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 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.

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eral gaze. On attempted left lateral gaze, he failed to ad-
duct his right eye (Figure 1) and complained of diplo-
pia, 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 sig-
nificant right-sided ptosis (Figure 2). Results of the rest 
of the examination were normal. Findings of the head com-
puted tomograph without contrast were unremarkable. 

Based on the fluctuating nature of the diplopia and 
isolated right medial rectus palsy, the absence of pupil-
lar involvement, and ptosis provoked by sustained ef-
fort, ocular myasthenia gravis was suspected. An intra-
venous 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 increas-
ingly lethargic and later unresponsive. Upon admission, 
computed tomography showed prominent subarach-
noid 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 nor-
mal appearances. The aneurysm was successfully throm-
bosed 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 com-
pressed nerve. The characteristic signs of oculomotor 
nerve palsy due to compression by intracranial aneu-
rysm 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 pu-
pils. Compressive lesions of the oculomotor nerve are 
thought to be characterized by an initial pupillary defi-
cit and, later, extraocular muscle paresis. In the cases of 
ischemic lesions of the oculomotor nerve, the extraocu-
lar muscles become paretic, but the pupil is reactive to 
light. These phenomena are explained by the topography-
ical arrangement of the pupillomotor fibers of the nerve 
and the pattern of their blood supply. These basic con-
cepts, however, have been challenged. Multiple case re-
ports 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 ex-
ist for the sparing of pupillomotor fibers. A widely ac-
cepted 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 as-
pect. Therefore, any compressive force acting from other 
directions spares this bundle and damages the somato-
motor axons. Alternatively, the position of the bundle may 
depend on 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 pres-
sure is evenly distributed and the smaller-diameter pu-
pillomotor 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 aneu-
rysms, 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 seg-
ment might explain the fluctuation of the symptoms in 
our patient. Because of their topographical relationship 
(Figure 4), basilar artery aneurysms may push on the 
ventral-medial aspect of the nerve first, resulting in par-
tial 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 accom-
panying its compression is thought to be originating from 
the wall of the expanding aneurysm. Other cases of in-
tracranial 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, cavernous sinus meningiomas, sphenoid ridge meningiomas, cavernous sinus chondrosarcomas, giant vertebrobasilar aneurysm, and carotid aneurysm 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 extraocular 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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Figure 4. Illustration of the anatomical relationship between a basilar aneurysm and the compressed oculomotor nerve.

REFERENCES


