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Grand Rounds

An unusual case of binocular oblique diplopia in an 82-year-old man

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History

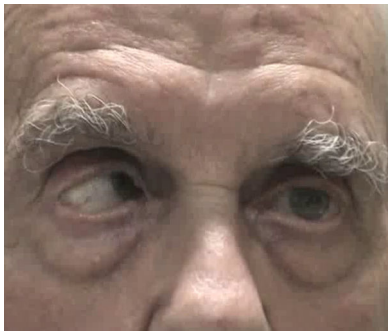
An 82-year-old man presented to the Neuro-Ophthalmology Clinic at Beth Israel Deaconess Medical Center for evaluation of binocular, oblique diplopia of 1 month's duration. His medical history was significant for benign prostatic hyperplasia, hypertension, and coronary artery disease. Review of systems revealed no additional major complaints. His family history was also unremarkable.

Examination

On examination, his pupils were symmetric, without any relative afferent defect. He was noted to have a left head tilt of 35°. A left head tilt was also present in the patient's driver's license photograph. There was a large-angle esotropia of approximately 50 prism diopters (PD). This esotropia measured approximately 45 PD on right gaze and >50 PD on left gaze. The right eye was also found to have a hypertropia of 20 PD in primary position, both at distance and at near. This worsened in left gaze to >25 PD and in right head tilt to 30 PD; it improved in right gaze to 6 PD and in left head tilt to 10 PD. There was also significant right inferior oblique overaction and complete abduction palsy of the left eye (Video 1). There was no globe retraction on adduction of the left eye, and no lid twitch, ptosis, or proptosis. Trigeminal function was normal. There was no facial asymmetry noted. The rest of the examination was unremarkable.

Ancillary Testing

A provisional clinical diagnosis was made of right trochlear nerve palsy and left abducens nerve palsy, with the assumption that one was chronic and the other new, because simultaneous onset seemed unlikely. Axial T1 postcontrast magnetic resonance imaging (MRI) revealed an enhancing lesion in the right ambient cistern along the course of the trochlear nerve, consistent with right trochlear nerve schwannoma (Figure 1) and thin section axial FIESTA images revealed left abducens nerve compression by an anterior inferior cerebellar artery (AICA) aneurysm (Figure 2).



Video 1. Sensorimotor examination revealing right hypertropia, right inferior oblique overaction, and complete abduction palsy of the left eye.

Treatment

The patient was offered endovascular coil embolization for treating the aneurysm as well as right inferior oblique recession and transposition of the left superior rectus muscle temporally, with bilateral medial rectus recessions, but he declined intervention.

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Digital Journal of Ophthalmology, Vol. 21

Digital Journal of Ophthalmology, Vol. 21

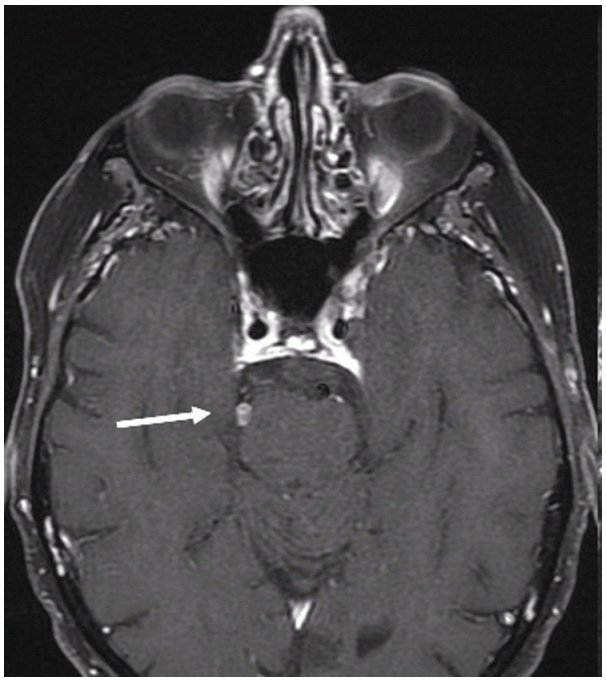

Figure 1. Axial T1 postcontrast image revealing an enhancing lesion in the right ambient cistern along the course of the right trochlear nerve (white arrow) consistent with right fourth nerve schwannoma.

