Cavernous Hemangioma of the Optic Chiasm: A Surgical Review

Matthew Crocker, M.R.C.S.,¹ Ruth DeSouza,² Andrew King, F.R.C.Path.,³ Steve Connor, F.R.C.R.,⁴ and Nick Thomas, F.R.C.S.(SN)⁵

ABSTRACT

Objectives: To report a new case of cavernous hemangioma of the optic chiasm and to review all previously published cases with regard to presentation, surgical treatment, and outcomes. **Design:** Case report and literature review. **Main Outcome Measures:** Cases identified through PubMed and published literature. Presentation states of patients in terms of visual loss and pituitary function. Surgical approaches, operations performed, and outcomes of the surgery. **Results:** thirty-nine previously reported cases were studied with the present case. All 40 patients presented with visual failure. Where documented, there was an 20% pituitary dysfunction rate. A total of 32 craniotomies were reported. Seventy-eight percent of patients underwent decompression including hematoma evacuation and partial or complete removal of the cavernoma, with improvement in visual function in 87% of these patients. The patients undergoing only biopsy showed stable visual function in 50% with further deterioration in 50%. **Conclusions:** We conclude that this rare condition can be managed with good outcomes in terms of visual improvement, provided a high index of suspicion is maintained and the goals of surgery for emergency patients are maintained to include chiasmal decompression. Although complete resection of the lesion is frequently possible, it should not be the primary aim of surgery.

KEYWORDS: Cavernoma, cavernous hemangioma, optic chiasm, apoplexy

Cavernous hemangiomas (CHM) are histologically benign vascular malformations composed of capillary tangles.¹ Although tethered to surrounding parenchyma, there is no involvement of parenchyma in the CHM itself.² CHM can occur anywhere in the brain and spinal cord but have been reported most frequently at subcortical sites on the

frontal and temporal lobes.³ The etiology of CHM may be spontaneous, syndromic, or familial, with the familial form often associated with multiple CHM.^{2,3} The familial form is reported in association with chiasmal CHM.⁴ CHM affects ~0.5% of the population, with only a small fraction of these having optochiasmal CHM. To date there are more

¹Department of Neurosurgery, St George's Hospital, London, United Kingdom; ²King's College, London, United Kingdom; Departments of ³Neuropathology, ⁴Neuroimaging and ⁵Neurosurgery, King's College Hospital, London, United Kingdom.

Address for correspondence and reprint requests: Matthew Crocker, M.R.C.S., Specialist Registrar, Department of Neurosurgery, St George's Hospital, Tooting, London SW17 0QT,

United Kingdom (e-mail: matthewcrocker@blueyonder.co.uk).

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than 30 reported cases of optochiasmal CHM, with the first described in 1958.⁵

Clinical presentation of CHM is determined by location. The frequency of asymptomatic cavernomas is widely debated at anything from 3 to 90%.⁶ Overall, the most common presentation of symptomatic CHM is seizures,^{7,8} but optochiasmal CHM has a different manifestation, most commonly episodic acute visual disturbance which may or may not improve.⁹ Subarachnoid hemorrhage and hematoma are also reported presentations of optochiasmal CHM.¹⁰ Optochiasmal CHM may present chronically with progressive visual loss, headache, and pituitary disturbance from recurrent hemorrhage. Signs of raised intracranial pressure and psychiatric disturbance are uncommon.¹¹ Symptoms may be precipitated by alcohol, pregnancy, and labor.¹⁰ The differential diagnosis of an optochiasmal CHM includes arteriovenous malformations, aneurysm, optic glioma, craniopharyngioma, other neoplasms, pituitary apoplexy, and infiltrative and inflammatory conditions.^{10,12}

Diagnosis of CHM using computed tomography (CT) or magnetic resonance imaging (MRI) scans may be incidental or following symptoms. CHM is rarely detected by conventional angiography. MRI is the preferred mode of imaging and characteristically shows a CHM as a central region of mixed signal intensities (indicating recurrent hemorrhage) surrounded by a rim of low-intensity signal of hemosiderin deposits in T1 and T2 images.^{11,12}

We report a case of optochiasmal CHM with the classic clinical and radiological findings. We review all previous cases in the available published literature with particular emphasis on the presentation, surgical management, and outcomes.

CASE REPORT

A 43-year-old man re-presented to our department with sudden visual failure. He had been under follow-up for the previous 7 years, having first presented with a mild bitemporal visual field defect

which had improved with conservative management. He had also been found to have pituitary dysfunction. Prior investigations had shown hypocortisolemia and growth hormone deficiency. He had been taking hormone replacement therapy.

