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Review

Cavernous malformations isolated from cranial nerves: Unexpected diagnosis?

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ABSTRACT

Objectives: Cranial nerves (CN) cavernous malformations (CMs) are lesions that are isolated from the CNs. The authors present three cases of CN CMs, for which MR was demonstrated to be critical for management, and surgical resection produced good outcomes for the patients. Surgical removal is the recommended course of action to restore or preserve neurological function and to eliminate the risk of future haemorrhage. However, the anatomical location and the complexity of nearby neural structures can make these lesions difficult to access and remove. In this study, the authors review the literature of reported cases of CN CMs to analyse the clinical and radiographic presentations, surgical approaches and neurological outcomes.

Patients and methods: A MEDLINE/Pub Med search was performed and revealed 86 cases of CN CMs. The authors report three additional cases in this study for a total of 89 cases. CMs affecting the optic nerve (CN II), oculomotor nerve (CN III), facial/vestibule-cochlear nerves (CN VII, CN VIII) have been described. The records of three patients were reviewed with respect to the lesion locations, symptoms, surgical approaches and therapeutic considerations. Clinical and radiological follow-up results are reported. Three patients (2 females, 1 male; age range 21–37 year) presented with three CN lesions. One lesion involved CN III, one lesion involved CN VII–CN VIII, and one involved CN II. The patient with the CN III lesion had a one-month history of mild right ptosis and diplopia. The patient with the CN VII–CN VIII lesion exhibited acute hearing loss and on the left and left facial paresis. The patient with the opticchiasmatic lesion presented with acute visual deterioration on the right and a left temporal field deficit in the left eye. Pterional and orbitozygomatic craniotomies were performed for the CN III lesion and the CN II lesion, and retrosigmoid craniotomy was performed for the cerebello-pontine angle lesion.

Results: All patients experienced symptom improvement after surgery. On MR follow-up, recurrence was excluded in all patients.

Conclusions: CN CMs present with specific symptoms and require complex surgical techniques for resection. These lesions are frequently symptomatic, because of the complexity of the origin tissue. Symptomatic CN CMs should be resected microsurgically and completely when possible to prevent further losses of nerve function, improve function, avoid recurrence, and to eliminate the risk of future haemorrhages. The authors discuss the therapeutic options and the radiological features of these infrequent localisation of CMs. Specifically, the authors focus on the role of magnetic resonance imaging in the identification of these rare lesions.

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Cavernous malformations (CMs) are angiographically occult vascular malformations that are pathologically defined as thin-walled capillary spaces without muscular tissue or intervening neural tissue. Because of their vessel architecture, CMs tend to cause repeated haemorrhages. The annual bleeding rate of patient has been reported to be 0.25%–20% [1,2], regardless of the location in the brain.

Most lesions occur in the subcortical brain parenchyma and typically cause headaches, seizures, or neurological deficits due to haemorrhage. Haemorrhaging is typically extra-lesional, but can occur internally, with repetitive bleeding that causes growth and ultimately mass effects on the surrounding structures [2–4].

CMs rarely arise from cranial nerves (CN), but CN CMs primarily affect the optic nerve/chiasm, and sporadically affect the oculomotor, facial, vestibulocochlear, and hypoglossal nerves. CMs typically cause dysfunction of the affected cranial nerve, either insidiously via mass effects or acutely due to haemorrhaging.

MR imaging is currently the best imaging method for the evaluation of these malformations [5–8], although MR imaging might not be completely and consistently diagnostic.

The authors present three cases of CN CMs, in which MR was critical for management, and surgical resection led to acceptable outcomes for the patients. We review the pertinent literature, both to outline a suitable diagnostic approach, and to clarify the indications for treatment and the surgical techniques that are applicable to the management of these complex lesions with the aim of identifying the best strategy.

1. Case presentation

Case 1. A 34-year-old woman presented with a 1-month history of mild right ptosis and diplopia. Myasthenia syndrome was supposed and subsequently excluded. On admission a neurological examination detected a mild third cranial nerve palsy on the right. Brain MR revealed a small mass located in the interpeduncular cistern at the level of the right oculomotor nerve. The lesion exhibited mixed signal intensities on both T1 and T2 sequences, and only very subtle enhancement following the administration of gadolinium (Fig. 1a).

