

Surgical management of cavernous malformation of the optic nerve with canalicular extension

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Abstract

Background: Cavernous malformations arising in a single optic nerves sparing the chiasm (intracranial prechiasmatic optic nerve) and expanding into the optic canal are extremely rare lesions. Published series or case reports regarding the surgical removal of these vascular malformations within this specific location are scarce.

Case Description: We present the first case to be published, of an intracranial optic nerve cavernous malformation with a contiguous canalicular component that was totally and successfully removed through a microsurgical pterional approach with excellent clinical outcome.

Conclusion: This pathology should always be considered in the differential diagnosis of optic neuropathy and visual loss. Early detection and surgical proposal are mandatory, warranting the prevention of permanent damage to visual pathways. Radical resection is challenging, but usually curative and associated with favorable visual outcomes.

Key Words: Cavernous malformation, cranial nerve, operative technique, optic canal unroofing, optic nerve, optic pathway

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INTRODUCTION

Cavernous malformations (CMs) are vascular malformations that can be found throughout the central nervous system.^[6] They rarely affect the cranial nerves.^[4,8,20] When arising in a single optic nerve (ON) sparing the chiasm (intracranial prechiasmatic ON) and expanding into the optic canal, they are even rarer and require specific surgical considerations.^[5,13,24] We present the first case in this topography to be reported in the literature.

CASE REPORT

A 40-year-old female with past medical history of migraine and no family history of CM, presented with

unilateral scotomas with a gradual onset over 7 months and progressive visual deterioration, referred as blurred vision of the right eye (RE), with a significant worsening few days before seeking medical care.

She was initially admitted at Ophthalmology outpatient clinic for observation. The ophthalmologic evaluation revealed a decreased visual acuity (7/10) and a visual field defect on the RE. Her left eye (LE) was completely normal (10/10, without field defect). The computerized campimetry (CC) disclosed a supero-inferior arciform defect in the RE. LE CC was normal [Figure 1].

Visual evoked potential responses showed results compatible with a right second cranial nerve dysfunction due to compressive optic neuropathy on the RE.

A noncontrast enhanced computed tomography (CT) scan was performed and revealed a spontaneously hyperdense focal nodular lesion, heterogeneous, with irregular contours and a punctate area of increased attenuation coefficient related to calcification, on the right optic-carotid cistern, adjacent to the ipsilateral ON. No surrounding edema or mass effect was documented. No evidence of subarachnoid or intraventricular hemorrhage [Figure 2].

Based on the CT scan result, the patient was referred to the outpatient clinic of the Neurosurgery Department. She completed the diagnostic neuro-imaging evaluation with a CT angiography (CTA) [Figure 3], a magnetic resonance imaging (MRI), and a MR angiography (MRA) of the brain.

The MRI showed a focal solitary well-defined rounded lesion involving the right ON in its intracranial prechiasmatic segment, sparing the chiasm. The T1-weighted noncontrasted sequence showed a spontaneously hyperintense lesion [Figures 4a-c]. In the T2-weighted MRI, the lesion exhibited a heterogeneous signal intensity and a peripheral hypointense rim [Figure 4d]. No large vessels adjacent to the lesion were identified.

The MRA demonstrated a hyperintense mass due to hemosiderin. No enlarged vessels were identified [Figure 5].

These features were considered to be consistent with an occult vascular malformation, presenting an inner hematoma (blood and blood by-products in various states of evolution) surrounded by a peripheral hemosiderin rim, suggesting the diagnosis of a CM of the intracranial segment of the right ON.

The patient complaints remained unchanged for one week. After this period of time, she was electively operated on, through a right pterional craniotomy. An intradural optic canal unroofing, through a drilling technique, was performed to completely expose and microsurgically remove the CM [Figure 6]. The CM was found over the superior surface of the ON at the entrance of the optic canal with an anterior canalicular extension.

