Pupil-Sparing, Painless Compression of the Oculomotor Nerve by Expanding Basilar Artery Aneurysm

A Case of Ocular Pseudomyasthenia

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**Background:** Oculomotor nerve paresis may have relatively benign but also life-threatening causes. Distinguishing between these is of great clinical importance.

**Objective:** To reveal a potential pitfall of the clinical evaluation of oculomotor nerve paresis.

**Patient:** Single case observation.

**Results:** A 56-year-old man had fluctuating diplopia and fatigable ptosis, promptly relieved by intravenous edrophonium, leading to the diagnosis of ocular myasthenia gravis. His pupillary function was intact. A few days after the initial diagnosis, he suffered a subarachnoid hemorrhage secondary to the rupture of a basilar artery aneurysm. His ocular symptoms were related to aneurysmal oculomotor nerve compression.

**Conclusion:** Patients with oculomotor nerve dysfunction need more detailed evaluation because the underlying cause cannot be safely determined on a clinical basis.

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ANEURYSMS ARISING FROM various segments of the arterial circle of Willis are the most common source of nontraumatic subarachnoid hemorrhage. Patients with intracranial aneurysms may have various premonitory symptoms. Focal neurological deficits occur from compression or ischemia secondary to arterial spasm. The oculomotor nerve is frequently compressed by an expanding aneurysm of the posterior communicating artery, but occasionally a basilar artery aneurysm can compress the oculomotor nerve, causing ipsilateral pupillary dilation and ptosis. The initial clinical symptoms of intracranial aneurysms can be very atypical and insidious and can perfectly mimic the features of other neurological diseases, such as migraine headache or ischemia of the oculomotor nerve.

We describe a 56-year-old man who had an isolated, painless, pupil-sparing oculomotor nerve lesion caused by an expanding basilar artery aneurysm that had the clinical features and laboratory findings typical of ocular myasthenia gravis. We draw attention to potential pitfalls of the evaluation of isolated oculomotor nerve lesions and suggest a more extensive evaluation for patients with this problem.

A 56-year-old white man without any significant past medical problems experienced double vision an hour after waking up in the morning. Double vision lasted for about 20 minutes, resolved for a few minutes, and then returned for an additional 20 to 30 minutes. He rested his eyes, and the diplopia resolved for the rest of that day. The next day, the patient had double vision in the morning for 20 minutes; it resolved temporarily after he closed his eyes and then returned for another 30 minutes. On the day of admission, he developed diplopia that did not resolve.

At the initial interview, he complained of seeing 2 images side by side. The separation of the images was more severe on the left lateral gaze. He denied any current or previous headache, dizziness, loss of consciousness, photophobia, weakness, numb sensation, neck pain, and nausea or vomiting.

Results of a neurological examination revealed equal, round pupils that reacted to light directly and consensually. (The pupils were tested in a darkened room.) Visual fields were full on confrontational testing. He had normal, conjugate upward and downward gaze and also intact right lat-
eral gaze. On attempted left lateral gaze, he failed to adduct his right eye (Figure 1) and complained of diplopia, consistent with a complete right medial rectus palsy. His palpebral fissures were equal at rest (Figure 1), but after a 30-second sustained upward gaze, he developed significant right-sided ptosis (Figure 2). Results of the rest of the examination were normal. Findings of the head computed tomograph without contrast were unremarkable.

Based on the fluctuating nature of the diplopia and isolated right medial rectus palsy, the absence of pupillary involvement, and ptosis provoked by sustained effort, ocular myasthenia gravis was suspected. An intravenous injection of 5 mg of edrophonium significantly improved right-eye adduction. The patient was initially given pyridostigmine. He was scheduled for repetitive nerve stimulation and a single-fiber electromyogram. Six days later, a family member found him to be increasingly lethargic and later unresponsive. Upon admission, computed tomography showed prominent subarachnoid hemorrhage with blood extending into the lateral ventricles. A 4-vessel cerebral angiogram revealed an 8-mm basilar apex aneurysm (Figure 3). The internal carotid and posterior communicating arteries had normal appearances. The aneurysm was successfully thrombosed by coiling. The patient made a good recovery and now lives independently.

