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Clinical challenges

The pescatorial sixth

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(In keeping with the format of a clinical pathological conference, the abstract and key words appear at the end of the article.)

1. Case report

A 56-year-old man came to the emergency room complaining of binocular horizontal diplopia since late that morning. He denied trauma, alcohol, or drug consumption and felt otherwise well. He had been diagnosed with diabetes 5 years previously and had been taking metformin for 3 years.

His initial examination showed limitation of abduction in both eyes. C reactive protein was 0.68 mg/L, and he had an elevated blood glucose of 178 mg/dL. A computed tomography scan of his brain was normal. Ophthalmology was consulted for a provisional diagnosis of bilateral diabetic microvascular abducens palsies.

Visual acuity was 20/20 in each eye, and visual fields were full to confrontation. There was no relative afferent pupil defect. Slit lamp examination was unremarkable, and intraocular pressures were 15 mm Hg in each eye. Fundus examination was consistent with mild nonproliferative diabetic retinopathy without macular edema. Pupils were symmetric in light and dark. The lids were slightly asymmetric, with 1 mm less palpebral fissure on the right. He had an esotropia increasing in right and left gaze, with complete bilateral limitation of abduction (Fig. 1).

What is the differential diagnosis of his ocular motor deficit at this point?

What is the next step?

2. Comments

2.1. *Comments by Jason Barton, MD, PhD*

The photographs show absence of abduction past the midline of either eye. This certainly could be consistent with bilateral VI nerve palsies, but before leaping to the conclusion that this is neuropathic, it is always wise to step back and consider the broader differential of abduction deficits and think of which

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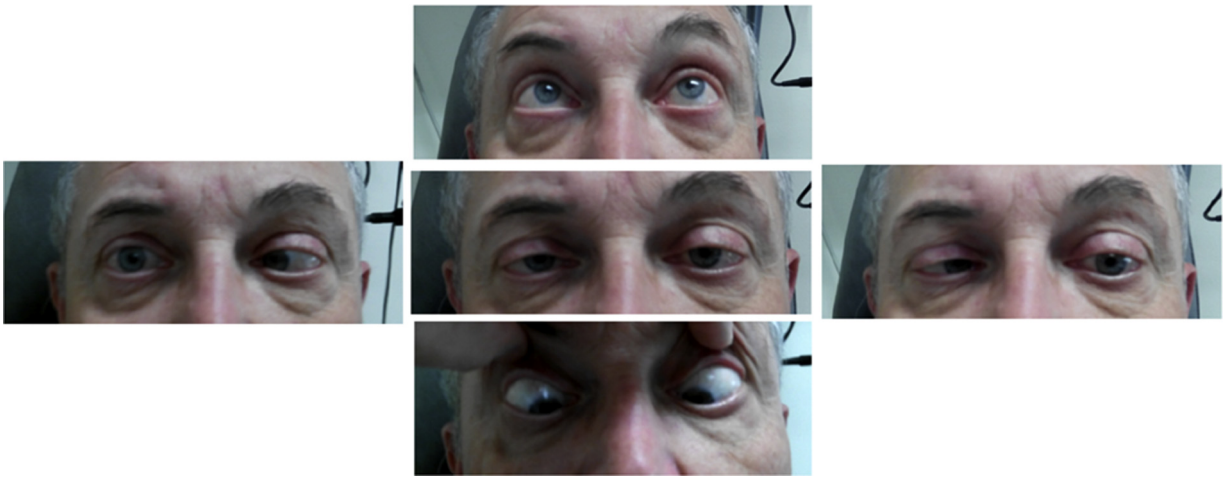

Fig. 1 – Gaze positions at presentation on the first day of the illness. He has bilateral complete limitation of abduction.

could present (1) bilaterally, (2) in a relatively isolated manner, and (3) in such an acute fashion.