RESULTS

Total removal of the right ON CM was performed with preservation of the ON. CM localization on the right intracranial prechiasmatic ON (sparing the

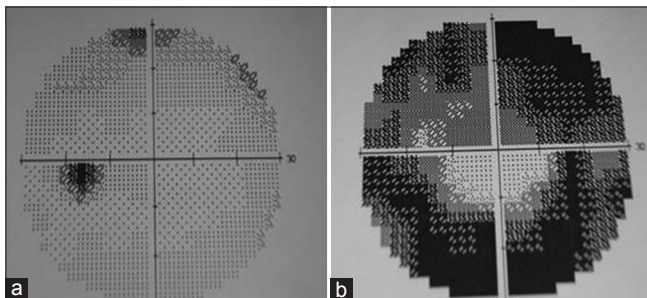


Figure 1: Preoperative CC. (a) LE CC: Normal; (b) Supero-inferior arciform defect in the RE



Figure 3: CTA demonstrating normal vessels. No lesions were documented

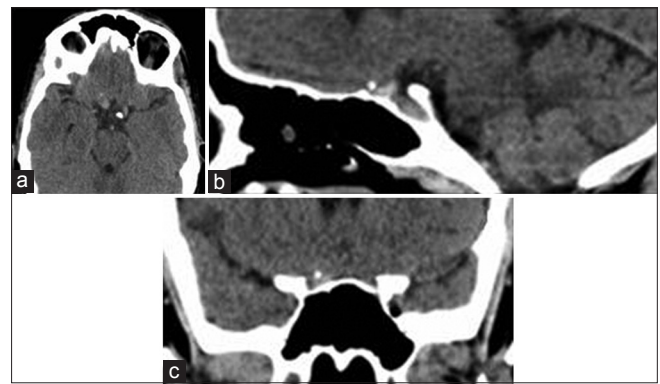


Figure 2: Noncontrast CT scan reveals a spontaneously hyperdense lesion with a punctate calcification. (a) Axial; (b) Sagittal; (c) Coronal views

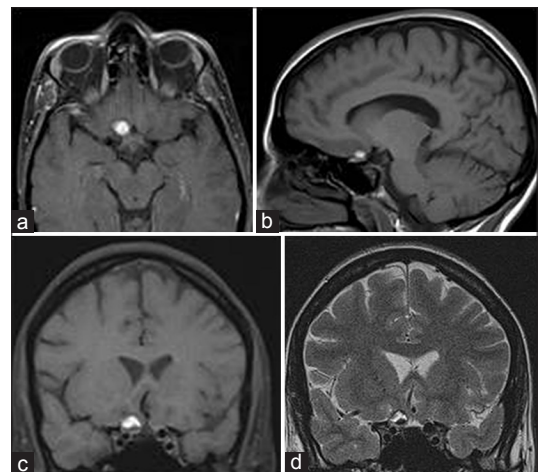


Figure 4: Preoperative MRI presenting a lesion affecting the right intracranial prechiasmatic optic nerve (sparing the chiasm). (a) Axial T1W; (b) Sagittal T1W; (c) Coronal T1W; (d) Coronal T2W

chiasm) with a canalicular component, as seen in the preoperative diagnostic images, was corroborated by the intraoperative findings. There were no procedure-related complications. The postoperative CT scan [Figure 7] and MRI [Figure 8] revealed complete resection of the optic CM and excluded any surgery-related complications.

The histopathological analysis of the resected lesion confirmed the preoperative diagnosis of CM [Figure 9].

The postoperative ophthalmologic evaluation revealed a significant improvement of the previous visual dysfunction with total recovery of the RE visual acuity (from 7/10 to 10/10) and visual field defect [Figure 10].

Eighteen months after surgery, the patient totally recovered; the visual deficits and the migraine symptoms also ceased, allowing the suspension of the prescribed medical therapy.

