Journal of Clinical Neuroscience 22 (2015) 258–267

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Review

Anterior visual pathway cavernous malformations

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ARTICLE INFO

Article history:
Received 8 April 2014
Accepted 28 July 2014

Keywords:
Cavernous malformation
Chiasmatic apoplexy
Optic chiasm
Optic nerve
Optic tract
Visual pathway

ABSTRACT

Anterior visual pathway cavernous malformations (CM) are rare diagnoses with poorly-defined natural history and management. A systematic review of all reports of anterior visual pathway CM was performed to identify all English-language articles with histopathologically-proven anterior visual pathway CM published from 1950 to December 2013. Patient demographics, presenting symptoms, CM location, treatment modality and clinical outcome were recorded and analyzed. The case of a 60-year-old woman from our institution with acute-on-chronic visual disturbance secondary to visual pathway CM is presented. Including the current patient, 70 cases of anterior visual pathway CM have been published to our knowledge. The average patient age is 34.8 ± standard deviation of 14.2 years, with a female preponderance (n = 37, 52.9%). The majority of patients had an acute (n = 44; 62.9%; 95% confidence interval [CI] 0.51–0.73) onset of symptoms. In at least 55.6% (n = 40) of patients, the cause of visual disturbance was initially misdiagnosed. The majority (91.4%; n = 64) of patients underwent craniotomy, with complete resection and subtotal resection achieved in 53.1% (n = 34; 95%CI 0.41–0.65) and 17.2% (n = 11; 95%CI 0.10–0.28) of all surgical patients, respectively. Comparing surgically managed patients, complete resection improved visual deficits in 59.0% (n = 20; 95%CI 0.42–0.75), while subtotal resection improved visual deficits in 50.0% (n = 5; 95%CI 0.24–0.76; $p = 0.62$). CM is an important differential diagnosis for suprasellar lesions presenting with visual disturbance. A high index of suspicion is required in its diagnosis. Expeditious operative management is recommended to improve clinical outcomes.

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1. Introduction

Cavernous malformations (CM) are common, usually quiescent [1] vascular malformations of the central nervous system (CNS), with an incidence of 0.3% to 0.7% [1,2] and comprising 10% to 15% of all vascular malformations [1]. In the CNS, they occur with decreasing frequency in the supratentorial region (80%), infratentorial region (15%) and spinal cord (5%) [3]. Extra-axial CM are extremely rare. Symptomatic CM situated in the cerebral hemispheres commonly present with headaches, seizures, and/or focal neurological deficits [4,5]. Brainstem CM present more frequently and with more devastating outcomes compared to supratentorial CM [6].

Anterior visual pathway CM is a specific subset of CM, first reported in the literature by Uihlein et al. in 1958 [7]. CM of the anterior visual pathway affects a combination of the optic chiasm, optic nerve and anterior optic tracts. Due to the eloquence of affected tissues, CM of the anterior visual pathway typically presents with acute visual disturbance, termed "chiasmal apoplexy" by Maitland et al. [8] On MRI, anterior visual pathway CM show characteristic changes, with a hypointense to isointense signal on T1-weighted images, and a heterogeneous "popcorn" appearance on T2-weighted images with mixed bright and dark signals. This dark signal is accentuated in gradient-echo T2* sequences, due to a "blooming-effect" caused by haemosiderin deposition in and around the CM [9,10]. Nonetheless, these MRI findings, even when coupled with the clinical presentation, are not pathognomonic. CM of the anterior visual pathway, presenting as suprasellar lesions, are notoriously difficult to diagnose and commonly mistaken as craniopharyngioma, glioma or meningioma.

A systematic review of the current literature pertaining to CM of the anterior visual pathway was performed to clearly elucidate the clinical presentation, distribution, diagnostic certainty, treatment and clinical outcome. The findings are discussed in the context of

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http://dx.doi.org/10.1016/j.jocn.2014.07.027
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a 60-year-old woman with CM of the optic chiasm and left optic nerve. She presented with acute visual loss and underwent a pterional craniotomy and total resection at our institution.

2. Illustrative case report

2.1. History and examination

This 60-year-old woman presented with a 3 day history of headache and acute visual loss in the left eye. Ophthalmological examination revealed visual acuity of 6/9 on the right, and perception of hand movements only in the left eye. Confrontational visual field testing demonstrated a left-sided superior temporal visual field defect together with a left relative afferent pupillary defect and left optic disc pallor on fundoscopy. There were no visual field deficits in the right eye. She reported no other symptoms and there were no other focal neurological deficits.

The patient initially presented to an ophthalmologist 16 years prior with decreased visual acuity of 6/12 in the left eye. She was commenced on oral corticosteroids on a presumptive diagnosis of optic neuritis and experienced significant improvement of her visual acuity. She was subsequently lost to follow-up until the current presentation.

2.2. Investigation

Inflammatory markers and pituitary hormone levels were normal. MRI revealed a 15.0 mm × 13.8 mm × 12.2 mm suprasellar lesion involving the left optic nerve, optic chiasm and left optic tract. It was isointense and hyperintense on T1-weighted imaging, and isointense and hypointense on T2-weighted imaging with minimal contrast enhancement. There was increased signal loss on susceptibility-weighted imaging (Fig. 1). There was no evidence of flow on either magnetic resonance angiography or computed tomographic angiography. No other intracranial lesions were evident. A haemorrhagic chiasmal glioma, craniopharyngioma or an atypical meningioma were considered amongst the differentials.