The patient underwent a right pterional craniotomy. As the medial portion of the sylvian fissure was opened and the basal arachnoid cisterns were dissected, a red, multiloculated lesion became visible (Fig. 1b). The lesion was a typical mulberry-like vascular malformation that was adherent but did not encircling the third cranial nerve. A microsurgical dissection was performed around the lesion. En bloc removal was not possible due to the thin lesion walls. Thus a meticulous piecemeal total excision was performed to preserve the anatomical integrity. Histopathological analysis revealed the lesion to be a cavernous haemangioma.

The post-operative course was uneventful. One year after surgery, the patient presented with only a negligible right ptosis, and a normal MR appearance (Fig. 1c).

Case 2. For two months, a 37-year-old male patient experienced vertigo, progressive sensorineural hearing loss on the left and the onset of left facial paresis (House-Brackmann Grade II–III). On admission, a neurological examination revealed no further abnormalities. Brain MR detected a mass in the left cerebello-pontine angle (CPA), which extended towards the internal auditory canal (IAC).

The lesion was heterogeneously hyperintense on T1-weighted (Fig. 2a) images and exhibited a hypointense ring on T2-weighted images. No enhancement due to the administration of a contrast agent was observed.

The patient underwent a left retrosigmoid craniotomy. After opening the CPA cistern, a lesion associated with CN VII and CN VIII was identified based on its typical reddish-blue colour and raspberry-like appearance that is typical of a cavernous hemangiomas. Neither the facial nerve nor the cochlear nerve was in contact with the lesion. After high-speed drilling of the IAC, the mass was excised en bloc, and the hemosiderin deposition, present in the vestibular nerve around the angioma was left in situ to avoid injuring the normal tissue.

Histological examination demonstrated the presence of a cavernoma. Postoperatively, the only improvements were in the patient's-facial function (House-Brackmann Grade I). One-year follow-up MR scans demonstrated that no recurrence had occurred (Fig. 2b). The sensorineural hearing loss was unchanged, and the facial function completely recovered.

Case 3. A 21-year-old female complained of progressively blurred vision for several months. Her visual symptoms suddenly worsened on the night before admission. A neurological examination revealed total visual loss in the right eye and a left temporal field deficit in the left eye. Brain MR revealed a nodular optochiasmatic mass that was inhomogeneously hyperintense on T1images and iso-hypointense on T2 images and apparently consisted of blood of different ages (Fig. 3a).

Surgical treatment was immediately planned. Via a right orbitozygomatic craniotomy, the lesion which consisted of dark blood was approached. The optic apparatus appeared oedematous, and had evidence of hemosiderin staining. Through this area a gentle dissection was performed and a complete removal of the cavernoma was achieved. The hemosiderin-stained tissue was left in situ to avoid damage to the complex structures.

Histological examination revealed a vascular malformation that corresponded to a cavernoma (Fig. 3b).

Postoperatively, the patient's visual examination results remained stable, and she experienced no complications.

Three months after the surgery an insignificant recovery of the visual deficit was registered in the right eye, and MR revealed the complete removal of the lesion. At a 7-month follow-up a mild recovery of the patient's visual acuity and an improvement in her field deficit were observed. Subsequent MR excluded recurrent cavernoma (Fig. 3c).

2. Discussion and conclusions

CMs are found in approximately 0.4%–0.9% of the population, and they account for 10%–20% of all cerebrovascular malformations.

CMs are classified as either intra-axial (i.e. in the brain stem, cerebellum, or cerebral hemisphere) or extra-axial (i.e. primarily in the floor of the middle cranial fossa, particularly the cavernous sinus). CMs typically occur in the supratentorial compartment (80%), and such lesions are followed in frequency by infratentorial lesions (15%) and lesions within the spinal cord (5%) [9–14].

CN CMs and especially those with isolated appearance are extremely rare as revealed by a literature review (Table 1).

CN CMs are associated with a risk of neurological deterioration during their natural evolution, As widely accepted in the literature, patients with symptoms experience inexorable progression to CN dysfunction [8–11].

Direct nerve injury, compressive ischaemia, oedema, and/or irritation due to intralesional haemorrhage are thought to be the causes of CN CMs.

Subarachnoid haemorrhages result from the bleeding of CN CMs and likely have less serious consequences than those expected from intra-axial haemorrhage which have an obviously different outcomes. However, bleeding from CN CMs can induce neurological impairment via mass effect that are not restricted to the cranial nerves but also influence the brainstem and have potentially severe effects.