Oculomotor nerve cavernous malformation

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Oculomotor nerve cavernous malformation (ONCM) is a rare vascular lesion involving the third cranial nerve. It is a subtype of **intraneural cavernous malformations (CMs)** that can occur in cranial or peripheral nerves. While cavernous malformations are more common in the brain and spinal cord, their occurrence in cranial nerves, particularly the **oculomotor nerve (CN III)**, is extremely rare.

Clinical Presentation

Patients with ONCM may present with symptoms related to oculomotor nerve dysfunction, including: - **Ophthalmoplegia** (partial or complete) - **Ptosis** (drooping of the upper eyelid) - **Diplopia** (double vision) - **Pupil involvement** (anisocoria, sluggish or nonreactive pupil) - **Headache** or retro-orbital pain

Pathophysiology

- ONCMs are **low-flow vascular malformations** composed of dilated, thin-walled blood vessels. - They are prone to **microhemorrhages**, which can cause progressive or sudden neurological deficits. - The **cavernous sinus segment of CN III** is a common location due to the vascular supply of the region.

Imaging Features

- **MRI** is the modality of choice:

1. **T2-weighted images**: Heterogeneous signal intensity due to blood degradation products.
2. **T2*-GRE/SWI**: Hypointense lesions due to hemosiderin deposition.
3. **T1-weighted images**: Variable signal intensity depending on hemorrhagic stages.
4. **No significant enhancement** on contrast-enhanced images.

- **Differential diagnoses** include:

1. Schwannoma
2. Meningioma
3. Tolosa-Hunt syndrome
4. Aneurysms of the cavernous ICA

Management

- **Observation**: If asymptomatic or minimally symptomatic, serial MRI may be recommended. - **Surgical resection**: Considered in cases with progressive symptoms, recurrent hemorrhage, or disabling diplopia. However, surgery is **challenging** due to the risk of damaging the nerve. - **Stereotactic radiosurgery (SRS)**: Rarely used but may be considered in select cases.

Prognosis

- Variable, depending on lesion size, location, and hemorrhagic events. - Neurological deficits may persist if significant nerve damage occurs.

Case reports

Martínez-Macho et al. from the University Hospital La Princesa describe a case of an oculomotor nerve cavernous malformation, which was managed conservatively. The authors also review the outcomes of other therapeutic options based on cases documented in the existing literature. Case report: A 36-year-old woman presented to the Emergency Department with a headache in the left temporal region, predominantly at night, along with mild left palpebral ptosis and binocular diplopia of 2 months duration. A brain MRI revealed a lesion exhibiting typical characteristics of CA in the left lateral region of the interpeduncular cistern, in close contact with the left oculomotor nerve. After considering treatment options and in consultation with the patient, a conservative management plan with periodic MRI follow-up was chosen. After 2 years of follow-up, the patient showed favorable progress. Although exceptional, CA should be considered in the differential diagnosis of other more common extra-axial lesions involving CN. The therapeutic management of a CA of the oculomotor nerve remains controversial due to the limited number of cases described in the literature. The authors suggest that for individuals with asymptomatic or mildly symptomatic CA affecting the oculomotor nerve, a conservative treatment approach is the most suitable choice to preserve neurological function. In cases characterized by progressive symptoms, a history of recurrent bleeding, or evidence of lesion enlargement on sequential imaging assessments, total microsurgical resection should be considered [1)].

A 47-year-old male with gradual CN III palsy. Initial imaging showed no significant findings, but a follow-up MRI revealed a growing lesion along CN III. Intraoperative findings confirmed a CN III CM. Diagnosing and treating CN III CM are complex. Radiological findings lack specificity, requiring

consideration of various diagnoses for patients with isolated CN III palsy and abnormal radiological findings.

For Zohdy et al. surgery is the gold standard, aiming for complete lesion removal while minimizing neurological complications [2)]

A 64-year-old male experienced diplopia and left-sided ptosis. Magnetic resonance imaging (MRI) revealed a lesion consistent with a CM affecting the cisternal segment of the left oculomotor nerve. The patient underwent a left-sided frontotemporal craniotomy for surgical resection of the lesion. Postoperatively, the patient's symptoms improved, and follow-up imaging confirmed the complete removal of the CM. This case underscores the importance of considering CMs in the differential diagnosis of oculomotor nerve palsy and highlights the efficacy of surgical intervention in such rare presentations [3)]

A 71-year-old male with a two-month history of progressive oculomotor nerve paralysis. Magnetic resonance imaging (MRI) suggested a CA of the oculomotor nerve. The patient underwent a complete resection of the lesion via a subtemporal approach, with preservation of nerve integrity. Histopathological examination confirmed the diagnosis of CA. Postoperatively, there was no improvement in the patient's oculomotor function. The study highlights that while early nerve-sparing surgical excision may offer the potential for functional recovery, delays in treatment often result in irreversible deficits. [4)]

A 33-year-old man experienced progressive left eye symptoms over six months, including pupil dilation, ptosis, and double vision. Neurological examination revealed left oculomotor nerve palsy. MRI showed a 7mm extra-axial mass near the left uncus and cavernous sinus, initially suggesting meningioma. Surgical exploration, however, revealed a reddish-brown nodular mass arising from the left oculomotor nerve. Histopathology confirmed it as a cavernous angioma. jkns

Postoperatively, the patient's oculomotor nerve palsy worsened but showed improvement after two months. At six months, diplopia persisted in right lateral gaze, with slight narrowing of the palpebral fissure and a dilated pupil. No recurrence was observed during an 18-month follow-up. This case underscores the importance of considering cavernous angiomas in differential diagnoses of cranial nerve dysfunctions and highlights the challenges in the preoperative identification and surgical management of these rare lesions [5)].

1)
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5)

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