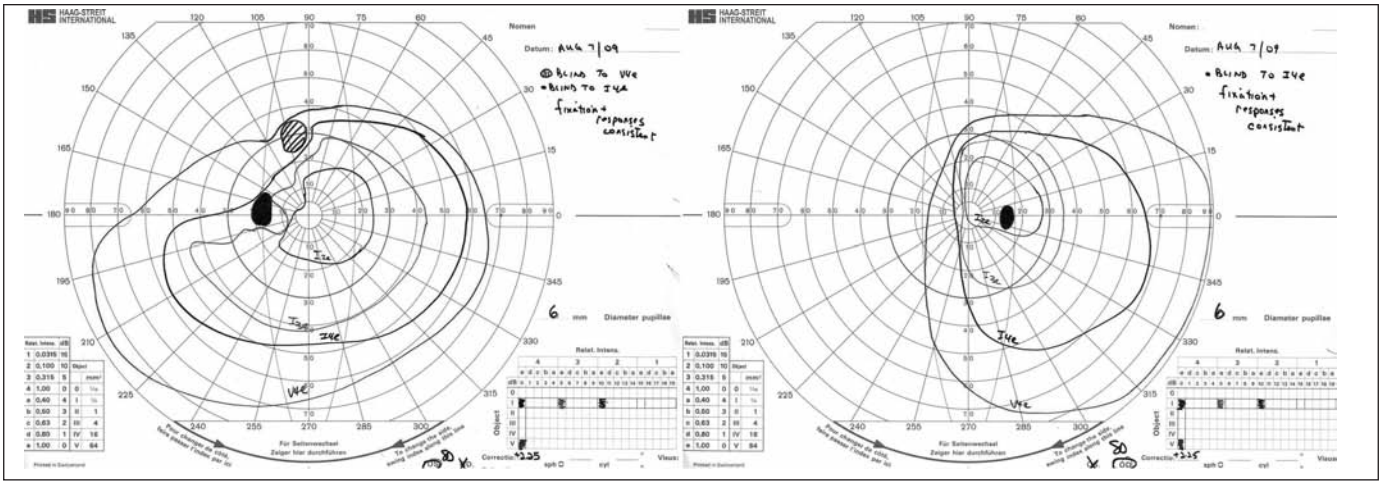

Figure 2: *Goldmann visual field showing an incongruous left homonymous hemianopia after initial hemorrhage of cavernous angioma of the optic chiasm.*

radiologic differential diagnosis. The radiologist and neurosurgeon involved both felt that cavernous malformation was still unlikely in that location, and glioma was the favoured radiologic diagnosis despite inconsistency with the age of the patient and the clinical presentation of sudden onset of visual loss and headache. Observation and follow-up MRI was recommended. After a few months of stability and improved vision, there was recurrent progressive visual loss with a now complete left homonymous hemianopia and a 0.6 log unit left RAPD. The MRI was repeated, and the most likely diagnosis was confirmed to be cavernous malformation of the optic chiasm (Figure 3). Surgical resection was successfully performed and histopathology confirmed the diagnosis of cavernous malformation. Visual acuity remained stable while driving Goldmann visual fields showed an increase from 95 to 130 degrees of contiguous horizontal visual field post-resection (Figure 4).

Discussion

Cavernous malformations are relatively common lesions with an incidence of 0.3-0.7% in the general population.[2] They exist in both sporadic and familial forms, with the latter more commonly presenting with multiple lesions in the central nervous system (CNS).[1] Cavernous malformations have been reported to affect cranial nerves;[3] although involvement of the optic apparatus is extremely rare, representing less than 1% of all cavernous malformations.[2] The lesions themselves are benign and do not contain or invade local neural tissue. These lesions are associated with recurrent and usually minor hemorrhage causing focal neurologic sequelae, especially in lesions associated with small structures such as cranial nerves, brainstem, or anterior visual pathways.

Because cavernous malformations of the optic chiasm are so rare, and since the clinical presentation can mimic other more common neuro-ophthalmic diagnoses, chiasmal cavernous malformations may be inadvertently excluded from the differential diagnosis of lesions affecting the anterior visual pathway apparatus, as was the case with our patient. The absence of endocrine dysfunction, ophthalmoplegia and associated diplopia can help distinguish a chiasmal malformation from pituitary apoplexy, since sudden expansion of the pituitary can compress oculomotor nerves in the adjacent cavernous sinuses, but this would not be expected with cavernous malformations which hemorrhage into surrounding brain parenchyma and may be associated with transient surrounding vasogenic edema and indistinct increase in surrounding T2 hyperintense signal on MRI (Figure 1b). Hemorrhage helps differentiate chiasmal malformation from optic glioma. Cavernous malformations

