# Chiasmal Apoplexy Due to Ruptured Cavernous Hemangioma of the Optic Chiasm

Gérard Mohr, M.D., F.R.C.S. (C), Jules Hardy, M.D., F.R.C.S. (C), F.A.C.S., and Pierre Gauvin, M.D., F.R.C.S. (C)

Departments of Neurosurgery and Ophthalmology, Notre-Dame Hospital and University of Montreal, Montreal, Canada

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A 30-year-old obese patient with a rapidly progressing chiasmal syndrome presented with acute bitemporal hemianopia, severe bilateral amblyopia, mental confusion, and lethargy. X-ray films of the skull showed a normal sella turcica and computed tomography scan as well as angiography revealed a vascular mass within the chiasmatic cistern. At operation, via left subfrontal approach, an intrachiasmal hematoma was evacuated and biopsies of the hematoma cavity revealed a cavernous hemangioma. Visual symptoms improved markedly postoperatively. This rare association of chiasmal apoplexy and intrachiasmal cavernoma is discussed.

KEY WORDS: Chiasmal apoplexy; Cavernous hemangioma; Chiasmal syndrome; Cavernoma

Cavernous hemangiomas (or cavernomas) are rare vascular malformations composed by venous channels without intervening nervous parenchyma. They are often silent but, at times, may rupture and lead to severe intracranial hemorrhage [18]. We report a case in which a cavernous hemangioma of the optic chiasm led to an acute chiasmal compression. Surgical findings and literature were reviewed.

## **Case Report**

A 30-year-old male patient with long-lasting obesity first complained in May 1979 (2 years prior to admission) of transient bilaterally blurred vision for about 6 weeks. In November 1980, he experienced another similar episode with transient decrease of visual acuity. At the end of January 1981, he again complained of similar ocular symptoms, which were then accompanied by temporooccipital headaches and generalized weakness. The first neuroophthalmologic examination on February 18, 1981, revealed the following: visual acuity—right eye, with correction 20/200, left eye, with correction 20/60; visual fields: hemianopic scotoma of the left eye and inferior temporal quadranopia of the right eye (Figure 1). The optic fundi were normal. Plain x-ray films of the skull revealed no abnormalities, except for a slight sellar enlargement. The optic foramina were not enlarged. The clinical impression at that time was chiasmal syndrome vs. hysteria.

The second ophthalmologic examination, performed on April 1, 1981, revealed the following changes: visual acuity O.D. 20/60, visual fields: enlargement of the left temporal scotoma and now complete right-sided hemianopia. The patient was admitted to Notre-Dame Hospital on April 20, 1981, because of headaches, rapid decrease of vision, mental confusion, and aggressive behavior for about 10 days. According to his family, the patient had shown hypersomnia for 6 months. On examination, the patient was obese, with slight gynecomastia without galactorrhea. He was stuporous, and at times considerably agitated and confused. Complete endocrine evaluation failed to show any hypothalamopituitary deficiency. Differentiated ophthalmologic examination was impossible because of lack of cooperation, but complete bitemporal hemianopia was suspected. The computed tomography (CT) scan showed a suprasellar mass enhancing with contrast and compressing the anterior portion of the third ventricle (Figure 2). Left carotid angiogram showed a vascular density in the suprasellar area, compatible with a small meningioma of the jugum sphenoidale or a tumorous process of the optic chiasm extending into the lamina terminalis, such as an optic glioma with recent hemorrhage. On April 30, 1981, a left frontotemporal craniotomy was performed; the spinal fluid was xanthochromic. The left

Address reprint requests to: Dr. Gérard Mohr, Division of Neurosurgery, Notre-Dame Hospital, 1560 Sherbrooke Street East, Montreal, H2L 4M1, P.Q., Canada.

