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SURGICAL NEUROLOGY INTERNATIONAL
SNI: Neurovascular, a supplement to Surgical Neurology International
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Editor: James I. Ausman, MD, PhD University of California, Los Angeles, CA, USA

Optic chiasmal cavernous angioma: A rare suprasellar vascular malformation

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Received: 11 March 16 Accepted: 27 May 16 Published: 01 August 16

Abstract

Background: Suprasellar cavernous malformation in the optic pathway is not commonly encountered. To date, there are only few reports present in the literature.

Case Description: The authors report a rare case of suprasellar optic pathway cavernous malformation in a 33-year-old female who presented with progressive visual loss. Her imaging revealed a large heterogeneous, hyperintense, hemorrhagic right suprasellar extra-axial complex cystic structure, causing mass effect on the adjacent hypothalamus and third ventricle displacing these structures. Gross total resection of the lesion was achieved utilizing a right frontal craniotomy approach. Histopathological examination confirmed the diagnosis of suprasellar chiasmal cavernous malformation.

Conclusion: Although visual pathway cavernous malformation is a rare event, it should be included in the differential diagnosis of lesions occurring suprasellarly in the visual pathway and hypothalamus.

Key Words: Cavernoma, cavernous angioma, hypothalamus, optic pathway, suprasellar region

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/2152-7806.187495
Quick Response Code:

INTRODUCTION

Cavernous malformations (CMs) are common low-flow lesions of the central nervous system (CNS), accounting for 10–20% of all vascular malformations with an incidence of 0.3–0.7% in the general population.[16,21] They are distinct, well-circumscribed lesions composed of a single endothelial cell layer with sinusoidal spaces and no muscular layer.[12] They are separated by a collagenous stroma without intervening brain parenchyma. They are most commonly encountered in the supratentorial region (80%), followed by the infratentorial region (15%), and spinal cord (5%).[25] Suprasellar occurrences of CM in the optic pathway are extremely uncommon. To the best of our knowledge, less than 80 cases have been reported in the literature.[13,29] Herein, we report a rare case of suprasellar optic pathway CM in a 33-year-old female. In addition, a literature review on suprasellar optic pathway CMs is presented.

CASE REPORT

History and examination

A 33-year-old female presented 3 months postpartum with a headache of moderate severity and progressive visual

How to cite this article: Abou-Al-Shaar H, Bahatheq A, Takroni R, Al-Thubaiti I. Optic chiasmal cavernous angioma: A rare suprasellar vascular malformation. Surg Neurol Int 2016;7:S523-6.
http://surgicalneurologyint.com/Optic-chiasmal-cavernous-angioma:-A-rare-suprasellar-vascular-malformation/

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.

Imaging

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 [Figure 1]. Computed tomography (CT) arteriography demonstrated a completely thrombosed center. The imaging findings were compatible with suprasellar CM.

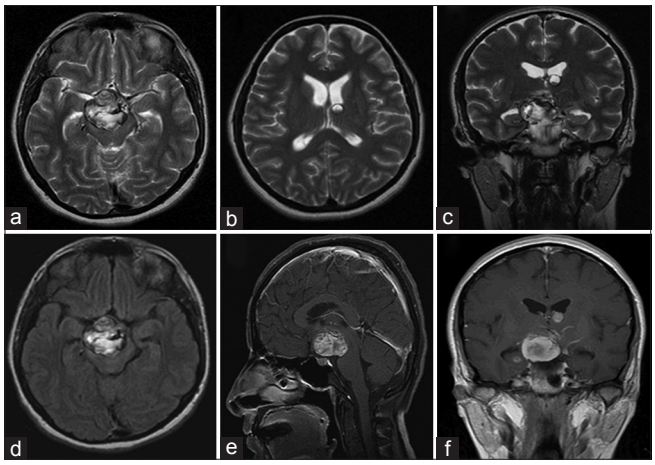

Figure 1: Preoperative magnetic resonance imaging of the brain demonstrating a heterogeneous hyperintense T1 and T2 suprasellar mass compatible with acute/subacute hemorrhagic lesion measuring 3.1 × 2.9 × 2.1 cm with no clear evidence of enhancement allowing for intrinsic T1 hyperintensity (a, c, d). There is some adjacent extension of blood products into the right hippocampal sulcus (c). There is also a left-sided moderately-sized venous angioma in the basal ganglia (c, f). This represents a large hypothalamic region/subependymal cavernous angioma with recent hemorrhage and associated surrounding mild edema mainly in the right basal ganglia and thalamus. Moreover, displacement and mass effect on the surrounding structures including basal ganglia, subcapsular brain parenchyma, as well as hypothalamus and displacement of the cerebral peduncle with a mass effect on the midbrain is noted (a, c, d-f). An additional hemorrhagic focus in the left frontal horn of the lateral ventricle close to the foramen of Monro region is also seen (b, c). Specifically, there is a mass effect and anterior displacement of the partially visualized pituitary infundibulum as well as a mild mass effect on the optic chiasm particularly on the right side compressing and displacing it (c, e, f). No additional susceptibility foci are noted

Operation and histopathological findings

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.

