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Hearing and Vision Loss in an Older Man

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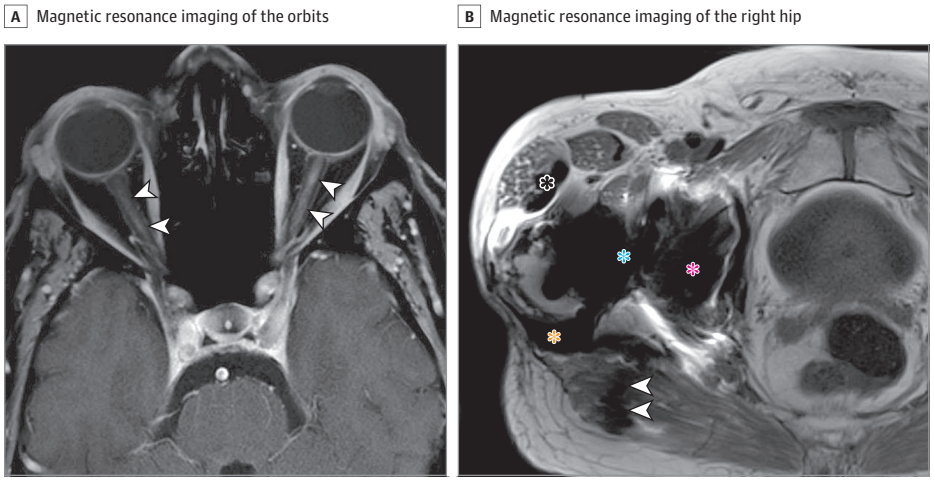

Figure. A, Axial postgadolinium, fat-suppressed, T-1–weighted magnetic resonance image of the orbits, demonstrating left greater than right enhancement of the optic nerves (arrowheads). The enhancement is centrally located within the optic nerves on coronal sections (not shown). No other lesions were seen in the brain. B, Axial T-2–weighted magnetic resonance image of the right hip, demonstrating susceptibility artifact of orthopedic hardware, as well as soft tissue heterotopic ossification (corroborated by plain-film radiographs), consistent with pseudotumor of metallosis with decompression into the right hip joint space (blue asterisk), the subiliac bursa (red asterisk), greater trochanter bursa (yellow asterisk) extending into the gluteus maximus muscle (arrowheads), and into a potential space between the tensor fascia lata and the rectus femoris (black asterisk).

A 63-year-old man with a history of well-controlled hypertension, lumbar stenosis, and hip arthroplasty with revisions presented with bilateral loss of vision and hearing for several weeks. He also reported a history of painful paresthesias at his feet beginning about 1 year prior.

On examination, his visual acuity was initially 20/80 OD and 20/200 OS; it deteriorated over the next 2 months to finger counting bilaterally. The Weber test gave a nonlateralizing result, and bone conduction hearing was absent bilaterally. There was length-dependent loss of vibratory sensation and absent reflexes at both lower extremities. Magnetic resonance imaging demonstrated abnormal enhancement of bilateral optic nerves (**Figure**, A). Optical coherence tomography and fluorescein angiogram were normal. Nerve conduction studies showed mild sensory axonal polyneuropathy. Laboratory tests were notable for elevated thyrotropin (63 mIU/ml), low levels of free thyroxine (T4) (0.2 ng/dL), and elevated levels of cerebrospinal fluid protein (94 mg/dL). Cerebrospinal fluid IgG index, oligoclonal bands, aquaporin 4 antibody, paraneoplastic panel, and protein electrophoresis test results were all normal. He was treated empirically with high-dose steroids and plasmapheresis, with no improvement in symptoms.

Four months after initial presentation, the patient reported new symptoms of chest tightness and dyspnea on exertion. An echocardiogram showed a left ventricle ejection fraction of 20% to 25%, which had been 55% to 60% at the last test 2 years prior. At 5 months after initial presentation, the patient developed acute worsening of dyspnea and was admitted for respiratory distress. He was found to have nonischemic cardiomyopathy and died on hospital day 13.

WHAT IS YOUR DIAGNOSIS?

A. Neuromyelitis optica (Devic disease)

B. Amyloidosis

C. Susac syndrome

D. Cobalt toxicity

Diagnosis

D. Cobalt toxicity

Discussion

The patient's prior hip replacement surgeries raised the possibility of cobalt toxicity as a cause of his constellation of symptoms. The patient had his first hip replacement surgery 7 years prior to presentation with a ceramic-on-ceramic implant and had required 2 revision surgeries. During the revisions, which occurred 2 years prior

to presentation, extensive soft-tissue metallosis was noted. Given this history of failed hip replacement, serum cobalt levels were checked and were consistently elevated, ranging between 280 to 779 ng/mL (a normal level is less than 1.5 ng/mL). A plain-film radiographic image of the hip showed that the hardware was intact, and there was heterotopic ossification in the surrounding soft tissue (not shown). Magnetic resonance imaging demonstrated a pseudotumor of metallosis (Figure, B). The patient was referred to orthopedic surgery for revision but died before surgical intervention.

The toxic effects of cobalt have been known since the 1960s, when the treatment of anemia with cobalt and the addition of cobalt to beer led to adverse effects.[1,2] In more recent years, joint replacements have become an increasingly recognized cause of systemic cobalt toxicity. Potential neurotoxic effects of cobalt include sensorineural hearing loss, optic neuropathy, and sensorimotor peripheral neuropathy.[3,4] Cognitive impairment has also been reported.[5] Other systemic effects of cobalt include cardiomyopathy and thyroid dysfunction.[6]

In this patient, bilateral vision loss with long segments of optic nerve enhancement immediately call to mind a demyelinating process, particularly neuromyelitis optica. The patient's male sex and the lack of oligoclonal bands could be consistent with a seronegative case of neuromyelitis optica; however, the concurrent hearing loss is atypical. Furthermore, many nondemyelinating disease processes can disrupt the blood-nerve barrier and lead to abnormal optic nerve enhancement. Amyloidosis can cause peripheral neuropathy and cardiomyopathy but do not explain the patient's presenting complaints of vision and hearing loss. Vision loss in Susac syndrome is because of occlusion of retinal arteries, and the syndrome is often associated with lesions in the corpus callosum. These findings were absent on fluorescein angiogram and magnetic resonance imaging. Cobalt toxicity is the unifying diagnosis that accounts for the patient's neurological decline, as well as his endocrine and cardiac dysfunction. Metallic debris from prior failed implants may have contributed to the metallosis.[7]

While cobalt toxicity is rare among the many patients who have benefited from joint replacement, there are an increasing number of people undergoing these procedures,[8] making cobalt toxicity a growing area of concern. Neurologists may be the first clinicians to evaluate a patient suffering from cobalt toxicity, given the frequency of neurologic symptoms.[4,5] The patient in this report had symptoms of peripheral neuropathy 1.5 years prior to his death from cardiomyopathy. If a patient with a history of a metal-containing implant develops symptoms that lead to concern of cobalt toxicity, serum cobalt levels and imaging of the joint should be obtained. If elevated cobalt levels and metallosis are discovered, surgical revision for source control should be considered urgently because it is considered the definitive treatment. Many of the effects of cobalt toxicity can be reversed with surgery. Conversely, failure to diagnose or treat expediently can have fatal consequences.[7,9,10]

ARTICLE INFORMATION

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