Previous imaging investigations had identified a hypothalamic and posterior chiasmal lesion. This had been monitored with serial MRI over a 5-year period, the most recent being 18 months prior to the current presentation (Fig. 1). There had been static appearances to the lesion. The lesion returned mixed T1- and T2-weighted signal on MRI. No calcification or fat density was demonstrated on CT and subsequent CT angiography revealed no vascular abnormality. A craniopharyngioma was considered the most likely radiological diagnosis.

The patient was seen in a clinic for a routine appointment with a 1-week history of worsening visual failure with a bitemporal field defect. He was admitted and a repeat MRI scan was performed (Fig. 2). This showed no change in the hypothalamic/posterior chiasmal component of the lesion; however, there were now associated signal abnormality and expansion of the anterior optic chiasm and posterior optic nerves. These changes were consistent with hemorrhagic degradation products.

A subfrontal craniotomy was performed through a right eyebrow incision. There was evidence of subacute hemorrhage surrounding the arachnoidea mater of the optic chiasm, which was fenestrated and drained. A fleshy mass lesion with moderate vascularity immediately dorsal to the chiasm was entered and removed with only peripheral scar tissue left.

Following the surgery the patient's visual function was immediately improved. His pituitary function was unchanged. He made an uncomplicated recovery from the surgery.

Histopathologically, there was an abnormal collection of distended vascular channels, some lined by thickened eosinophilic walls (Fig. 3). There was surrounding reactive brain tissue including Rosenthal fibers and some hemosiderin-laden macrophages. The features were those of a CHM.

Figure 1 (A) T1-weighted sagittal and (B) T1-weighted sagittal post gadolinium images of the pituitary fossa, and (C) T2-weighted axial image of the brain. There is a hypothalamic/posterior chiasmal lesion that measures 1.4 cm in maximum axial dimension. It returns heterogeneous T2-weighted signal characterized by hyperintensity and areas of peripheral and punctate central hypointensity. Nodular areas of T1-weighted hyperintensity are demonstrated with minimal gadolinium enhancement. Craniopharyngioma was felt to be most likely, with partially thrombosed aneurysm, cavernoma, hemorrhagic glioma, and dermoid also being considered.

REVIEW OF PUBLISHED CASES

We searched PubMed for previously reported cases of optochiasmal CHM using a variety of search criteria. References from previous reports were then

cross-checked to ensure that no cases were missed. The published cases are tabulated (Table 1) in order of year of publication. Some papers reported more than one case and are denoted by alphabetical suffix. We have excluded cases in which there was no

Figure 2 (A) T1-weighted coronal and (B) T1-weighted sagittal post gadolinium images of the pituitary fossa, and (C) T2-weighted axial image of the brain (one section inferior to Fig. 1). There is no change to the hypothalamic/posterior chiasmal lesion relative to Fig. 1. There is a new area of signal abnormality more anteriorly. This returns T2-weighted hyperintensity with peripheral T2-weighted hypointensity and patchy central T1-weighted hyperintensity. This expands the optic chiasm and prechiasmal optic nerves without associated gadolinium enhancement. Features are consistent with hemorrhagic degradation products.

pathological confirmation of cavernoma and those diagnosed at post mortem, and we included the present case.

Individual cases were assessed for age and gender of patient and presentation mode, whether chronic, chronic with acute exacerbation (usually meaning an acute chiasmal syndrome), or acute only

(less than 3 weeks). The nature of the presentation in terms of impairment of visual acuity, visual field defect, and pituitary function was noted. The pre-operative suspected diagnosis and size of the lesion are included. The surgical approach and procedure in terms of total/subtotal resection are included as well as an approximate assessment of outcome in

Figure 3 Histology from the lesion reveals a cavernous hemangioma composed of numerous distended blood-filled channels (hematoxylin and eosin stain, magnification not available).

terms of visual improvement. Other important aspects are specified.

A total of 40 previous cases including our present case are identified from 32 previous reports and are tabulated (Table 1).^{2,5,9-40}

Of those traced, incomplete data were available in most cases, varying from age of patient (missing from only 2 of 40 cases) to pituitary function (missing from 29 of 40 cases).

There was slight female preponderance with 22 cases female, 15 male, 3 gender not reported. Mean age (reported in 38 of 40 cases) was 32 years (age range, 8 to 58 years).

