

Optic chiasma cavernous malformation

Epidemiology

Suprasellar occurrences of [cavernous malformations](#) (CM) in the [optic chiasm](#) are extremely uncommon, representing less than 1% of all CNS CMs ^{1) 2)}.

To the best of the knowledge of Abou-Al-Shaar et al. less than 80 cases have been reported in the literature ³⁾.

Clinical features

Patients with these lesions typically present with [chiasmal apoplexy](#), characterized by sudden [visual loss](#), [acute headaches](#), [retroorbital pain](#), and [nausea](#) ⁴⁾.

These symptoms typically occur after a period of transient blurry vision and headaches. In addition, [hypopituitarism](#) from direct compression of the [pituitary stalk](#) has been reported in the literature ^{5) 6)}.

Diagnosis

On CT scan, optic pathway CMs appear as well-demarcated hyperdense lesions with or without calcifications ⁷⁾.

MRI

MR imaging of cavernous hemangioma of the optic chiasm ⁸⁾

MRI is considered the most sensitive and specific imaging modality for the diagnosis of CM ⁹⁾.

On T1-weighted images, CMs of the optic pathway demonstrate a hypointense to isointense appearance, whereas on T2-weighted images, they appear as heterogeneous “popcorn” lesions with mixed hyperintense and hypointense signals.

The [Hypointensity](#) can be delineated further in the gradient-echo T2* images due to [hemosiderin](#) deposition in and around the CM. In addition, following intravenous gadolinium administration, minimal or no enhancement can be observed in the CM ^{10) 11)}.

It has been reported that CMs of the optic nerve and tract may show nerve thickening on coronal views, whereas CMs of the optic chiasm often appear as focal round masses ¹²⁾.

Angiography is usually not helpful in diagnosing CMs because it does not delineate the lesion due to the low internal flow and high incidence of thrombosis ¹³⁾.

Differential diagnosis

CMs of the optic pathway are commonly misdiagnosed as [optic neuritis](#), [optic glioma](#), [meningioma](#), [craniopharyngioma](#), [venous angioma](#), [arteriovenous malformation](#), thrombosed [intracranial aneurysm](#), and [pituitary apoplexy](#), [histiocytosis](#), [hypothalamic glioma](#), [tuber cinereum hamartoma](#) and [metastasis](#) ^{14) 15)}.

Cavernoma should be considered when a solid suprasellar mass has hemorrhage (mimicking cystic-adamantinomatous craniopharyngioma).

Cavernoma and suprasellar meningioma are rarely associated. Holland and Symon report a patient, whose recovery after removal of the meningioma was complicated by haemorrhage from the cavernoma. This occurrence has not been previously reported ¹⁶⁾.

Treatment

Surgical removal is the recommended treatment to restore or preserve vision and to eliminate the risk of future hemorrhage. However, the anatomical location and eloquence of nearby neural structures can make these lesions difficult to access and remove.

The surgical approach should allow optimal exposure of the lesion using the shortest route and with minimal brain retraction. Various surgical approaches have been reported in the literature including pterional, orbitozygomatic, supraorbital, subfrontal, and transbasal interhemispheric approaches. Almost half of the cases reported in the literature were managed through the frontotemporal approach ¹⁷⁾.

Biopsy is contraindicated for these lesions due to the high risk of bleeding and symptomatic worsening ^{18) 19)}.

Reviews

2006

In their meticulous review of the literature, Lehner et al. found 42 previously reported patients with vascular malformations within optic nerves, chiasm, or optic tracts, 30 of them being cavernous hemangiomas. The optic chiasma was involved in 38 patients (90.5%) and a total excision of the tumor was performed in 21 cases ²⁰⁾.

Case reports

2016

A 33-year-old female presented 3 months postpartum with a headache of moderate severity and progressive visual loss in both eyes. On examination, the patient's Glasgow coma scale (GCS) was 15/15. Visual field examination showed left homonymous incomplete hemianopia. Her visual acuity was 20/25 in the right eye and 20/30 in the left eye. Her discs and macula were healthy bilaterally. Extraocular movements were intact and pupils were reactive. The rest of her examination was unremarkable. Complete endocrine workup was normal.

Magnetic resonance imaging (MRI) revealed a large heterogeneous, hyperintense, hemorrhagic right suprasellar extra-axial complex cystic structure measuring 31 × 30 × 90 mm on T1-weighted images. There was mass effect on the adjacent hypothalamus and third ventricle displacing them toward the left and superiorly in addition to the optic pathway. The pituitary stalk was displaced toward the left. The lesion encased the right posterior cerebral artery and displaced the right carotid artery laterally.

