CASE REPORT

Acute Transient Oculomotor Nerve Palsy from Presumed Cavernous Angioma in an Infant

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ABSTRACT

Cavernous angiomas of the cranial nerves are extremely rare, and those of the oculomotor (third) cranial nerve are rarer still, with no previous cases having been reported in the literature. We report herein the first case of a presumed cavernous angioma involving the subarachnoid portion of the left third nerve presenting as an acute left third nerve palsy in an infant. The child was followed without intervention and the palsy completely resolved. Given the poor functional results with attempted surgical excision and the potential for spontaneous improvement in oculomotor nerve function, it is reasonable to follow patients with these lesions without intervention.

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Introduction

Cavernous angiomas are vascular malformations composed of large sinusoidal capillary spaces that do not invade surrounding neural tissue. These lesions are most frequently intracerebral, but in rare instances have been reported to occur in the peripheral nervous system. Cavernous angiomas of the cranial nerves are extremely rare,¹ and those of the oculomotor (third) cranial nerve are rarer still, with nine previous cases having been reported in the literature.^{2–9} We report herein the first case of a presumed cavernous angioma of the subarachnoid portion of the left third nerve presenting as an acute, but transient left third nerve palsy in an infant.

Case report

A 3-month-old male infant with no known medical history and an uncomplicated prenatal course and delivery presented 3 days after the acute onset of left eyelid ptosis, decreased extraocular motility with an associated left exotropia and hypotropia, and anisocoria with the left pupil larger and less reactive than the right to direct and consensual light stimulation. By

history, as well as by photography performed by his parents, the child's appearance previously had been normal (Figure 1), and his ptosis and ocular motility had already begun to improve (Figures 2 and 3). On induced tropia testing, he had a fixation preference for the right eye. All findings were consistent with a left oculomotor nerve paresis. The child did not exhibit any evidence of discomfort or irritation at this time and was afebrile. Magnetic resonance (MR) imaging revealed enlargement of the initial subarachnoid portion of the left oculomotor nerve that was hyperintense on T1-weighted images, hypointense on T2-weighted images using a constructed interference steady-state (CISS) sequence (Figure 4). It enhanced after intravenous injection of gadolinium (Figure 5) and on post-contrast susceptibility-weighted sequences (Figure 6). An MR angiogram showed, no evidence of an aneurysm, arteriovenous malformation, or other high-flow vascular lesion. Based on the MR findings and clinical examination, a diagnosis of probable cavernous angioma of the left oculomotor nerve was made. As the patient was already improving by history and given the risks of surgical intervention, a decision was

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Figure 1. Appearance of patient 2 weeks before onset of left oculomotor nerve paresis.

Figure 2. Appearance of patient at onset of left oculomotor nerve paresis. Note obvious left ptosis, exotropia, and anisocoria (A) as well as limitation of adduction of left eye on attempted right gaze (B).

Figure 3. Appearance of patient at presentation 3 days after onset of left oculomotor nerve paresis. Note marked improvement in ptosis and ocular alignment compared with presentation (see Figure 2).

Figure 4. Magnetic resonance imaging at time of presentation. Axial image obtained by constructive interference in steady state (CISS) sequence, a strong T2-weighted gradient echo technique shows a lesion corresponding to the initial subarachnoid portion of the left oculomotor nerve (large arrow). Note relationship of the lesion to the left posterior cerebral artery (small arrow).

Figure 5. Magnetic resonance imaging (MRI) at the time of presentation. Coronal multiplanar reconstructed image using volumetric interpolated breath-hold examination (VIBE) sequence shows that the oculomotor nerve lesion (arrow) is located just beneath the left posterior cerebral artery at the junction of the P1 and P2 segments.

Figure 6. Magnetic resonance imaging (MRI) at time of presentation. Axial, post-contrast, susceptibility-weighted image (SWI) shows that the lesion (arrow) is vascular in nature.

made to follow the patient closely without intervention. Two weeks after the acute episode, his examination had become almost completely normal (Figure 7), with no ptosis or ophthalmoparesis, but very mild residual anisocoria.

Figure 7. Appearance of patient 18 days after onset of left oculomotor nerve paresis shows complete resolution. Note lack of ptosis and normal ocular alignment. There is very slight residual anisocoria.