The pattern of deterioration was established in all patients and classified as acute in 14 cases (35%), acute on chronic in 11 cases (28%), and chronic in 15 cases (38%). Visual failure was present in all cases with 35 having reduced visual acuity, 4 normal acuity, and 1 unreported, and 35 having a visual field deficit, 1 a normal field, and 4 unreported. Preoperative pituitary function was reported to be abnormal in only 2 cases and normal in 8.

A suspected diagnosis before surgery was offered in 27 patients with a mixture including retrobulbar neuritis (more common in the pre-MRI era), tumors both unspecified and craniopharyngioma, and a correct preoperative diagnosis of

CHM in 11 of 27 patients (41%). Lesion size ranged from 5 to 30 mm where reported.

The surgical approach used was documented in 32 patients. Only 1 patient underwent transphenoidal surgery¹⁵ and having failed to improve underwent left subfrontal craniotomy. Of the total 32 cranial approaches used, 7 (22%) were frontal or subfrontal (left or right), 17 (53%) were frontotemporal or pterional (left or right), 2 were anterior interhemispheric, 3 were orbitozygomatic, and 3 were unspecified craniotomy.

The surgical procedure was documented in 38 patients. Biopsy only was performed in 4 patients (11%). Biopsy and hematoma evacuation were performed in another 4 patients (11%). Partial removal was performed in 10 patients (26%) and total removal achieved in 21 patients (55%).

Postoperative visual function was recorded in 35 cases. Postoperative improvement in vision was seen in 29 patients (83%), stabilization of vision in 6 patients (17%), and worsening in 1. Of the 34 patients undergoing a decompression procedure (hematoma removal, partial or complete lesion removal), 29 (85%) experienced improvement in visual function, 4 were stable, and 2 visual outcomes were unreported.

Of the patients undergoing biopsy only, 2 (50%) subsequently experienced visual deterioration while the other 2 (50%) stabilized. Two patients subsequently received radiotherapy. One patient who had undergone subtotal resection of the lesion was reported as later showing recurrence. Diabetes insipidus was noted in 2 patients postoperatively.

DISCUSSION

Optochiasmal CHM is a most unusual variant of CHM. The presenting symptoms clearly range from asymptomatic lesions through patients with gradually progressive visual or pituitary disturbance to those patients presenting acutely with a chiasmic apoplexy syndrome. A range of treatment

Table 1 Summary of All Published Cases of Optochiasmal Cavernous Hemangioma

Authors	Age	S	Pres	VA	Fields	Pit	Dx	Size (mm)	Approach	Surgery	Vision	Notes
Uihlein and Rucker, 1958 ⁵	12	M	AoC	↓	↓	—	RBN	—	Craniotomy (unspecified)	Resection, extent unspecified	↑	Postoperative radiotherapy
Klein et al, 1979 ¹³	30	M	AoC	↓	↓	→	—	—	Right frontal craniotomy	Biopsy and hematoma evacuation	↑	
Mohr et al, 1985 ¹⁴	30	M	C	↓	↓	→	T	Small	Left frontotemporal	Decompression	↑	
Reilly and Oatey, 1986 ¹⁵	36	F	A	↓	↓	—	PA	—	Trans-sphenoidal followed by left subfrontal craniotomy	Biopsy and hematoma evacuation	↑	
Maruoka et al, 1988 ¹⁶	24	F	A	↓	↓	→	—	10	Right subfrontal craniotomy	Complete resection	↑	
Castel et al, 1989 ¹⁷	23	F	AoC	↓	→	—	—	6–8	Right frontotemporal	Biopsy and hematoma evacuation	→	
Corboy and Galetta, 1989 ¹⁸	44	F	AoC	↓	↓	—	RBN	—	Craniotomy	Biopsy only	↓	Initial visual improvement but subsequent worsening
Hassler et al, 1989 ¹⁹	24	F	C	→	↓	—	AN	5	—	Complete resection	↑	
Regli and de Tribolet, 1989 ²⁰	28	F	A	↓	↓	—	—	6	Pterional craniotomy	En bloc resection	↑	
Tien and Dillon, 1989 ²¹	32	F	C	↓	↓	—	—	15	Craniotomy	Biopsy then RXT	→	
Zentner et al, 1989 ²²	35	M	C	→	↓	—	AN	10	Right pterional craniotomy	Complete excision	↑	
Lejeune et al, 1990 ²³	26	F	C	↓	↓	—	CHM	—	Right subfrontal craniotomy	Biopsy and hematoma evacuation	↑	
Steinberg et al, 1990a ²⁴	58	M	A	—	↓	—	T	—	Right pterional craniotomy	Biopsy	→	No change in field deficit; VI nerve palsy improved
Steinberg et al, 1990b ²⁴	33	F	AoC	↓	↓	—	CHM	10	Right pterional craniotomy	Complete resection	↑	