Computed tomography (CT) arteriography demonstrated a completely thrombosed center. The imaging findings were compatible with suprasellar CM.

The patient underwent right frontal craniotomy and gross total resection of her suprasellar intrachiasmatic large infiltrative hemorrhagic CM. Organizing blood clots with reactive fibrohistiocytic and inflammatory reaction admixed with some ectatic vascular channels suggestive of a vascular malformation were noted. There were small foci admixed with granulation tissue, showing some dilated cavernous spaces that would be compatible with a vascular malformation such as cavernous angioma. On immunohistochemistry, the lesion was CD163+, CD20 rare, CD3+, CD34+, CD31+, CD38+, CTK–, EMA plasma cells, GFAP–, S100 dendritic cells, SMA vascular smooth muscle.

The patient had an uneventful operative course. Her visual acuity improved to 20/20 in both eyes. [Extraocular muscles](#) showed mild limitation of both eyes in an upward gaze. Otherwise, she was stable with no neurological deficits. Follow-up MRI at 12 months revealed complete removal of the suprasellar hemorrhagic CM with no evidence of a residual lesion or recurrence ²¹⁾.

Cavernous malformation of the optic chiasm: Neuro-endoscopic removal ²²⁾.

Trentadue et al. report a case in which the finding was incidentally detected in a 49-year-old man. They describe the imaging characteristics of the lesion in such a rare location, highlighting the role of magnetic resonance imaging (MRI) (specifically 3 Tesla) in the management of asymptomatic patients ²³⁾.

2015

A 48-year-old female presented with an insidious history of progressive visual loss. Magnetic resonance imaging (MRI) showed a CM in the suprasellar region. The patient was operated via a right pterional approach with a complete lesion removal. The postoperative course was uneventful. Early postoperative ophthalmological examination revealed minimal improvement of the vision in the left eye ²⁴⁾.

2014

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 $34.8 \pm$ 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 ²⁵.

2012

Ning et al. report a 28-year-old male presenting with left homonymous hemianopsia. Magnetic resonance imaging (MRI) revealed an occupied lesion located in the right side of the optic chiasm, and a clinical diagnosis of chiasmal CM was made. Microsurgical excision was performed via anterolateral pterional craniotomy. The patient showed good recovery with slight improvement of the visual field deficits after the operation. No CM recurrence was discovered during the follow-up MRI scans ²⁶.

2011

Rheinboldt and Blase report the case of a 31-year-old male who presented to the ER with a 1-week history of progressively worsening, throbbing, left retro-orbital headache, ptosis, and subjective worsening of short-term memory function. Initial review of systems and laboratory data were noncontributory. Non-contrasted CT demonstrated a large hyperdense mass centered in the suprasellar cistern without evidence of dissecting extra-axial hemorrhage. Though the initial appearance mimicked a basilar tip aneurysm or another primary extra-axial suprasellar pathology such as a hemorrhagic or proteinaceous craniopharyngioma, germinoma, or optic glioma, a second smaller, clearly intra-axial, hyperdense lesion was observed in the left periventricular forceps major white matter. Consideration for multiple cavernomas versus hypervascular metastatic disease such as renal malignancy, thyroid malignancy, or melanoma was raised. CTA confirmed normal intracranial vasculature. Subsequent MRI images showed an acutely hemorrhagic mass centered at the left paramedian hypothalamus and tuber cinereum with numerous secondary foci, demonstrating mature hemorrhagic elements and confirming the diagnosis of multiple cavernomas ²⁷.

2008

A 33-year-old female who suffered from a recurrence of an intrachiasmatic cavernous malformation is presented. She had already undergone surgery in 1991 and 2001 and was admitted to our hospital

with reduced vision in the right eye. After MRI, and diagnosis of recurrence of the cavernoma, a neurosurgical operation was performed using the pterional approach. The intraoperative situation was documented with micro photographs. The postoperative course was uneventful. The female described a minimal improvement of her vision. No postoperative complications were observed. To our knowledge, microsurgically complete extirpation of a recurrence of an intrachiasmatic cavernoma has not yet been reported in the literature ²⁸⁾.

2007

Santos-Ditto et al. present the case of a female patient who developed chiasmatic apoplexy and menstrual alterations. CT scanning showed a suprasellar hemorrhage. She underwent surgery with the presumptive diagnosis of pituitary tumor. At surgery, we find a brown-grayish lesion involving left optic nerve and chiasm. Cavernous angioma was diagnosed by histopathology. Cavernous angiomas constitute nearly 15% of all central nervous system vascular malformations. Location at the optic pathway is very rare, but must to be ruled out in the diagnosis of a patient with chiasmatic and/or optic apoplexy. Surgery is useful in preventing worsening of the previous deficit or a new visual defect ²⁹⁾.