**COMMENT**

The oculomotor nerve compression in this patient had many atypical features that obscured the diagnosis. One of these was the intact pupillomotor function of the compressed nerve. The characteristic signs of oculomotor nerve palsy due to compression by intracranial aneurysm have been known since the 19th century. In 1958, Rucker reported 64 cases of oculomotor compression by aneurysms in which 97% of patients had dilated pupils. Compressive lesions of the oculomotor nerve are thought to be characterized by an initial pupillary deficit and, later, extracocular muscle paresis. In the cases of ischemic lesions of the oculomotor nerve, the extracocular muscles become paretic, but the pupil is reactive to light. These phenomena are explained by the topographical arrangement of the pupillomotor fibers of the nerve and the pattern of their blood supply. These basic concepts, however, have been challenged. Multiple case reports and a nationwide survey have revealed cases of oculomotor compressions in which the pupils were not affected. Sparing of the pupils therefore does not always exclude compressive etiology. Various explanations exist for the sparing of pupillomotor fibers. A widely accepted explanation is that the pupillomotor fibers are not evenly distributed on the surface of the nerve but rather form a distinct bundle on its dorsomedial-medial aspect. Therefore, any compressive force acting from other directions spares this bundle and damages the somatomotor axons. Alternatively, the position of the bundle may be different in certain people so that compressive forces from the usual directions do not affect it. Another theory is that in cases of slowly evolving compressions the pressure is evenly distributed and the smaller-diameter pupillomotor fibers, which are more pressure-resistant, may escape the damage.

Rarely, oculomotor nerve palsy has been described as a result of basilar artery aneurysms. In cases in which the oculomotor nerve was compressed by basilar aneurysms, the involvement of the nerve tends to be milder, probably because of the greater mobility of the nerve in the interpeduncular fossa. The mobility of the nerve segment might explain the fluctuation of the symptoms in our patient. Because of their topographical relationship, basilar artery aneurysms may push on the ventral-medial aspect of the nerve first, resulting in partial oculomotor nerve paresis.

Another unusual phenomenon seen in our patient was the total lack of headache. As the oculomotor nerve does not contain pain sensation fibers, the headache accompanying its compression is thought to be originating from the wall of the expanding aneurysm. Other cases of intracranial aneurysm without history of headaches have been described; therefore, the absence of headache does not exclude the presence of an aneurysm.
In addition to the atypical features discussed above, the most misleading feature of the initial manifestation of the aneurysm in our patient was its striking similarity to that of ocular myasthenia gravis. The patient reported the highly characteristic fluctuating diplopia that improved after rest and the reversible ptosis provoked by sustained upward gaze. These features, in the presence of a positive Tensilon test result, were very suggestive of a disorder of the neuromuscular junction. There are a few disorders described in the literature that may imitate the features of myasthenia gravis. Tumors in the pineal region,9 cavernous sinus meningiomas,10,11 benign superior orbital fissure syndrome,12 sphenoid ridge meningiomas, cavernous sinus chondrosarcomas,13 giant vertebrobasilar aneurysm,13 and carotid aneurysm14 were diagnosed initially as myasthenia gravis, sometimes based on a false-positive Tensilon test result. However, many of the patients had headaches on initial examinations. The Tensilon test has several limitations, including the subjectivity of the interpretation. In ocular myasthenia, the patient’s reliability is not a concern because isolated extracocular muscle palsy cannot be simulated. Therefore, in our case, the improvement of right-eye adduction could be safely interpreted as an objective sign. The increase in strength following administration of edrophonium in our patient may have multiple explanations. Edrophonium increases the transmission in intact nicotinic neuromuscular junctions and the muscarinic cholinergic synapses. The latter is the reason for edrophonium’s adverse effects. By inhibiting the cholinesterase enzyme, edrophonium may enhance the cholinergic transmission of the still-intact or only partially damaged fibers, resulting in a transient increase of strength. Another explanation of the improved strength is the known direct stimulating effect of edrophonium on the postsynaptic (muscle) membrane. These observations demonstrate that a positive Tensilon test result is a sensitive but not specific tool to diagnose a neuromuscular junction problem. It alone cannot confirm the diagnosis of ocular myasthenia gravis.

We propose a more detailed evaluation of patients with a painless, isolated oculomotor nerve lesion. Regardless which extraocular muscles are involved and to what extent or how the pupils are affected, these patients should have brain magnetic resonance imaging with gadolinium enhancement. If no compressive lesion is visualized, a magnetic resonance angiograph may detect aneurysms. If the magnetic resonance angiograph is nonrevealing but the suspicion of aneurysm is high, a computed tomographic angiogram or conventional angiogram should be obtained. The diagnosis of ocular myasthenia or a diabetic-vascular oculomotor nerve lesion should be made only after excluding a compressive lesion.

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