Ferreira and Ferreira, 1992 ²⁵	8	M	A	↓	↓	—	—	—	Right frontotemporal craniotomy	Decompression and resection	→	
Hwang et al, 1993 ²⁶	42	M	AoC	↓	↓	→	T	—	Left frontotemporal craniotomy	Decompression	↑	
Perez-Lopez Fraile et al, 1995 ²⁷	25	F	C	↓	↓	→	—	—	—	Resection	↑	Perioperative thalamostriate infarct (recovered)
Shibuya et al, 1995a ²⁸	18	F	C	↓	↓	→	CP	30	Anterior interhemispheric	Complete excision	↑	Transient DI postop
Shibuya et al, 1995b ²⁸	60	F	A	↓	↓	—	CHM	5	—	Complete excision	↑	
Warner et al, 1996 ²⁹	32	F	AoC	↓	↓	—	CHM	13	Left pterional	Complete excision	→	
Arrue et al, 1999a ³⁰	—	—	C	↓	↓	—	CHM	20–30	Right frontopterional craniotomy	Partial removal	↑	
Arrue et al, 1999b ³⁰	—	—	AoC	↓	—	—	—	20–30	Left frontal craniotomy	Total removal	↑	
Iwai et al, 1999 ³¹	31	F	A	↓	↓	—	CHM	—	Right frontotemporal	Complete excision	↑	
Christoforidis et al, 2000 ³²	38	M	A	→	↓	—	CHM	—	—	—	—	
Elmaci et al, 2000 ³³	28	F	A	↓	↓	—	AVM	—	Right frontotemporal	Complete excision	↑	
Paladino et al, 2001 ³⁴	58	F	C	↓	↓	→	T	14 × 8	Right eyebrow subfrontal	Complete resection	↑	
Shaikh et al, 2002 ³⁵	42	M	A	↓	↓	—	T	—	Right pterional	Complete excision	↑	Diabetes insipidus postop
Deshmukh et al, 2003a ²	34	M	AoC	↓	—	—	—	—	Orbitozygomatic craniotomy	Complete excision	↑	Familial history
Deshmukh et al, 2003b ²	29	F	A	↓	—	—	—	—	Orbitozygomatic craniotomy	Subtotal resection	↑	Recurred
Deshmukh et al, 2003c ²	28	F	C	↓	↓	—	—	—	Orbitozygomatic craniotomy	Complete excision	↑	
Kehagias, 2003 ³⁶	27	M	C	↓	↓	—	—	—	—	Removal	—	
Glastonbury et al, 2003 ³⁷	25	F	A	↓	—	—	CHM	—	—	Complete excision	—	
Milea et al, 2005 ³⁸	29	F	C	↓	↓	—	—	—	—	Biopsy and complete resection	↑	
Shkarubo et al, 2005 ³⁹	28	—	A	↓	↓	↓	T	—	—	Clot removal and debulking	↑	
Lehner et al, 2006 ⁹	39	F	A	→	↓	—	CHM	—	Left pterional craniotomy	Complete excision	↑	

Table 1 (Continued)

Authors	Age	S	Pres	VA	Fields	Pit	Dx	Size	Approach	Surgery	Vision	Notes
Muta et al, 2006 ¹²	14	M	C	↓	↓	→	CP	15	Anterior basal interhemispheric	Biopsy	↓	Deterioration after postop bleed
Crocker et al, 2007 (present case)	43	M	AoC	↓	↓	↓	CP	14	Right eyebrow subfrontal	Decompression/debulking	↑	
Hempelmann et al, 2007 ¹¹	38	M	AoC	↓	↓	—	CHM	—	Pterional	Decompression/complete excision	↑	Transient visual deterioration postop
Ozer et al, 2007 ¹⁰	15	M	C	↓	↓	—	CHM	—	Left pterional craniotomy	Subtotal removal	↑	
Santos-Ditto et al, 2007 ⁴⁰	39	F	C	↓	↓	—	PA	—	Frontotemporal craniotomy	Complete resection	→	

S, sex of patient; Pres, presentation; VA, visual acuity; Pit, pituitary function on presentation; Dx, preoperative diagnosis; RBN, retrobulbar neuritis; T, tumor; PA, pituitary apoplexy (unspecified cause); AN, aneurysm; CHM, cavernous hemangioma; CP, craniopharyngioma; AVM, arteriovenous malformation; AoC, acute on chronic; C, chronic; A, acute.

—, unspecified; RXT, radiotherapy.