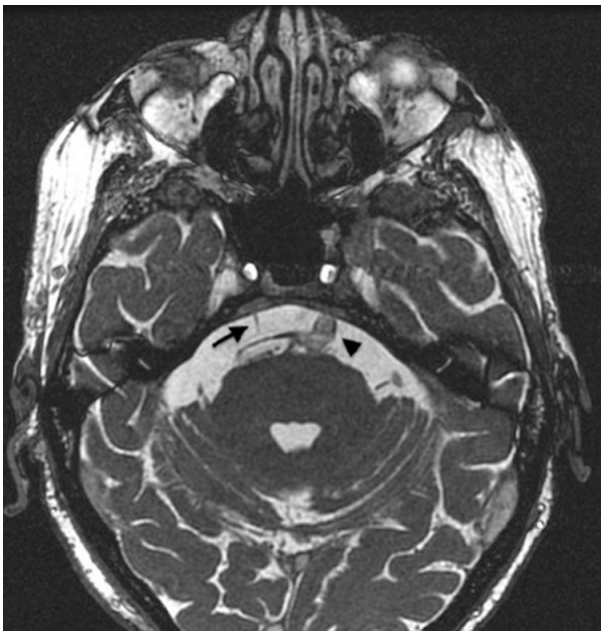

Figure 2. Thin-section axial FIESTA image revealing left sixth nerve compression by an anterior inferior cerebellar artery aneurysm (black arrowhead). The cisternal segment of the normal sixth nerve is seen on the right (black arrow).

Differential Diagnosis

The differential diagnosis for this case includes myasthenia gravis, Graves' ophthalmopathy, Miller-Fisher syndrome and leptomeningeal disease causing multiple cranial neuropathies. If the patient had myasthenia gravis, his symptoms and examination findings might fluctuate, and his examination might show a fatiguable ptosis and/or a Cogan's lid twitch. Graves' ophthalmopathy would classically be associated with restrictive limitation in extraocular movement and could include other signs, including proptosis, lid retraction, and/or scleral show. Miller-Fisher syndrome, like myasthenia, can cause extraocular motility impairment that may be difficult to localize anatomically but is typically associated with ataxia and areflexia. Leptomeningeal disease causing multiple cranial neuropathies would likely have other neurologic and possibly systemic manifestations.

Diagnosis and Discussion

Based on these findings, right trochlear schwannoma was the likely etiology of the right trochlear nerve palsy while left abducens nerve compression from the AICA aneurysm explained the left abducens nerve palsy. Subsequent correspondence with the patient's optometrist confirmed the fact that the trochlear nerve palsy was longstanding. Longstanding trochlear nerve palsies can often be differentiated from more acutely acquired palsies by the presence of a compensatory head tilt, as exhibited in this patient, and/or large vertical fusional amplitudes. The concurrence of right trochlear and left abducens nerve palsies, as seen in the present case, is rare and results in complex extraocular muscle dysfunction.

Acquired trochlear nerve palsies are most commonly secondary to head trauma, followed by microvascular ischemia and compressive lesions. They are much less commonly due to intrinsic neoplasms of the trochlear nerve.[1] Trochlear nerve schwannomas are extremely rare in patients without neurofibromatosis. The first case of trochlear schwannoma was described by King[2] in 1976, and fewer than 50 cases have since been reported in the literature. Patients may be asymptomatic or present with loss of function of the affected cranial nerve. Larger tumors may cause mass effect on adjacent cranial nerves or present with symptoms of brainstem compression.[1] MRI is useful for both diagnosis and follow-up of these lesions, which appear well demarcated, lie along the course of the nerve, and show homogenous enhancement.[3] Patients may be managed conservatively with serial scans; surgical intervention may be reserved for patients exhibiting signs of brainstem compression.[1]

Digital Journal of Ophthalmology, Vol. 21

Abducens nerve palsy is most commonly related to microvascular ischemia. The incidence of abducens nerve palsy associated with intracranial aneurysms is less than 4%.[4] Mechanisms by which an aneurysm may cause abducens nerve dysfunction include direct compression and an irritant effect of subarachnoid blood from rupture. Neurovascular compression of the abducens nerve may also result from dolichoectatic vertebral or basilar arteries, which may compress the nerve close to the root.[4,5] Compression of the nerve in the prepontine cistern from an AICA aneurysm has rarely been described. Thin section MRI using CISS/FIESTA sequence is the imaging modality of choice for evaluating neurovascular compression of the abducens nerve.[5] Aneurysms causing ophthalmoplegia may be treated by endovascular coiling although, especially in older patients and patients with microvascular risk factors, this treatment does not usually result in complete resolution of nerve palsy, and there may often be residual diplopia, especially in upward or downward gaze.[4,6]

Literature Search

The authors searched PubMed on July 27, 2014, for English-language articles (1950-present) using the following search terms: *diplopia*, *trochlear schwannoma*, *anterior inferior cerebellar artery aneurysm*, *fourth nerve palsy, trochlear palsy, abducens palsy, sixth nerve palsy*.

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