Discussion

This is the first published report of a presumed cavernous angioma involving the subarachnoid portion of the oculomotor nerve in an infant. Cavernous angiomas of the cranial nerves, especially those involving the ocular motor nerves, are extremely rare. In a review of 58 cranial nerve angiomas,¹ 21 were located within one of the optic nerves or chiasm, 26 involved the facial or auditory nerves, five cases involved the oculomotor nerve, three cases involved the trochlear nerve, one case involved the trigeminal nerve, and one case involved the hypoglossal nerve. There are a total of nine cases involving the oculomotor nerve in the literature. Five presented with sudden onset of a third nerve paresis and occurred without precipitating factors in adolescent or adult males (Table 1).^{2,3,5,8} The acute clinical presentation of these cases was thought to be due to hemorrhage within the lesion. The remaining four cases presented with slowly progressive symptoms and signs. One of these cases was a young man who experienced progressive third nerve dysfunction over a 6-month period.⁶ Another patient was a young woman with galactorrhea, amenorrhea, and a visual field defect from compression of the pituitary stalk and optic chiasm by the lesion.⁴ The third patient presented with chronic facial pain that had no clear relationship to the lesion.⁷ The final patient was a 71-year-old man with a 2-month history of a progressive oculomotor nerve paresis.⁹ Interestingly, two of the reported cases occurred in patients with Roberts Syndrome,^{2,5} a rare genetic condition associated with craniofacial

and ocular anomalies, mental retardation and phocomelia of the limbs.

At issue in the rare cases of symptomatic known or presumed cavernous angiomas of the oculomotor nerve is their appropriate management. There would appear to be three options: 1) remove the lesion completely, including resection of a portion of the involved nerve; 2) perform a partial resection, attempting to leave the nerve intact; and 3) observe without intervention. As cavernous angiomas may rebleed, it is reasonable to consider complete resection of an oculomotor nerve angioma if a patient already has a complete third nerve palsy.^{2,3,5,8,9} However, even complete oculomotor nerve pareses can recover, and, indeed, there is a report of one patient with a cavernous angioma of the oculomotor nerve, who experienced complete recovery of oculomotor nerve function 6 months after partial resection of the angioma.⁸ Furthermore, the author who published the first case of a cavernous angioma involving the oculomotor nerve, for which he performed a complete resection,² subsequently wrote a letter to the editor of another journal stating that he regretted his decision to do so and now thought that simply opening the angioma and removing its contents as completely as possible without disrupting the nerve would be the best course of action.¹⁰ On the other hand, the patient reported by Obaid et al. underwent this procedure and had no return of oculomotor nerve function.⁹

A longitudinal study of intracranial cavernous angiomas in 122 patients reported a hemorrhage rate of 0.6% annually for patients who never had a previous hemorrhage; however, this rate increased to 4.5% per year for patients who already had one

Table 1. Review of published cases of pathologically proven cavernous angiomas involving the oculomotor nerve.

Article/date/ref #	Age	Gender	Presenting sign/symptom	Treatment	Post-operative oculomotor nerve function
Scott et al. (1983) ²	14	Male	Acute 3rd nerve palsy	Complete excision (nerve transection)	None (unchanged)
Yamada et al. (1986) ³	33	Male	Acute 3rd nerve palsy	Complete excision (nerve transection)	None (unchanged)
Matias-Guiu et al. (1990) ⁴	36	Female	Pituitary stalk compression	Partial excision	None (worsened)
Ogilvy et al (1992) ⁵	28	Male	Acute 3rd nerve palsy	Complete excision (nerve transection)	None (unchanged)
Park et al (2005) ⁶	33	Male	Subacute 3rd nerve palsy	Partial excision	Partial (unchanged)
Itshayek et al (2007) ⁷	25	Female	Facial pain	Partial excision	Full (unchanged)
Wolfe et al (2011) ⁸	69	Male	Acute 3rd nerve palsy	Partial excision	None (unchanged)
	26	Male	Acute 3rd nerve palsy	Partial excision	Full (improved)
Obaid et al (2014) ⁹	71	Male	Subacute 3rd nerve palsy	Complete excision (nerve left intact)	None (unchanged)

or more hemorrhages.¹¹ It is not known if these rates apply to cranial nerve angiomas. Thus, it is impossible to predict the long-term outcome of observation alone, as recurrent hemorrhages could result in a permanent complete or incomplete neuropathy or even subarachnoid hemorrhage. In addition, patients with recurrent hemorrhage from an oculomotor nerve cavernous angioma could develop aberrant regeneration of the nerve, resulting in significant visual dysfunction. On the other hand, the functional outcomes from attempted resection of cavernomas causing acute oculomotor nerve palsy also would appear to be poor (Table 1). Thus, we would recommend observation rather than intervention—at least initially—in those rare patients with cavernous angiomas of the oculomotor nerve.

Although the time course of onset and resolution of the lesion in a matter of days is typical of a cavernous angioma, we have no pathologic confirmation of this diagnosis. A recent study¹² reported several cases of cranial nerve schwannomas that produced cranial nerve palsies that spontaneously resolved, and in some cases, recurred; however, in the majority of these cases, the palsies resolved slowly over the course of several months not over a few days to a week. Thus, although we cannot say definitively that our patient harbors a cavernous angioma, the rapid onset and equally rapid resolution, as well as the lack of any progression of the child's third nerve paresis over more than 3 years, make this diagnosis the most tenable.

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