In visual deficits columns: ↑, improving; →, stable; ↓, worsening.

In pituitary function column: →, stable; ↓, worsening.

Papers reporting more than one case are denoted by alphabetical suffix.

strategies have been employed to deal with the variety of clinical scenarios that may arise.

An understanding of the natural history of optochiasmal CHM itself is vital to be able to formulate a rational management plan for these patients. The precise data in terms of year on year risk of hemorrhage are not known for cavernoma in this specific location and are unlikely to become so, given the rarity of the condition. There are no clear histological or radiological differences between optochiasmal CHM and those found elsewhere in the CNS. The differences in presentation therefore seem to arise simply due to the specific location of the lesion: this concept is not new.³ It is therefore reasonable to assume that these lesions have a similar natural history in terms of hemorrhage rate as those elsewhere in the central nervous system. This has been quoted as 0.6% annual risk of hemorrhage increasing to 4.5% for lesions that have previously bled.^{31,41} Of the cases reviewed and the new case presented here, there seems to be a far greater annual risk of hemorrhage overall than the risk proposed for CHM in general. This can likely be explained by the specific location of the lesions: smaller hemorrhages from CHM that would be undetected clinically if occurring in a cortical location are clinically more obvious if occurring in the optic chiasm. It may therefore be that the true hemorrhage rate for cavernomas in general is much higher than that quoted^{31,41} but simply that minor hemorrhages from such cavernomas are more likely to be clinically silent. A similar hypothesis has been proposed to explain the more aggressive behavior of spinal cord cavernoma.⁴² This hypothesis would support the higher rates of hemorrhage documented for brainstem cavernomas as well.⁴³

Chiasmal compression syndromes from hemorrhagic pituitary tumors probably form the most comparable clinical situation for which good outcome data are available. Emergency surgery for so-called pituitary apoplexy from a hemorrhagic tumor has been reported as having a good outcome in terms of visual function,⁴⁴ although comparable outcomes for the visual loss in pituitary apoplexy have been found in surgically and nonsurgically

treated patient groups,⁴⁵ and conservative strategies have therefore been recommended for pituitary apoplexy patients with nonprogressive visual symptoms.⁴⁶ However, this series also reported a relatively low incidence (22%) of recurrence of the pituitary tumor in the nonsurgical group of patients. Clearly, in optochiasmal CHM the prospects for recurrent hemorrhage in the future are high, based on the propensity for recurrent small hemorrhages, and a more aggressive surgical approach is likely to be necessary. Lehner and associates felt that an aggressive surgical strategy aimed at total resection was justified as the primary treatment.⁹

The cases reviewed have covered many different treatment strategies. There are the most conservative approaches, which are analogous to no surgery at all. The natural history of severe perichiasmal hemorrhage from CHM is severe visual deficit that may partially recover; however, such patients are most likely to have subsequent minor hemorrhages with continuing incremental visual decline. The presented literature shows a range of outcomes from surgery for chiasmal syndromes from CHM: there are few instances of worsening deficit from surgery with most patients experiencing partial improvement, usually with substantial functional gain.

One operative strategy employed has been that of simple biopsy.^{11,12} This strategy has been justified simply to minimize operative risk. Our experience with the present case is that a more extensive procedure to decompress the chiasm while obtaining a pathological diagnosis should be possible by the skilled surgeon with appropriate microsurgical experience. We do not believe that simple biopsy in the context of acute visual failure can be considered an adequate operation. Unlike other causes of chiasmal compression such as prolactinoma, active nonsurgical treatments are not available. Biopsy may therefore be performed with perioperative diagnostic histopathology to guide the rest of the procedure, but it should not be performed as an operation in its own right for optochiasmal CHM, especially in the context of an acute deterioration. In addition, biopsy has been

proposed as increasing the likelihood of bleeding from optochiasmal CHM.¹⁰

All operative strategies have involved craniotomy of some form. Approaches used have been a variety of subfrontal approaches including eyebrow, orbitozygomatic, and more lateral approaches, including pterional. The surgical approach must be dictated by the laterality of the lesion: for more lateral lesions a pterional approach may be adequate.

