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CLINICAL CHALLENGES

PETER SAVINO AND HELEN DANESH-MEYER, EDITORS

Once upon a Cataract Surgeon

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

Case Report

A 60-year-old male ophthalmologist presented with 3 months of progressive binocular oblique diplopia, headache, and mental status changes. He had become increasingly forgetful, developed a tremor of his right hand and foot, and gradually found that he could not safely perform cataract surgery. His coworkers reported that increasingly over time he would make incorrect word choices in conversation and had difficulty expressing his thoughts. He felt "unstable" when he stood or walked. On several occasions over the following months, he believed that he heard household noises and that there were "strangers outside my house," although there was no one there. His past medical, surgical, and ocular histories were all unremarkable. There was hypertension in his family. His medications included piracetam, telmisartan, and aspirin. He did not smoke, but drank alcohol socially.

Best corrected visual acuity was 20/25 in the right eye and 20/20 in the left. Confrontation visual fields and fundus examination were normal. Pupils were symmetric and reactive without relative afferent pupillary defect. He had intermittent mild lid retraction and an intermittent chin-up position of about 5 degrees. He had reduced elevation of both eyes, more so the left, that could be partially overcome with the Doll's head maneuver (Fig. 1). He had a 6–7 prism diopter esotropia in primary, right, and left gaze. There was also a subtle left hypertropia in right gaze, which increased to 3 diopters in left gaze and to 4–6 prism diopters in upgaze. He was orthotropic in down gaze. There was no nystagmus. Slit-lamp biomicroscopy was normal, and intraocular pressures were 16 mm Hg in both eyes.

What is the differential diagnosis? What further work-up is indicated?

Comments

COMMENTS BY JASON J.S. BARTON, MD, PHD

By history this man has cognitive problems that include memory and word-finding difficulty, as well as hallucinations and delusions. He has some sort of asymmetric tremor and possibly gait and balance

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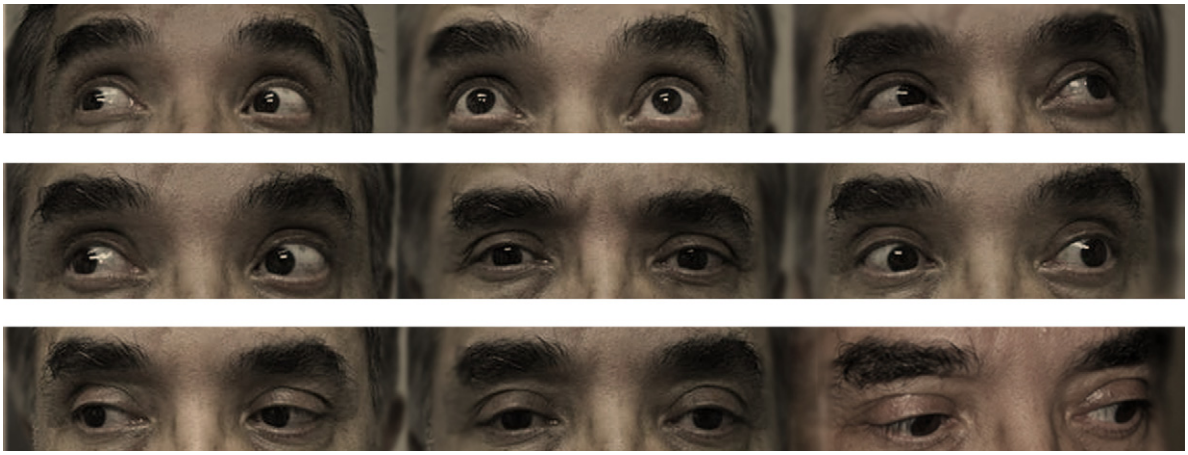

Fig. 1. Demonstration of underaction of elevation OU and esotropia in primary position, left and right gaze.

problems. A neurological examination would be invaluable to clarify the nature of the abnormal movement and balance issues, whether he has gait ataxia, and to determine if there are amnestic or aphasic elements correlating with his cognitive complaints. The neuro-ophthalmologic findings suggest a comitant esotropia and an incomitant left hypertropia. The latter could fit with a right inferior oblique paresis, but this pattern is more often mimicked by an incomitant skew deviation from brainstem disease. A comitant esotropia of rapid onset is not likely an abducens palsy. It could reflect a secondary decompensation of a pre-existing esophoria, but may also be due to divergence insufficiency or occur with cerebellar disease.

All of these findings point to a diffuse or multifocal process that may involve at a minimum the cerebrum, cerebellum, and brainstem. Regarding etiology there are two observations. First, he was engaged in the high-risk lifestyle of a cataract surgeon. Second, his problems are progressing over a relatively brief interval of a few months, though the tempo is a little uncertain. These leave a substantial list of potential causes to be considered.

Degenerative disorders that span this breadth of the neuraxis are somewhat rare. Diffuse Lewy body disease can cause cognitive changes with hallucinations, altered sleep–wake cycles, and parkinsonism, the latter of which could be responsible for gait instability.[10] The tempo of this disorder is usually a little slower than what is happening to this man, though.

As for infection, although the tempo is a bit slow for most classic bacterial processes, Whipple disease can cause diffuse cognitive changes as well as focal brainstem signs including a variety of ocular motor disorders. There is, however, no mention of lymphadenopathy, arthralgias, or gastrointestinal symptoms such as weight loss, diarrhea, or abdominal pain.[8] Nevertheless, about 4% of cases of this rare disorder present without systemic features, and as it can be treated successfully with antibiotics, the diagnosis should not be missed. Tuberculosis could create multifocal central dysfunction, sometimes even if the chest x-ray is unremarkable. Among viral conditions progressive multifocal leukoencephalopathy is the main possibility, often causing subacute onset of a variety of visual disturbances along with ataxia and dysphasia, though unlikely diplopia or brainstem involvement.[4] Also, this is more common in the immunocompromised. Prion disorders can cause rapidly progressive cerebral, cerebellar, and brainstem dysfunction; if his "tremor" turns out to be myoclonus, this would heighten suspicion of this class of diseases.

Among toxic disorders, mercury poisoning stands out as a candidate for a constellation of cognitive problems, hallucinations, visual problems, and possibly ataxia.[20] This is rarely encountered nowadays, however. Toluene encephalopathy can present with behavioral changes, hallucinations, ataxia and diplopia, but this is predominantly seen in children.[13]

Turning to metabolic conditions, the presentation of Wernicke encephalopathy is classically considered the triad of confusion or impaired consciousness, ataxia with imbalance, and ocular motor disorders, which include sixth nerve palsies, internuclear ophthalmoplegia, and gaze palsies.[25] It usually presents more acutely than seen here in a patient with malnourishment or alcoholism, and its earlier stages are often characterized by anorexia, nausea, and vomiting. Hyperintensities on magnetic resonance imaging (MRI) fluid-attenuated inversion-recovery (FLAIR) imaging in the thalami, mamillary bodies, and brainstem tegmentum are classic, but their sensitivity for the diagnosis is not yet certain.[30]