Myopathies can cause abduction deficits. Chief among these is thyroid ophthalmopathy,[58] one of whose favorite targets is the medial rectus muscle. Restriction of this muscle consequent to this inflammatory process can cause an abduction deficit. This may be bilateral, but in most patients the tempo is a slowly progressive course. A more rapid "malignant" course occurs rarely, but would usually be associated with more chemosis and proptosis. Another restrictive myopathy is entrapment of the medial rectus by a fracture of the medial orbital wall; however, the trauma necessary to cause this is not the kind of small historical detail a patient would neglect to mention. Although not strictly a myopathy, sagging eye syndrome can present with diplopia and an esotropia because of downward displacement of the lateral rectus from degeneration of the lateral rectus-superior rectus bands in both eyes.[8] This rarely causes complete loss of abduction.

How about disorders of the neuromuscular junction? Myasthenia gravis is always a possibility. It is often said that if the ocular motor pattern does not conform to a nerve palsy, one should think of myasthenia, but it would be more to the point to say that, even if it does fit a typical neuropathic pattern, one should still think of myasthenia.[4] Myasthenia can mimic internuclear ophthalmoplegia,[18] superior oblique palsies,[47] and gaze palsies, for example, and it can be bilateral and develop rapidly, although progressing to complete abduction deficits over less than a day seems a bit dramatic. Botulism may also present with ophthalmoplegia, but there is usually also internal ophthalmoplegia, with blurred vision from impaired accommodation and fixed, dilated pupils, as well as bulbar and systemic weakness and signs of autonomic dysfunction. Nevertheless, on rare occasions there may be only ocular signs.[14]

Neuropathies include both focal nerve lesions, in this case bilateral VI nerve palsies, and diffuse nerve lesions, such as Miller-Fisher syndrome and Guillain-Barré syndrome. Acute unilateral VI nerve palsies have a long list of possible etiologies—with trauma, neoplasia, and ischemia being the leading contenders—affecting the nerve anywhere from its fascicular course in the pons, through the subarachnoid space, and into the cavernous sinus.[48] The emphasis changes somewhat when the palsy is simultaneously bilateral.[26] Bilateral fascicular VI nerve palsies can occur but usually have other cranial nerve or long tract signs, typically weakness from damage to motor tracts in the basis pontis. Nevertheless, this has been described with multiple sclerosis.[28] More commonly, bilateral VI nerve palsies arise from problems in the subarachnoid space,[26] such as trauma,[50] increased or decreased intracranial pressure, hemorrhage, infectious, inflammatory, or neoplastic processes in the cerebrospinal fluid or arising from the clivus. Because there are connections between the right and left cavernous sinus, lesions here sometimes present with bilateral signs. Bilateral abducens palsies have been described with cavernous sinus fistulae[22]; dilated conjunctival veins and proptosis would be clues to this diagnosis. Thrombosis, pituitary apoplexy, and neoplastic lesions of the cavernous sinus could also cause bilateral ocular motor palsies.

Unilateral or bilateral abducens palsies are described as unusual regional variants of Miller-Fisher syndrome or Guillain-Barré syndrome.[45,63] Other features of these syndromes may eventually develop over the subsequent days, namely ataxia and areflexia in Miller-Fisher syndrome or weakness and paresthesiae in Guillain-Barré syndrome.

Divergence insufficiency is a central vergence disorder in which there is a relatively comitant esotropia at distance and orthotropia at near;[30] however, this is not associated with complete loss of abduction. Wernicke encephalopathy is associated with a variety of central ocular motor deficits, including abducens palsies and gaze palsies, along with confusion and ataxia.

Various types of congenital strabismus can look like abducens palsies, including bilateral type I Duane syndrome[12] and Mobiüs syndrome.[56] Patients are generally aware of the longstanding nature of these deviations and do not complain of diplopia, but occasionally an observant physician may note this in an obtunded patient who cannot provide that history, and panic ensues until the typical distinguishing signs of these disorders are noted, such as the narrowing of the palpebral fissure on adduction in Duane syndrome[12] and the facial palsy of Mobiüs syndrome.[56] Decompensation of a congenital esophoria can cause new-onset horizontal diplopia but not likely complete loss of abduction in one or both eyes.