It is interesting to note that in the present case and two of those reviewed, although craniopharyngioma was often a presupposed radiological diagnosis, trans-sphenoidal surgery was only undertaken once, in the pre-MRI era.¹⁵ This is presumably because similar to our case, although these cavernomas were considered to be craniopharyngiomas by virtue of their signal characteristics and symptoms at presentation, they were all in a suprasellar location without sellar expansion. The case in which initial surgery was by the trans-sphenoidal route was followed by subfrontal craniotomy 2 weeks later when the clinical state of the patient and the CT findings were unchanged. We feel it likely that such patients would have a poor outcome from trans-sphenoidal surgery in terms of difficulty with adequate chiasmatic decompression and would therefore not advocate it. A similar rationale for chiasmal arteriovenous malformations has been proposed.⁴⁷

Chiasmal decompression in cases of optochiasmal CHM will rely on two aspects: drainage or evacuation of hematoma from recent hemorrhage, and removal of the cavernoma itself. Several of the reviewed cases have reported drainage of subacute hematoma either in isolation or combined with partial or complete resection of the cavernoma itself. We advocate that in patients presenting with apoplectic symptoms, a procedure with minimal manipulation of an optic apparatus already at risk should be the goal, to minimize the prospects of iatrogenic visual deficit. In this context we would therefore advise drainage or evacuation of the hematoma, biopsy to confirm diagnosis, and resection of part or all of the cavernoma only if the pathologic anatomy proved highly favorable. If any doubt exists

over the extent of manipulation required, then the resection of the cavernoma should be avoided or at least deferred until visual stability is confirmed. Because a limited lesion resection would be preferred in this context, a direct and less extensive cranial approach may be used, most probably a subfrontal approach or variant.

There are, however, very serious implications for the partially treated cavernoma in this context. The cavernoma remnant must be considered to have a risk of symptomatic bleeding, and some patients, especially those with good visual outcomes, will merit further treatment of the remnant. One of the reviewed patients went on to have postoperative radiotherapy, and did not have long-term recurrence.⁵ Stereotactic radiotherapy has been used to treat cavernomas throughout the brain with good results. However, there are recognized tolerance limits of the anterior visual pathway beyond which risks of optic neuropathy increase unacceptably.⁴⁸ There is a lack of data to support firm conclusions regarding this specific problem which could arise in such patients, and all that can be said is that treating cavernoma remnants nonsurgically (i.e., with stereotactic radiotherapy) in this region may be more difficult than in other regions of the brain. We still feel strongly that the risks of an aggressive complete resection for the patient in chiasmal distress are very high and in most cases the definitive surgery can be performed in an elective manner, if necessary. The prospect of this is apparently low, as few of the cases in which subtotal resection was performed have been reported as having recurrent or residual components requiring treatment. However, results of partial removal of cavernomas in the brainstem have shown increased risk of hemorrhage; thus there is a fine line between chiasmal manipulation to achieve complete resection and incomplete surgery.⁴⁹

The elective procedure for the patient without an acute event is more difficult to approach. Clearly, in this context, acute hematoma evacuation will not be performed and hence chiasmal decompression may only be achieved by resection of part or all of the cavernoma. In this context the surgical

approach must be generous and should involve an extended subfrontal approach, perhaps including orbitozygomatic craniotomy. This may be extended pterionally in the case of a chiasmal cavernoma with a major anterior and unilateral component.

None of the cases reviewed mentioned the growing use of endoscopic techniques in the surgery for this region.⁵⁰ Given that we have advocated a less extensive approach and a more simple decompressive surgery for the patient presenting with an acute syndrome, we feel that endoscope-assisted surgery may have a role in the management of the acutely presenting patient. As such techniques grow in popularity it is likely that such a case will be managed by an endoscopically adept surgeon and we look forward to hearing of the challenges posed and outcomes.

CONCLUSIONS

We have presented a new case of optochiasmal CHM and reviewed a further 40 from published work. We find that good outcomes can be achieved in terms of improvement of visual deficit with craniotomy and microsurgical decompression and resection of the cavernoma if anatomical factors allow. A good outcome for the patient will depend on maintaining an index of suspicion for this rare diagnosis in the patient presenting with chronic stepwise visual failure.

REFERENCES

1. Lobato RD, Perez C, Rivas JJ, Cordobes F. Clinical, radiological, and pathological spectrum of angiographically occult intracranial vascular malformations: analysis of 21 cases and review of the literature. *J Neurosurg* 1988;68: 518–531
2. Deshmukh VR, Albuquerque FC, Zabramski JM, Spetzler RF. Surgical management of cavernous malformations involving the cranial nerves. *Neurosurgery* 2003;53:352–357
3. Moriarity JL, Clatterbuck RE, Rigamonti D. The natural history of cavernous malformations. *Neurosurg Clin N Am* 1999;10:411–417