A 15-year-old boy presented with an extremely rare optochiasmatic cavernous angioma. He was admitted to a special hospital with the complaint of blurred vision persisting for 1 month. Magnetic resonance imaging and biopsy of the lesion were inconclusive. He was admitted to the neurosurgical clinic after worsening of the visual symptoms 9 months later. Repeat magnetic resonance imaging showed optochiasmatic cavernous angioma which had doubled in size. The lesion was removed completely without any problem. Postoperatively his visual complaints remained stable, but had improved after 1 year. Optochiasmatic cavernous malformation should be treated by surgical excision, whereas biopsy is useless and may result in enlargement ³⁰⁾.

A 38-year-old male patient who suffered from acute onset of severe headache and progressive loss of vision. The vascular malformation of the optic pathways was completely removed via a pterional approach. This is the first reported instance of complete resection of a cavernoma involving the optic nerve, the chiasm, and the optic tract ³¹⁾.

2006

Muta et al. report a 14-year-old boy with cavernous malformation of the optic chiasm. He had a 2-year history of gradually worsening visual disturbance. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a suprasellar mass, findings compatible with craniopharyngioma. The mass was biopsied and histological examination confirmed cavernous malformation. On the second day after the biopsy, he suffered chiasmal apoplexy due to intratumoural haemorrhage, lost visual acuity and developed a field cut. Cavernous malformations arising from the optic nerve and chiasm are extremely rare; only 29 cases have been reported to date. Most patients manifested acute visual acuity and visual field disturbances. Although MRI findings of cavernous malformations in the brain parenchyma have been reported, MRI findings on the optic nerve and chiasm may not be completely diagnostic. Of the 29 documented patients, 16 underwent total resection of the lesion

without exacerbation of their preoperative symptoms; in some cases, resection was complicated by risk of damage to the surrounding neural tissue. As patients may suffer intratumoural haemorrhage after biopsy or partial removal of the lesion, the advisability of surgical treatment of cavernous malformations of the optic nerve and chiasm must be considered carefully ³²⁾.

In their meticulous review of the literature, Lehner et al. found 42 previously reported patients with vascular malformations within optic nerves, chiasm, or optic tracts, 30 of them being cavernous hemangiomas. The optic chiasma was involved in 38 patients (90.5%) and a total excision of the tumor was performed in 21 cases. Lehner et al. published a patient with a cavernous haemangioma of the optic chiasma and left optic tract who presented with an acute defect of the right visual field and severe retro-orbital pain. They succeeded in total excision of the malformation via a neuronavigationally guided approach. In the postoperative course, vision of our patient improved immediately and was found to be completely normal three months after the surgical intervention. Considering this patient and the published cases in the literature, they are of the opinion that microsurgical excision is a safe and efficient treatment for these rare pathologies ³³⁾.

2005

Shkarubo et al. describe a rare case of chiasmatic apoplexy whose cause was chiasmatic cavernoma. In addition to acute visual disorders suggesting the involvement of the left optic nerve, chiasma, and left visual pathway, 23-year-old patient had endocrine disorders as polyuria, polydipsia, which first suggests craniopharyngioma and glioma of the chiasma. A capsule and hematoma clots were removed from the thickened left optic nerve and left chiasmatic half during surgery. Only did a morphological study involving immunohistochemical analysis permit identification of the process as hemorrhage from cavernous micromalformation with the formation of hematoma ³⁴⁾.

1989

Three patients with cavernomas of the optic nerve, chiasm, or optic tract are presented. All suffered progressive visual loss due to local hemorrhage and the space-occupying effects of the vascular malformation. Computed tomography scans revealed small lesions with mild contrast enhancement in the suprasellar and parasellar cisterns, whereas angiography was unremarkable. Magnetic resonance imaging was helpful in our cases both for diagnosis and for planning surgical approach, showing typical signs of cavernomas as confirmed by subsequent surgery and histological examination. The clinical and intraoperative findings are presented ³⁵⁾.

1984

Buonaguidi et al. report a very rare case of an intrasellar cavernous hemangioma mimicking, clinically and neuroradiologically, the presence of a nonfunctioning pituitary neuroendocrine tumor. It was possible to diagnose this benign, congenital vascular malformation only through a histological examination ³⁶⁾.

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