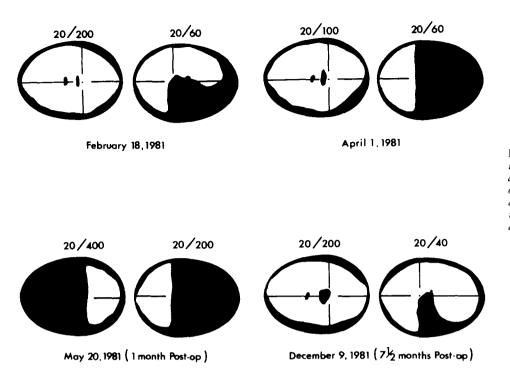
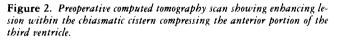


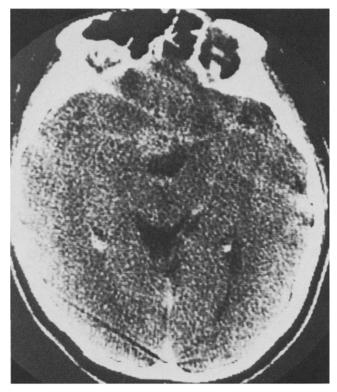
Figure 1. Sequential visual fields showing rapid progression of chiasmal syndrome until acute bitemporal hemianopia occurred immediately preoperatively. Seven and one-half months postoperatively, there was remarkable improvement of visual fields and visual acuity.

optic nerve was exposed and found to be massively enlarged. The optic chiasm was dilated and the left optic tract was also considerably swollen. There was evidence of an intrinsic bluish mass inside the chiasm. The pellucid membrane was entered and dark fluid blood escaped under pressure (Figure 3). Biopsies were taken from the wall of this cystic cavity. The postoperative course was satisfactory, despite some transient agitation requiring sedation. Microscopic examination revealed the presence of a cavernous hemangioma consisting of several venous channels separated by fibrous septa providing evidence of previous hemorrhages (siderophages); some veins contained organizing thrombi (Figure 4). The mental confusion disappeared gradually, and the patient was discharged 4 months postoperatively, with a visual acuity of 20/40 (right eye) and 20/200 (left eye). The chiasmatic syndrome had largely resolved, leaving a right temporal quadranopia and a left residual central scotoma with marked atrophy on the left side (Figure 1). These findings have remained stable since that time. The repeat CT scan performed 1 year later shows no evidence of residual malformation.

## Discussion

Cavernous hemangiomas are considered to be congenital blood vessel hamartomas occuring mostly in the cerebral hemispheres and (in decreasing order of incidence) in the basal ganglia, brainstem, and cerebellar hemispheres. They may be multiple and may occur outside the central nervous system in the lungs, kidneys, and liver [16]. Clinically, these lesions may present with symptoms of headache, sometimes diagnosed as pseudotumor cerebri [15]; epilepsy, mostly focal seizures





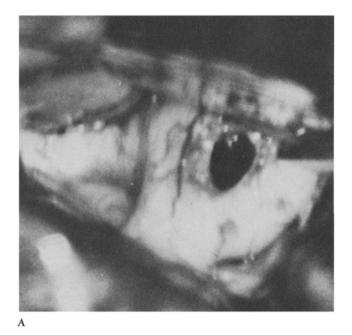
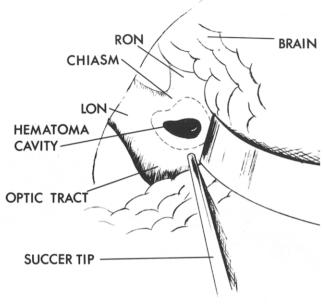
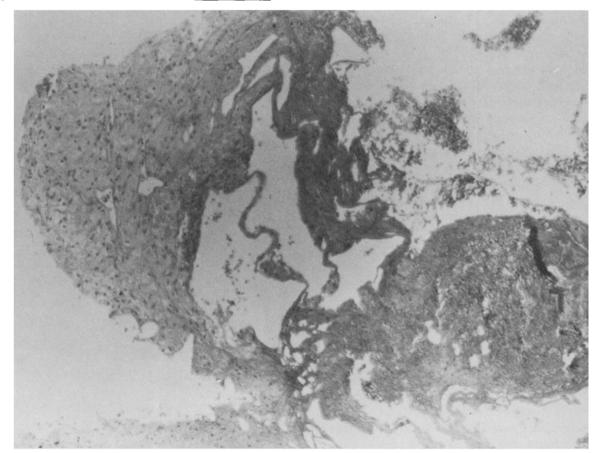


Figure 3. Operative photography (A) and sketch (B) showing massively swollen optic chiasm and left optic nerve ( $\underline{LON}$ ) and the hematoma cavity within the optic chiasm.

Figure 4. Photomicrograph of biopsy specimen taken from wall of bematoma cavity (<u>HE</u>,  $\times 100$ ), showing venous channels (<u>center</u>) separated by fibrous septae, and containing organizing thrombus (<u>right lower corner</u>).