Postoperative course

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 [Figure 2].

DISCUSSION

Optic pathway CMs are rare benign vascular lesions, representing less than 1% of all CNS CMs.[13,29] They are commonly discovered between the 2nd and 4th decades of life. Optic pathway CMs affect both genders, with a slight female predominance. Patients usually present due to the development of visual deficits. Acute visual disturbance, headache, nausea, and retroorbital pain, collectively known as "chiasmal apoplexy," represent

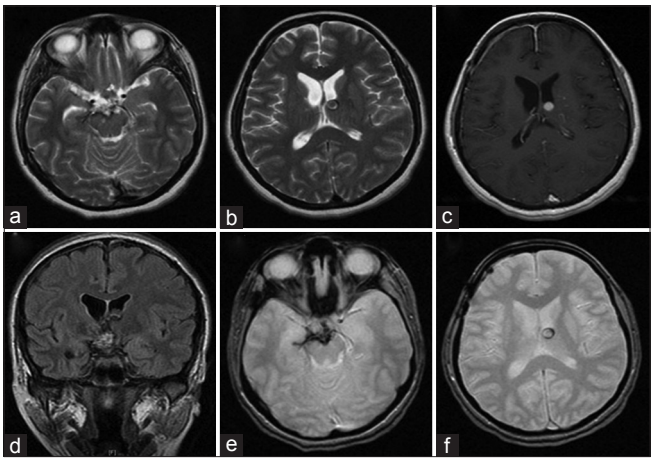

Figure 2: Postoperative magnetic resonance imaging of the brain depicting complete removal of the suprasellar hemorrhagic cavernous malformation with no evidence of a residual lesion or recurrence with some adjacent extension of blood products into right hippocampal sulcus (a-d) and no hemosiderin staining of the ventricular surface on the gradient-echo imaging (e, f)

the most commonly encountered symptoms among optic pathway CM patients.[13,14,29] These symptoms typically occur after a period of transient blurry vision and headaches. In addition, hypopituitarism from direct compression of the infundibulum has been reported in the literature.[2,22,27] Our patient presented with headache and left homonymous incomplete hemianopia.

The pathogenesis of CMs remains elusive. They are thought to arise during the early periods of embryogenesis and grow according to blood changes and malformative mechanisms.[19,24,28] However, not all CMs arise embryologically, as some de novo cases occurring after radiation therapy have been reported in the literature.[3,15,18]

The natural history of optic pathway CMs is also not clear. It is thought that optic pathway CMs have a higher tendency to bleed than cerebral ones because of the eloquence of the optic pathway region.[11,13,17] Interestingly, the rate of optic pathway CM hemorrhage is higher in females.[1,13]

It is not easy to diagnose CMs of the optic pathway preoperatively. It is not uncommon for them to be misdiagnosed as optic neuritis, and a great number of these patients receive corticosteroid therapy.[5,29] Interestingly, some of those patients will spontaneously recover from their symptoms, misattributing it to the effects of the corticosteroid therapy. However, to date, there is no evidence supporting corticosteroid therapy in CM.

MRI is considered the most sensitive and specific imaging modality for the diagnosis of CM.[23] 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, as seen in our patient. 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.[4,6] 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.[13] On CT scan, optic pathway CMs appear as well-demarcated hyperdense lesions with or without calcifications.[26] 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, as was demonstrated in our patient.[20]

It is important to note that none of these imaging features is pathognomonic as they can be encountered in other conditions. Thus, preoperative diagnosis of optic pathway CMs is extremely difficult and challenging. CMs of the optic pathway are commonly misdiagnosed as optic neuritis, optic glioma, meningioma, craniopharyngioma, venous angioma, arteriovenous malformation, thrombosed aneurysm, and pituitary apoplexy.[13,29] In our patient, we initially had a differential diagnosis of craniopharyngioma, glioma, or germ cell tumor. Therefore, it is important to include CM in the differential diagnosis of suprasellar optic pathway lesions.

The treatment of choice for optic pathway CM is complete surgical resection of the lesion. Surgical resection of these lesions is considered a challenge because of their deep location and eloquence. Complete surgical resection of the CM is essential in order to prevent regrowth and bleeding.[7,9,11,17,20] It results in resolution of the symptoms in the majority of patients.[13,29] Decompression and subtotal resection of the lesion may improve the symptoms; however, they carry the risk of rebleeding into residual CM.[13] Biopsy is contraindicated for these lesions due to the high risk of bleeding and symptomatic worsening.[7,20]

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.[13] In our patient, we utilized the frontal craniotomy to optimally expose the lesion, and we achieved gross total removal of the CM with this approach.

Spontaneous recovery has been reported in the literature, especially in children.[8,9] Radiation therapy has been utilized in a few patients with optic pathway CM.[10,30] However, its role in CMs of the optic pathway is still controversial. Due to the eloquence of this region, radiation therapy can have major and devastating complications; thus, its utility in such lesions is limited. Therefore, gross total resection of optic pathway CMs remains the standard care of therapy.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors declare no conflicts of interest regarding the production of this article. The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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