

# CR

## CASE REPORT

*E. P. C. M. S.*

# Cerebral Cavernous Malformation presenting as an Oculomotor Nuclear Syndrome

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### ABSTRACT

We wish to present a patient who had unilateral third nerve palsy and contralateral superior rectus muscle paralysis as a result of a lesion in the midbrain produced by a cerebral cavernous malformation. This oculomotor nuclear syndrome is a rare presentation of mesencephalic trauma, neoplasms, and vascular conditions and is the third case described in the literature associated with cerebral cavernous malformation. Relevant neuroimaging is shown.

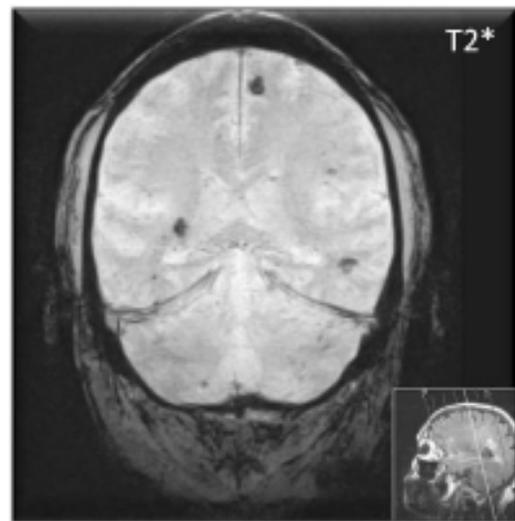
### INTRODUCTION

The oculomotor nuclear syndrome is a distinct ophthalmoplegia with partial or complete paralysis of the third cranial (oculomotor) nerve ipsilateral to the lesion and a paralysis of the superior rectus muscle contralateral to the lesion. This presentation is due to the superior rectus muscle innervation which comes from the contralateral third cranial nerve. This syndrome is usually seen in vascular, traumatic or neoplastic lesions of the mesencephalon (midbrain) and can present with other neurological deficits. We wish to report a case of cerebral cavernous malformation that presented with a left oculomotor nuclear syndrome.

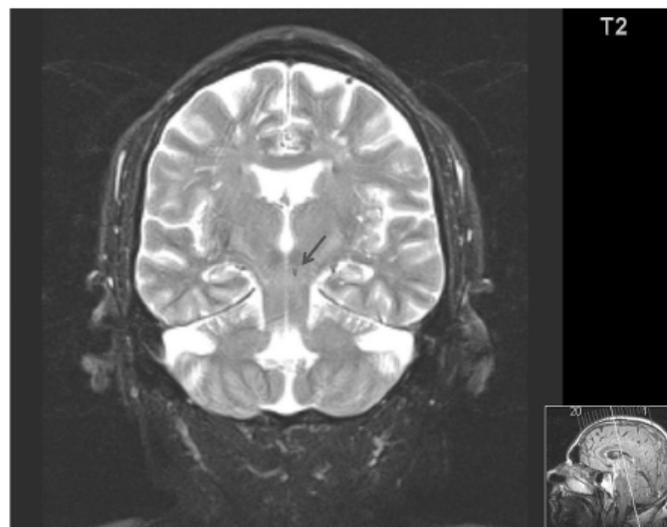
### CASE

A 62-year-old Caucasian right-handed man, casual worker with history of chronic arterial hypertension, came to the emergency department complaining of sudden onset of diplopia and continuous, moderate global headache for two days. On physical examination the blood pressure was 145/100mmHg with a regular heart rate of 98bpm. Complete dysfunction of the left third cranial nerve was evidenced by ipsilateral palpebral ptosis, paralysis of vertical gaze and ocular adduction, and a dilated, unreactive pupil. The external ocular movements in the right eye were normal except for paralysis of the upward gaze. The rest of the neurologic and physical examination was normal as well as initial laboratory work.

Magnetic Resonance Imaging (MRI) of the brain showed multiple supra and infra tentorial lesions with mixed signal intensity and a central reticulated core surrounded by a dark ring lesions best seen in T2\*, consistent with cerebral cavernous malformation (CCM) (Image 1). A 5 mm hypointense ringed lesion with inner hyperintensity was identified in the mesencephalon in T2 weighted coronal cuts. That lesion was accounted for the patient's symptoms and signs (Image 2). Surgical intervention was not considered due to the localization of the lesion. Symptoms and signs have remained unimproved; and two years after the initial presentation he has not had any new neurologic symptom.



**Image 1.** MRI of the brain. T2\* window. Coronal cut showing multiple supra and infratentorial with mixed signal intensity and a central reticulated core surrounded by a dark ring lesion.



**Image 2.** MRI of the brain. T2 weighted window, coronal cut. Arrow pointing at the left mesencephalic lesion accounted for the patient's current symptoms.

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Genetic testing showed a 7p13-15 mutation that produced an anomalous gene CCM2, implicated in the cerebral cavernous malformation.

**Discussion:**

CCMs are vascular malformations of clustered enlarged capillary with a single layer of endothelium with slow blood flow and little to no neural tissue. The size of the lesion can vary usually proportional to the amount of bleeding the lesions have sustained.<sup>1</sup> Approximately 50-75% of people with CCMs will have symptoms such as seizures, focal neurologic deficits, headaches, and cerebral hemorrhage. CCMs would usually present between the second and fifth decades of life.<sup>2,3</sup>

The neurological and ophthalmic findings in this patient were compatible with a left oculomotor nuclear syndrome (ONS). The upward gaze paralysis in the right eye is explained by the crossed innervation of the right superior rectus.<sup>11</sup> CCM is diagnosed by MRI; the characteristic lesion of mixed signal intensity with a central reticulated core surrounded by a dark ring is presumed to be produced by deposition of hemosiderin from prior hemorrhages.<sup>4</sup> In 1994 Zabramski et al<sup>5</sup>, described four types of lesions on spin echo and gradient echo signal that could be correlated histopathologically and clinically. Also, three genes have been linked to the familial form of CCM: KRIT1, CCM2, and PDCD10. Molecular genetic testing for all three genes is available on a clinical basis. CCM2 mutations, such as seen in this patient, account for 13%-20% of all familial CCMs.<sup>7-9</sup>

Currently, there is no medical treatment to control this condition and surgical removal have been successfully attempted for lesions associated with intractable seizures or focal deficits from recurrent hemorrhage or mass effect<sup>10</sup>, but this is limited to the accessibility of the lesion. Asymptomatic lesions are not routinely treated. This is the third documented case of a mesencephalic CCM presenting as an oculomotor nuclear syndrome.<sup>11, 12</sup>

Sometimes the oculomotor nuclear syndrome has other accompanying brain stem signs. Oculomotor nuclear syndrome with contralateral hemiplegia (Weber's syndrome) indicates concomitant involvement of the corticospinal tract in the midbrain. Also, contralateral ataxia and action kinetic tremor (Benedikt's syndrome) indicates involvement of the red nucleus in the midbrain.

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