4. Malik S, Cohen BH, Robinson J, Fried A, Sila CA. Progressive vision loss: a rare manifestation of familial cavernous angiomas. *Arch Neurol* 1992;49:170-173
5. Uihlein A, Rucker CW. The neurosurgeon's role in acute visual failure. *AMA Arch Ophthalmol* 1958;60:223-229
6. Brunon J, Nuti C. Natural history of cavernomas of the central nervous system. *Neurochirurgie* 2007;53(2-3):122-130
7. Voigt K, Yaargil MG. Cerebral cavernous haemangiomas or cavernomas—incidence, pathology, localization, diagnosis, clinical features and treatment: review of the literature and report of an unusual case. *Neurochirurgia (Stuttg)* 1976;19:59-68
8. Awad I, Jabbour P. Cerebral cavernous malformations and epilepsy. *Neurosurg Focus* 2006;21:e7
9. Lehner M, Fellner FA, Wurm G. Cavernous haemangiomas of the anterior visual pathways: short review on occasion of an exceptional case. *Acta Neurochir (Wien)* 2006;148:571-578; discussion 578
10. Ozer E, Kalemci O, Yucesoy K, Canda S. Optochiasmatic cavernous angioma—unexpected diagnosis: case report. *Neurol Med Chir (Tokyo)* 2007;47:128-131
11. Hempelmann RG, Mater E, Schroder F, Schon R. Complete resection of a cavernous haemangioma of the optic nerve, the chiasm, and the optic tract. *Acta Neurochir (Wien)* 2007;149:699-703
12. Muta D, Nishi T, Koga K, Yamashiro S, Fujioka S, Kuratsu J. Cavernous malformation of the optic chiasm: case report. *Br J Neurosurg* 2006;20:312-315
13. Klein LH, Fermaglich J, Kattah J, Luessenhop AJ. Cavernous hemangioma of optic chiasm, optic nerves and right optic tract: case report and review of literature. *Virchows Arch A Pathol Anat Histol* 1979;383:225-231
14. Mohr G, Hardy J, Gauvin P. Chiasmal apoplexy due to ruptured cavernous hemangioma of the optic chiasm. *Surg Neurol* 1985;24:636-640
15. Reilly PL, Oatey PE. Optic nerve apoplexy: report of two cases. *J Neurosurg* 1986;64:313-316
16. Maruoka N, Yamakawa Y, Shimauchi M. Cavernous hemangioma of the optic nerve: case report. *J Neurosurg* 1988;69:292-294
17. Castel JP, Delorge-Kerdiles C, Rivel J. Cavernous angioma of the optic chiasm. *Neurochirurgie* 1989;35:252-256
18. Corboy JR, Galetta SL. Familial cavernous angiomas manifesting with an acute chiasmal syndrome. *Am J Ophthalmol* 1989;108:245-250
19. Hassler W, Zentner J, Petersen D. Cavernous angioma of the optic nerve: case report. *Surg Neurol* 1989;31:444-447
20. Regli LP, de Tribolet N. Chiasmatic apoplexy caused by rupture of a cavernoma. *Neurochirurgie* 1989;35:102-103
21. Tien R, Dillon WP. MR imaging of cavernous hemangioma of the optic chiasm. *J Comput Assist Tomogr* 1989;13:1087-1088
22. Zentner J, Grodd W, Hassler W. Cavernous angioma of the optic tract. *J Neurol* 1989;236:117-119
23. Lejeune JP, Hladky JP, Dupard T, Parent M, Hache JC, Christiaens JL. Optic chiasmatic apoplexy. *Neurochirurgie* 1990;36:303-307
24. Steinberg GK, Marks MP, Shuer LM, Sogg RL, Enzmann DR, Silverberg GD. Occult vascular malformations of the optic chiasm: magnetic resonance imaging diagnosis and surgical laser resection. *Neurosurgery* 1990;27:466-470
25. Ferreira NP, Ferreira MP. Optic nerve apoplexy caused by a cavernous angioma: case report. *Neurosurgery* 1992;30:262-264
26. Hwang JF, Yau CW, Huang JK, Tsai CY. Apoplectic optochiasmal syndrome due to intrinsic cavernous hemangioma: case report. *J Clin Neuroophthalmol* 1993;13:232-236
27. Pérez López-Fraile I, Tapiador Sanjuán MJ, Eiras Ajuria J, Giménez Mas JA. Cerebral cavernous angiomas in pregnancy: two cases and a review of literature. *Neurologia* 1995;10:242-245
28. Shibuya M, Baskaya MK, Saito K, Suzuki Y, Ooka K, Hara M. Cavernous malformations of the optic chiasma. *Acta Neurochir (Wien)* 1995;136(1-2):29-36
29. Warner JE, Rizzo JF III, Brown EW, Ogilvy CS. Recurrent chiasmal apoplexy due to cavernous malformation. *J Neuroophthalmol* 1996;16:99-106
30. Arrue P, Thorn-Kany M, Vally P, et al. Cavernous hemangioma of the intracranial optic pathways: CT and MRI. *J Comput Assist Tomogr* 1999;23:357-361
31. Iwai Y, Yamanaka K, Nakajima H, Miyaura T. Cavernous angioma of the optic chiasm: case report. *Neurol Med Chir (Tokyo)* 1999;39:617-620
32. Christoforidis GA, Bourekas EC, Baujan M, Drevelengas A, Tzalonikou M. Neuroradiology case of the day: case 1—cavernous hemangioma of the optic chiasm. *AJR Am J Roentgenol* 2000;175:888-891
33. Elmaci I, Ates G, Kurtkaya O, Necmettin Pamir M. Chiasmal cavernous malformation: a rare cause of acute visual loss. *J Neurosurg Sci* 2000;44:226-229
34. Paladino J, Rotim K, Pirker N, Glunčić V, Jurić G, Kalauz M. Minimally invasive treatment of cavernous angioma of the optic chiasm: case report. *Minim Invasive Neurosurg* 2001;44:114-116
35. Shaikh A, Benjamin L, Kerr R. Chiasmal cavernous angioma: a rare case of progressive visual loss. *Eye* 2002;16:655-657
36. Kehagias DT. A case of headache and disordered vision: cavernous hemangioma of the optic chiasm (2003:8b). *Eur Radiol* 2003;13:2552-2553
37. Glastonbury CM, Warner JE, MacDonald JD. Optochiasmal apoplexy from a cavernoma. *Neurology* 2003;61:266
38. Milea D, Karachi C, Bonneville F, Mokhtari K, Lehoang P, van Effenterre R. Optochiasmal cavernoma: a rare cause of unilateral visual loss. *Rev Neurol (Paris)* 2005;161:605-607
39. Shkarubo AN, Serova NK, Tropinskaia OF, Shishkina LV, Pronin IN. Chiasmatic cavernoma. *Zh Vopr Neirokhir Im N N Burdenko* 2005;20-21; discussion 21-22