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[11]; hydrocephalus, and intracranial hemorrhage [7]. They usually become symptomatic during the second and third decade of life, and they have a marked tendency to become symptomatic during pregnancy, due to vascular engorgement.

Radiologically, the cavernomas show calcifications on plain skull films in 25% of the cases; dense calcifications have led to the term "hemangioma calcificans." On CT scan, they appear as well-demarcated, round-shaped densities containing various amounts of calcium with mild contrast enhancement and usually no significant mass effect [6,14]. At angiography, there is absence of enlarged feeding vessels and little or no venous drainage, but a vascular blush can be observed at the capillary phase, particularly if magnification is used [4,14].

Pathologically, the cavernous hemangiomas are characterized by a dense aggregation of blood vessel sinusoids consisting of irregular venous channels separated by thin septa: The vascular walls are lined by endothelium and contain no smooth muscle or elastic fibers, and the septa are formed by fibrous, collagenous stroma. Contrary to arteriovenous malformations, there is absence of intervening nervous or glial tissue between the sinusoids.

Cavernous hemangiomas involving the visual system are extremely rare [4] and we were only able to identify two reports in the literature that could be compared to our case. Uihlein and Rucker [16,17], in 1958, reported the case of a 12-year-old boy with rapidly progressing chiasmal symptoms, headaches, and lethargy. At emergency craniotomy, an acute hemorrhage within a hemangioendothelioma of the left optic nerve and the left half of the optic chiasm was evacuated, resulting in marked improvement of vision in the right eye. In 1979, Manz et al [9] reported a 30-year-old patient with rapid decrease of vision in his right eye for 3 weeks, along with bilateral scotomata and bitemporal hemichromatopsia, who then developed a Marcus-Gunn pupil and left temporal hemianopia with subarachnoid hemorrhage. This patient underwent surgery and was found to have a large cavernous hemangioma of the optic chiasm extending into the optic nerves and right optic tract.

Chiasmal apoplexy, characterized by an acute chiasmal syndrome with intrachiasmatic hemorrhage, has been described as a clinical entity by Maitland et al [8]. These investigators reported four patients with acute chiasmal syndrome related to intrachiasmal hemorrhages, three of which had cryptic vascular lesions (that is, negative biopsies at operation), and one an optic glioma with secondary hemorrhage. A pure intrachiasmatic arteriovenous malformation of the optic chiasm was reported by Roski et al in 1981 [13].

Most acute chiasmal syndromes are related to suprasellar tumors with intratumoral hemorrhages. In such instances, acute pituitary apoplexy [10], due to intraadenomatous hemorrhage, sometimes necrosis, will be suspected on x-ray studies of the skull showing sellar enlargement and acute pituitary failure, often accompanied by severe extraocular motor disturbances, due to acute cavernous sinus compression. Rapid transsphenoidal decompression will lead to recovery of visual function and normal consciousness. In cases where x-ray studies fail to demonstrate any sellar enlargement, intrachiasmal apoplexy must be suspected in the presence of an acute chiasmal syndrome without extraocular motor disturbances, and craniotomy with a subfrontal approach to the optochiasmatic region must be considered without delay. Acute chiasmal syndromes may otherwise be related to optic gliomas, intrachiasmatic craniopharyngiomas [5], or acute demyelination, such as observed in Devic's neuromyelitis optica. Large suprasellar aneurysms originating from the anterior cerebral-anterior communicating complex or from the carotid-ophthalmic junction may also mimic acute chiasmal syndromes, and cerebral angiography remains an important part of the diagnosis of all rapidly evolving chiasmal syndromes.

#### Summary and Conclusion

Chiasmal apoplexy, characterized by acute hemorrhagic intrachiasmal compression, severe psychic changes with mental confusion, memory disturbances due to distension of the mamillary bodies and basal hypothalamus, and severe headaches due to subarachnoid hemorrhage were recognized as a distinct clinical entity by Maitland et al in 1982. We report a case of ruptured cavernous hemangioma of the optic chiasm who presented a typical chiasmal apoplexy with an acute chiasmal syndrome, mental confusion, and subarachnoid hemorrhage. Rapid decompression by craniotomy and subfrontal approach with biopsies of the wall of the chiasmatic hematoma cavity ensured the diagnosis and was followed by important visual improvement.

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