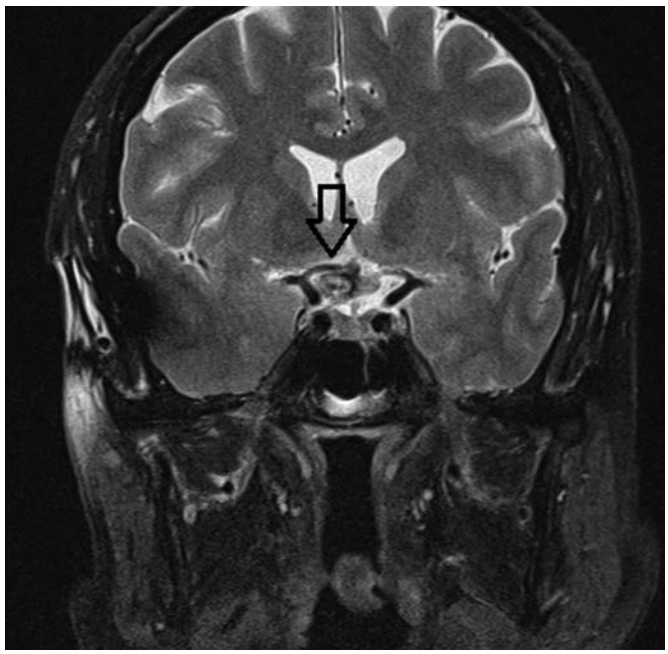

Figure 3: *Coronal T2 weighted MRI brain with fat suppression, showing cavernous malformation eccentrically located to the middle of the optic chiasm.*

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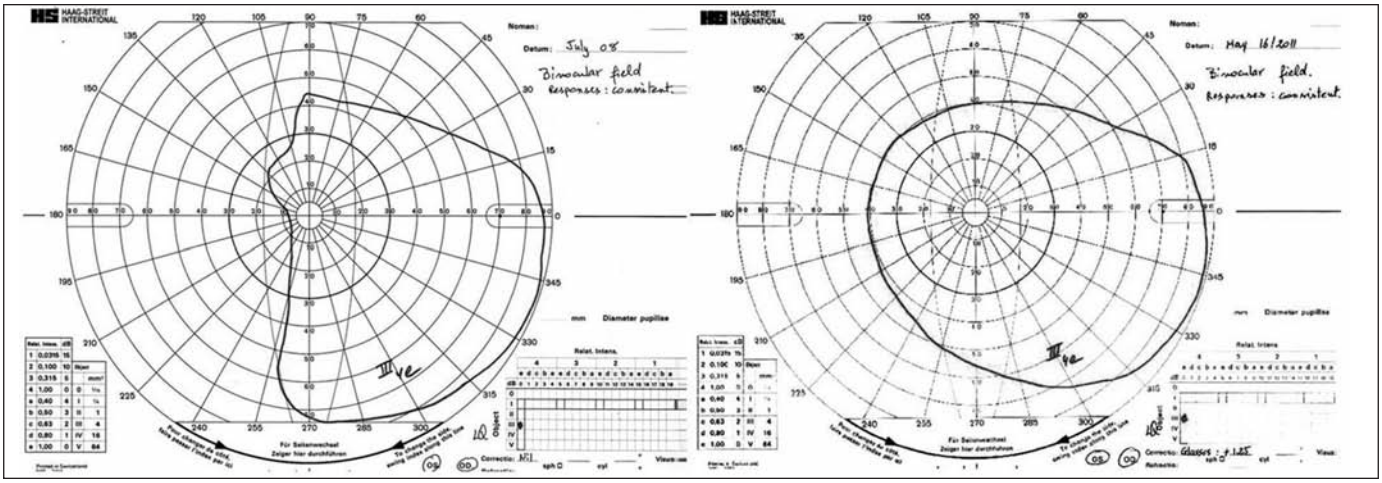

Figure 4: *Comparison of driving Goldmann visual fields from initial presentation and post-surgical resection shows an increase from 95 to 130 degrees horizontal contiguous visual field.*

often appear well demarcated on CT with foci of hyperdensity and calcifications resulting from calcified intra-lesional vasculature.[3] An MRI is the most sensitive and specific imaging modality for identifying cavernous malformations, especially when gradient echo or susceptibility weighted sequences are performed; however, these sequences are often only added when cavernous malformation is included in the clinical differential diagnosis, and may have to be specifically requested.[4] Tiny cavernous malformations appear as punctate foci of low signal, becoming more conspicuous on gradient echo sequences, due to paramagnetic susceptibility (calcium and/or hemosiderin). Larger cavernous malformations have a classic "popcorn-like" appearance, visible in Figures 1b in the axial plane and in Figure 3 in the coronal plane. They are typically well defined, rounded or lobulated, with a very characteristic rim of low signal that again exhibits paramagnetic susceptibility. This rim is well illustrated in Figure 1b. Variation from this typical radiographic appearance on MRI can occur making pre-operative diagnosis difficult.[1] Angiography is often of limited diagnostic value due to the low internal flow of lesions and high incidence of thrombosis making a differentiation between cavernous malformation and thrombosed aneurysm difficult.[3]

Gross total resection is the preferred surgical treatment, but surgery on the anterior optic pathway always carries a high risk of permanent visual loss. Fortunately, a cleavage plane between the malformation and normal neural tissue generally exists, which can allow successful resection to be performed, hopefully resulting in improved visual field and acuity.[2,5] The importance of considering neurosurgical excision of these precariously located hemorrhagic lesions is accentuated by the potential for visual field improvement. In our case, successful neurosurgical intervention allowed the patient to meet the visual field requirements for driving, and maintain his livelihood and quality of life. This report contributes to our understanding of the variability of the clinical and radiologic features of anterior optic pathway cavernous malformations, and underscores the importance of considering cavernous malformation in the differential diagnosis of hemorrhagic lesions of the optic chiasm.

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