2.3. Operation

A neuronavigated left pterional craniotomy was undertaken for definitive diagnosis and management. The left Sylvian fissure was split to expose the basal cisterns. Inspection under the operating microscope revealed an enlarged, oedematous left optic nerve with a bluish hue. The optic nerve sheath was incised, exposing an angiomatous lesion intimately involving the distal left optic nerve. The lesion extended to affect approximately one-third to one-half of the left-sided aspect of the optic chiasm anteroposteriorly and also the epicentre of the optic chiasm medially. The lesion was resected using microsurgical techniques. The underlying left optic nerve itself was atrophic. The optic tracts bilaterally and the contralateral right optic nerve was inspected and found to be free of any lesion. Extent of lesion involvement is diagrammatically illustrated in Supplementary Figure 1.

2.4. Histopathological findings

Histology revealed a CM in close association with neural tissue (Fig. 2). The lesion was composed of thin-walled cavernous channels, some capillaries, and occasional vessels with thicker hyaline walls but no arteries or veins (Fig. 3A). Some sections demonstrated luminal sclerosis and focal calcification of walls. Elastic–Van Gieson stain for elastin demonstrated no elastin fibres in the vessels. There was focal acute and chronic haemorrhage and perivascular lymphocytic cuffing in neural tissue around the malformation. One fragment showed necrosis of vessel walls and focal acute inflammation (Fig. 3B).

2.5. Postoperative course

The patient recovered well without any postoperative complications and was discharged on the sixth postoperative day. At 4 months post-surgery, her visual acuity remained unchanged. Humphrey Esterman binocular perimetry demonstrated an incomplete bitemporal hemianopia with relative sparing of the left superior temporal quadrant (Fig. 4).

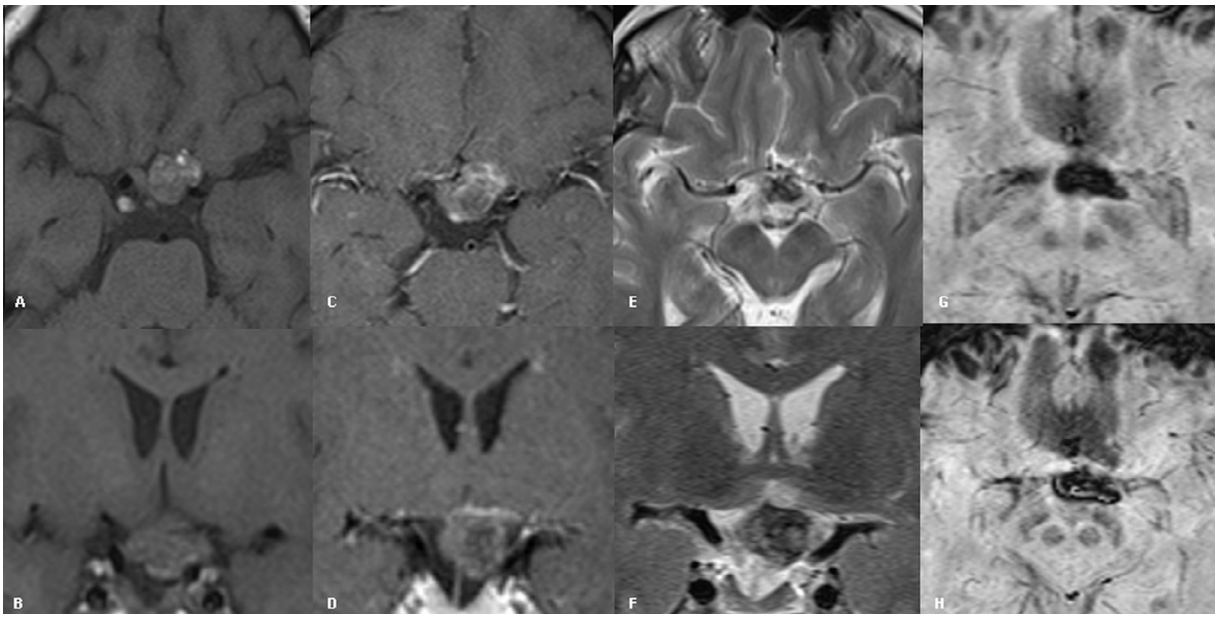

Fig. 1. Preoperative cerebral MRI demonstrating cavernous malformation involving the optic chiasm left optic nerve and left optic tract. Axial (A) and coronal (B) T1-weighted imaging of the chiasmatic and left optic nerve components with focal hyperintensity likely secondary to methaemoglobin. Axial (C) and coronal (D) T1-weighted gadolinium-enhanced imaging showing minimal enhancement. Axial (E) and coronal (F) T2-weighted imaging demonstrating heterogeneous hyperintensity and hypointensity consistent with blood products of differing ages. Axial (G, H) susceptibility-weighted imaging with increased signal loss due to paramagnetic blood products particularly peripherally due to haemosiderin-lined wall.

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