40. Santos-Ditto RA, Santos-Franco JA, Pinos-Gavilanes MW. Cavernous angioma of the second cranial nerve and chiasmatic apoplexy. *Neurocirugia (Astur)* 2007;18:47–51
41. Kondziolka D, Lunsford LD, Kestle JR. The natural history of cerebral cavernous malformations. *J Neurosurg* 1995;83:820–824
42. Deutsch H, Jallo GI, Faktorovich A, Epstein F. Spinal intramedullary cavernoma: clinical presentation and surgical outcome. *J Neurosurg* 2000;93(suppl):65–70
43. Ferroli P, Sinisi M, Franzini A, Giombini S, Solero CL, Broggi G. Brainstem cavernomas: long-term results of microsurgical resection in 52 patients. *Neurosurgery* 2005;56:1203–1212; discussion 1212–1214
44. Zhang X, Fei Z, Zhang W, et al. Emergency trans-sphenoidal surgery for hemorrhagic pituitary adenomas. *Surg Oncol* 2007;16:115–120
45. Gruber A, Clayton J, Kumar S, Robertson I, Howlett TA, Mansell P. Pituitary apoplexy: retrospective review of 30 patients—is surgical intervention always necessary? *Br J Neurosurg* 2006;20:379–385
46. Sibal L, Ball SG, Connolly V, et al. Pituitary apoplexy: a review of clinical presentation, management and outcome in 45 cases. *Pituitary* 2004;7:157–163
47. Lavin PJ, McCrary JA III, Roessmann U, Ellenberger C Jr. Chiasmal apoplexy: hemorrhage from a cryptic vascular malformation in the optic chiasm. *Neurology* 1984;34:1007–1011
48. Kenai H, Yamashita M, Nakamura T, Asano T, Sainoh M, Nagatomi H. Tolerance dose in gamma knife surgery of lesions extending to the anterior visual pathway. *J Neurosurg* 2005;102(suppl):230–233
49. Esposito P, Coulbois S, Kehrli P, et al. Place of the surgery in the management of brainstem cavernomas: results of a multicentric study. *Neurochirurgie* 2003;49:5–12
50. Kabil MS, Shahinian HK. Application of the supraorbital endoscopic approach to tumors of the anterior cranial base. *J Craniofac Surg* 2005;16:1070–1074