DISCUSSION

CMs constitute the most common type of angiographically occult vascular malformation.^[9,16,17,25] They represent 10-15% of all vascular malformations.^[1,15] Histologically they are composed of endothelial lined dilated vessels (caverns) packed together without intervening neural parenchyma.^[27] Their prevalence in the general population has been estimated to be roughly 0.1-0.5%.^[19,26] Sporadic and familial forms exist. The familial form affects 30-50%

of patients harboring a CM, and they are associated with a greater incidence of multiple lesions.^[23]

They may occur anywhere in the central nervous system, most frequently in the supratentorial compartment (70-80%), followed by the infratentorial (15%), spinal cord (5%), and, rarely, affecting the cranial nerves.^[6] CMs affecting the ON, oculomotor, trochlear, trigeminal, facial, vestibulocochlear, glossopharyngeal and hypoglossal nerves have been described.^[4,8,14,20] They exhibit unique clinical particularities, which should suggest a high index of suspicion and instigate the possibility of early surgical treatment.

Neuroimaging characteristics are suggestive but usually nondiagnostic. The high sensitivity of MRI makes it the neuroimaging modality of choice in the evaluation of patients with suspected CMs, allowing an early detection of these lesions. The MRI

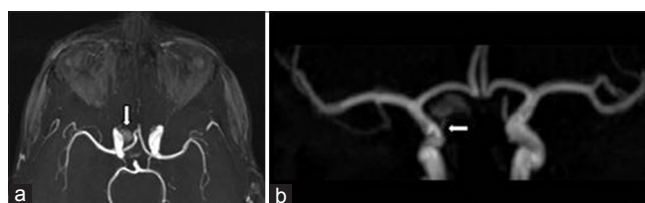


Figure 5: Preoperative MRA: (a) The lesion appears as an hyperintense mass (arrow) due to hemosiderin; (b) 3D reconstruction demonstrating an intimate relation of the right ophthalmic artery (arrow) with the ipsilateral optic mass

