Ossification of the Posterior Longitudinal Ligament Misdiagnosed as Multiple Sclerosis

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Introduction

Ossification of the posterior longitudinal ligament (OPLL) has been recognized as "Japanese Disease". However, OPLL has been well-documented in the United States affecting non-Asian patients as well.[1-4] The prevalence among the African-American population is very low.[5] Magnetic resonance imaging noninvasively provides the most useful information about the degree and extent of spinal cord compression.[6]

Case Report

A 55-year-old African-American male was admitted to the hospital for left lower limb and right upper limb weakness. The patient had walking difficulty mainly due to the left foot drop. A CT scan of the brain showed an old one centimeter left peri-ventricular left infarct. An MRI of the brain showed advanced demyelinating plaques scattered in his brain.

The patient was started on pulse steroids for suspicion of multiple sclerosis (MS) and was discharged after partial relief of his symptoms. He was re-admitted, after being lost to follow-up, four months later for the same weaknesses described above in addition to poor balance and staggering gait. Again, the patient was started on pulse steroids for MS based on the findings of the first MRI of the brain, while the report of the new MRI was pending. With minimal resolution of his symptoms on steroids and the new brain MRI report of no active MS, an MRI of the cervical spine was obtained (Figure 1).

Neurosurgery confirmed the presence of OPLL along with complex osteophytes formation and probable congenital cervical canal narrowing. According to the neurological, clinical, and radiological findings, the patient had Grade IV myelopathy according to the Nurick Scale. The patient underwent an anterior cervical decompression. The patient's neurological symptoms were relieved dramatically in the 48 hours post-operation.

Figure 1. MRI of the cervical spine.

Discussion

Ossification of the cervical posterior longitudinal ligament (OPLL) represents a continuum of disease beginning with hypertrophy of the posterior longitudinal ligament (PLL) followed by progressive coalescence of centers of chondrification and ossification. Initial hypertrophy of the PLL due to fibroblastic hyperplasia is followed by increased collagen deposition and progressive mineralization and cartilaginous in-growths leading to ossification centers that eventually mature into Haversian canals.

A genetic locus for OPLL most likely is located close to the human leukocyte antigen (HLA) site on chromosome 6-p.[4,7] Patients with diffuse idiopathic skeletal hyperostosis (DISH), half of whom have OPLL, test positive for that HLA antigen.[8] An autosomal-dominant mode of OPLL inheritance frequently is inferred as one quarter of the siblings of OPLL patients manifesting the disease and demonstrating two concurrent strands of HLA.[9-11] The pathophysiological similarity between DISH and OPLL and the fact that both are linked to HLA antigen make this antigen an area of intense investigation.

One quarter of North American and Japanese patients with cervical myelopathy exhibit OPLL.[12-15] OPLL is found in C2–4 70% of the time, 15% in T1–4, and 15% in L1–3. Neural injury occurring in the presence of OPLL stems from direct mechanical or indirect ischemic compromise. Cervical OPLL also appears twice as often in males as in females.[12,16-18]

OPLL in the African-American population has scarce data on its true incidence in that population.[19] Even in DISH, the more diffuse variant, the demographics in the United States showed a much higher incidence in the white rather than the African-American or Native American population.[20]

Two myelopathy grading scales are used: the Nurick Scale and the Japanese Orthopaedic Association (JOA) Scale. The Nurick Scale includes:

- Grade 0: intact, mild radiculopathy without myelopathy
- Grade I: mild myelopathy
- Grade II: mild-to moderate myelopathy
- Grade III: moderate myelopathy
- Grade IV: moderate to severe myelopathy
- Grade V: severe myelopathy, quadriplegia.[13,14,16,21-23]

The JOA Scale catalogues the severity of myelopathy by using a 17-point scale.[24-26]

Direct anterior resection of OPLL results in improved postoperative outcomes. In the study by Fessler, et al.,[27] Nurick grades improved 86% of the time following anterior surgical approaches. These patients improved an average of 1.24 Nurick grades, whereas those undergoing laminectomy improved only 0.07 Nurick grades. Better clinical outcomes are encountered following anterior resection rather than posterior decompression of OPLL.[12,14,19,28] This approach was adopted in our patient with satisfactory results.

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