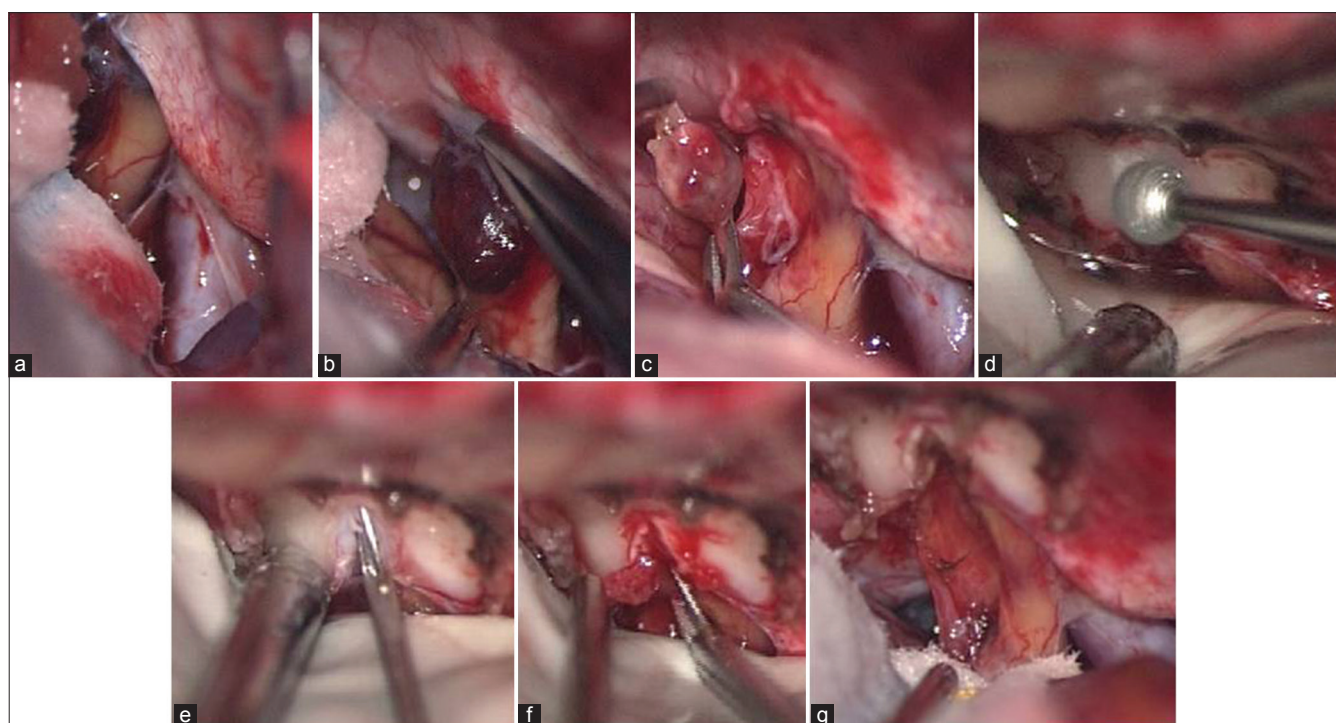


Figure 6 : Intraoperative microsurgical images: (a) Right optocarotid cistern exposition; (b, c) Microsurgical resection of the CM arising from the intracranial segment of the right optic nerve at the entrance of the optic canal ; (d) Unroofing of the optic canal through a drilling technique using a 3 mm diamond drill; (e) Opening of the dura of optic canal; (f) Resection of the canalicular component of the CM; (g) Optic nerve and the emergence of the ophthalmic artery after complete removal of the CM

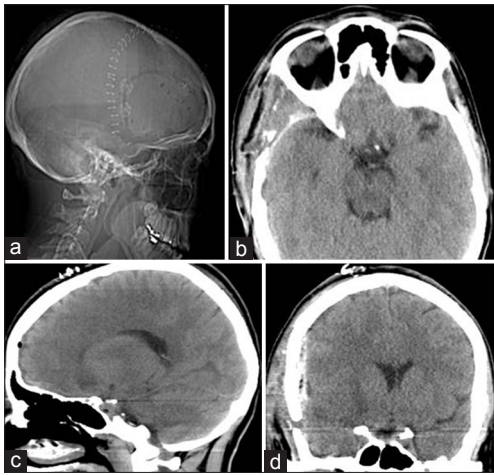


Figure 7: Postoperative CT scan confirming total removal of the CM and excluding surgery-related complications. (a) Lateral scout view exhibiting the surgical approach (right pterional craniotomy); (b) Axial; (c) Sagittal; (d) Coronal views

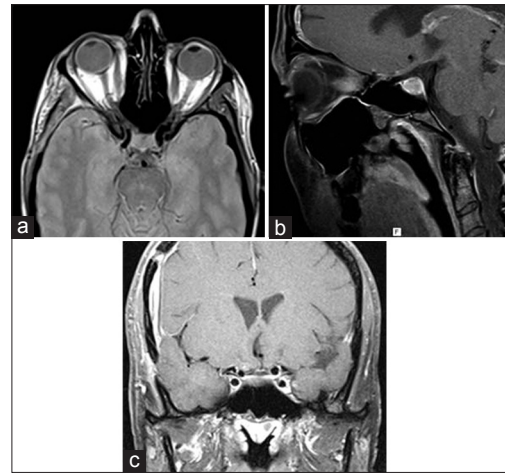


Figure 8: Postoperative T1-weighted MRI confirming total removal of the CM. (a) Axial; (b) Sagittal; (c) Coronal views

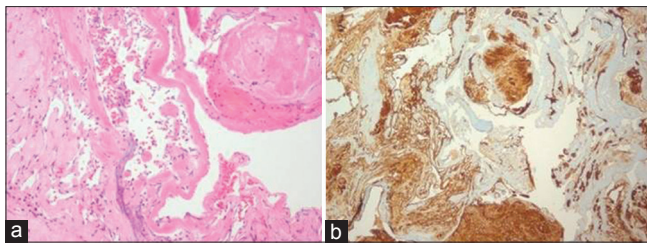


Figure 9: (a, b) Thin and thick walled vascular spaces lacking elastin and smooth muscle fibers. Focally thickened and hyalinized walls of vascular channels with focus of calcification. Peripheral margin of gliotic tissue containing hemosiderin-laden macrophages

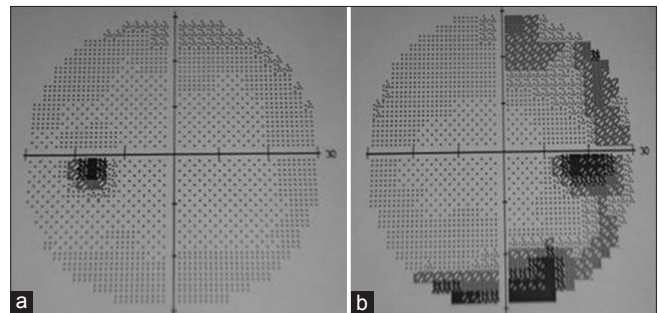


Figure 10: Postoperative CC documented a significant recovery of the previous visual field defect: (a) left eye; (b) right eye

appearance of CMs is mostly related to hemorrhage in evolution and is highly suggestive of the diagnosis. In some patients, the diagnosis may not be evident until surgery and differentiation from neoplasm may be possible because most metastases greater than 5 mm can have vasogenic edema, while CMs have little or no edema.^[21]

From a clinical standpoint, these lesions show a great variability both morphologically and in terms of their behavior. Natural history studies suggest that most lesions have a relatively benign course. This is particularly true when the lesions are located superficially in the supratentorial compartment. Reported rates of hemorrhage range between 0.25% and 4.2% per person/year.^[7,11,22,26]

Clinical presentation is diverse, including seizures (23-50%), headaches (6-52%), and focal deficits (20-45%) arising from mass effect due lesion growth or bleeding, or caused by perilesional hemosiderin deposition. A wide variety of focal neurological deficits have been reported with CMs, being the signs and symptoms directly related to the location of the CM. Up to 40% of patients may be asymptomatic.^[2,3,7,22,26,29] Lesions are increasingly being

detected as incidental MRI findings.^[15] Asymptomatic cranial nerve CMs should be managed conservatively and followed-up annually with MRI.^[3]

Because of the eloquence of the affected tissue of origin, cranial nerve CMs are more prone to become symptomatic and, unlike CMs in other locations, their clinical behavior is aggressive, seriously threatening cranial nerve function. Even a small bleed can cause clinically significant neurological deficits.^[12] Symptomatic patients with CMs of the cranial nerves experience inexorable progression to cranial nerve dysfunction.^[8] When arising within the optic pathways, usually they result in visual acuity loss and field defect that can, potentially, be correctable with prompt surgery.^[10,18]

A retrospective review of the current literature on optic pathway CMs show higher rates of vision preservation in patients who underwent complete lesion removal.^[5,12,13,24] For these reasons, the authors recommend that all symptomatic patients must be treated, being surgical resection the gold standard of care. Surgery is indicated to prevent (re) hemorrhage, progression of the cranial nerve dysfunction, with concomitant vision improvement or simply its preservation. Gross total resection (GTR) is the mainstay of treatment, since residual lesions have a

high risk of recurrence and lead to progressive symptoms, both requiring a more definitive, difficult, and dangerous surgery.

Treatment strategies for deep-seated CMs or those located in eloquent areas include observation, radiosurgery, and microsurgery. CMs of the ON constitute a special subgroup. Because of their highly eloquent location, severe neurological symptoms are more likely to occur than in other locations. Symptomatic patients should be surgically treated, being GTR, through microsurgical techniques, the state-of-the-art treatment option and the only efficient modality of care.

In the present case, the CM was found over the superior surface of the ON at the entrance of the optic canal with an anterior canalicular extension.

Some considerations were taken into account to safely remove this vascular malformation: (i) Before surgery, it was important to correctly predict the preoperative diagnosis; otherwise, difficulties at this stage could lead to problems in selecting the most appropriate strategy for treatment. CMs can be diagnosed in the vast majority of cases with high accuracy on MRI thanks to their specific appearance. (ii) The possibility of presence of multiple lesions should be ruled out before surgery, whenever this is pertinent. In this patient, an isolated solitary supratentorial CM was diagnosed on cranial MRI. Having in mind the lack of family history of CMs, the absence of spinal symptoms and the rarity of spinal CMs in patients harboring a solitary intracranial CM, a spinal MRI study was not performed. (iii) Choosing the optimal approach was critical due to the eloquent location of the CM. The best approach was a straight-line corridor to the lesion, with minimal manipulation of the surrounding neural structures, allowing a safe and cautious lesion dissection, in order to perform a GTR. (iv) An intradural approach was executed. Opening of the sylvian fissure and basal cisterns was performed sharply with microsurgical dissection. (v) Unlike in other vascular malformations, it was not possible, in this case, to completely expose the whole lesion and circumscribe it before starting its resection, due to its particular topography and lobulated morphology. The bulky component of the CM was overlying and seated on the prechiasmatic component of the ON, at the entrance of the posterior aspect of the optic canal. This portion of the CM, due to its dimensions, completely blocked the optic canal posteriorly, covering its superior bony border, making it impossible to proceed with its safe unroofing without removing the extra canalicular component of the CM first. (vi) Identifying and establishing a clear dissection plane between the CM and ON, at the very beginning of dissection, was an important step. Hematomas, when present, should be evacuated. (vii) As the ON is vascularized from its sheath in a centripetal way it was not be mobilized. (viii)

After removal of the intracranial part of the CM and decompressing the ON, we removed the canalicular component of the lesion. Dura of the orbital roof was, at this stage, completely exposed. It was incised creating a small window on the superior surface of the optic canal. Optic canal unroofing was then carefully performed through a drilling technique using a diamond ball under generous irrigation to avoid thermal injury to the ON. Structures surrounding the drilling area were covered with a wet piece of operative rubber glove to prevent deposition of bone dust in the subarachnoid space. Cottonoids were not used because they could get entangled to the rotating drill causing severe damage. (ix) Opening of optic canal dura was then performed, and the canalicular segment of the CM was subsequently safely removed. Portions of the lesion that had already been separated from the nerve were sharply excised. Care was taken at this stage, not to harm the ophthalmic artery. (x) A blunt separation of the lesion away from the intracanalicular ON was performed. (xi) Careful microsurgical dissection proceeded until a GTR was achieved. The hemosiderin-stained tissue should was not resected once it was not part of the CM and its resection could damage the ON or ophthalmic artery. (xii) Some surgeons prefer an extradural approach to open the optic canal with a subsequent translation into an intradural approach. Once the major component of the CM was lying on the superior surface of the ON at the entrance of the posterior aspect of the optic canal, we adopted an intradural approach. We considered safer to indentify the ON and the supralying CM in an early stage, decompress it gaining some room for microsurgical dissection with almost none ON manipulation. (xiii) Due to its specific appearance on MRI, the lesion was preoperatively diagnosed as a CM. Being considered a slow blood flow vascular malformation, and due to the above-mentioned reasons, we first removed the extra canalicular component of the CM and subsequently the canalicular one. No additional hemorrhage risk was added by assuming a careful piecemeal resection. Actually, many CMs in other eloquent locations, like in the brainstem for example, are also conveniently managed through a gradual cautious piecemeal resection.^[8,23,28] (xiv) Meticulous hemostasis with bipolar coagulation at a low current intensity and abundant irrigation with saline solution, must avoid contact with the ON and its vascularization.

CONCLUSION

CMs affecting the ON with extension into the optic canal are extremely rare lesions. We present the first case in this topography to be reported in the literature. This pathology should always be considered in the differential diagnosis of optic neuropathy and visual loss. In the context of neurological deficit, early detection and surgical proposal is mandatory, warranting the prevention of permanent

damage to visual pathways. MRI is the neuroimaging modality of choice in the diagnosis of these lesions.

The mainstay of treatment is radical resection, which is challenging, but usually curative and associated with favorable visual outcomes. Symptomatic patients or enlarging (progressive) CMs on serial MRI, should undergo resection. Surgeon's experience, anatomical knowledge, and microsurgical skills are important factors that influence the final outcome.

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