

Case Report

Spastic Paraparesis in a Middle Aged Female Unmasked a Rare Cause of Myelopathy: Ossification of the Posterior Longitudinal Ligament (OPLL) - A Case Report

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Abstract

Ossification of the posterior longitudinal ligament (OPLL) is a rare but potentially devastating clinical entity of degenerative myelopathy. Highest reported prevalence of OPLL is in East Asian countries. The cervical spine is more commonly affected followed by thoracic and lumbar spine. The clinical manifestations range from asymptomatic to myelopathy or myeloradiculopathy. Although rare, thoracic OPLL is more severe than cervical OPLL and usually requires operative intervention. CT scan is the investigation of choice to determine the extent and thickness of the ossification. Treatment ranges from observation in patients with minimal symptoms to surgical decompression followed by stabilization for patients with myelopathy. Here we discuss a middle-aged lady who presenting with a slowly progressive spastic paraplegia with bladder dysfunction, without sensory involvement. She was being treated conservatively as a case of spondylotic degenerative disease, but showed no improvement. Finally proper evaluation of her radiologic finding and clinical correlation revealed the diagnosis of OPLL. She underwent laminoplasty with decompressive surgery. The outcome was proved to be curative.

Key words: ossification of posterior longitudinal ligament (OPLL), myelopathy, laminoplasty

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Introduction

The ossification of the posterior longitudinal ligament (OPLL) of the cervical spine is an inflammatory process that results in the replacement of the posterior longitudinal ligament (PLL) with lamellar bone. OPLL causes spinal canal stenosis, which leads to different degrees of neurological manifestations.¹ OPLL can involve any region of the spine; with the cervical spine being the most commonly affected level followed by the thoracic and lumbar spine. OPLL was first described in the 1960s, is also known as “Japanese disease” as it was first noticed in the Japanese population.² The epidemiology of OPLL has been the subject

of many studies. Initially, it was believed to be endemic to the Asian population. The prevalence of OPLL is reported to be as high as 3.0% in Asians, and ranges from 0.1% to 1.7% in North Americans and Europeans.^{3,4} The exact aetiopathogenesis of OPLL is not entirely understood to date. A broad spectrum of metabolic, genetic, and environmental factor has been studied and implicated for its development and progression. This case report describes a 40-year-old lady with spastic paraparesis, ultimately diagnosed as a case of thoracic OPLL which is less common presentation of this rare disease.

Case summary

Mrs. A, 40-year-old, married, Muslim, homemaker hailing from Pabna with no known co-morbidities got admitted in PMCH with the complaints of weakness of both lower limbs for last 1 year. Weakness was acute in onset which she first noticed while trying to get out of bed in morning and was slowly progressive over the year. She needed support to walk after taking few steps. She also complained of mild tingling sensation in both lower limbs but no objective sensation abnormality. Her upper limb was functioning normally and she could maintain her upright position. For

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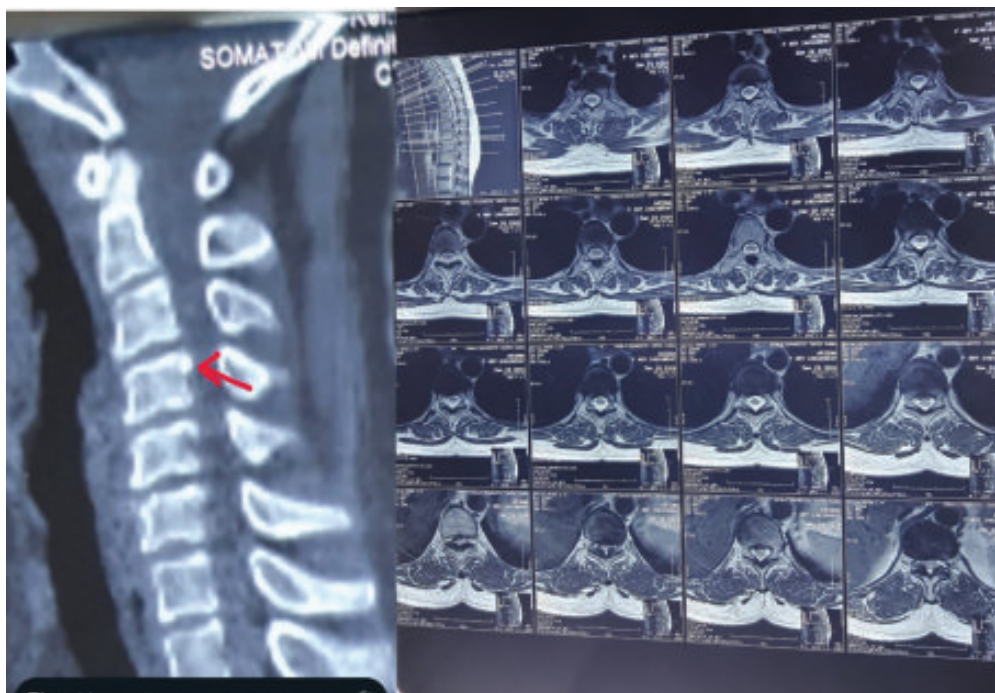
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last few months she is suffering from urinary incontinence, not accompanied by any burning, urgency, hesitancy or change in urinary color or volume. She denied any recent or remote history of trauma, low back pain or any other bony pain. She has no history of fever, weight loss, respiratory tract infection, diarrhoea, previous weakness of any limb, visual disturbance or any surgical intervention. On general examination, her vitals were within normal limit. There was no anaemia, no bony tenderness or any gibbus. Motor system examination of lower limb showed, normal muscle bulk, increased tone on both sides. Muscle power was 3/5 on left side, 4/5 on right side both proximal and distally. Ankle clonus was present, knee & ankle jerks were exaggerated, both plantar responses were extensor, coordination was intact. Motor examination of upper limbs revealed no neurological deficit. All modalities of sensation were intact in all four limbs. Other system examination revealed no abnormality. We considered the possibilities of Spastic paraparesis due to degenerative disease, neoplasia, multiple sclerosis & Pott's disease. On investigation, CBC revealed mild neutrophilic leukocytosis with WBC count 11.29 k/mcl with neutrophil 66% and normal RBC and Platelet count. Urine RME revealed pus cells: 6-8/HPF, Creatinine 0.66 mg/dL. X-Ray of Chest P/A view revealed normal finding and X-ray of dorsolumbar spine (B/V) revealed degenerative changes in D6 to L5 level. CT scan was done initially and later MRI of Thoracic Spine with survey of whole spine

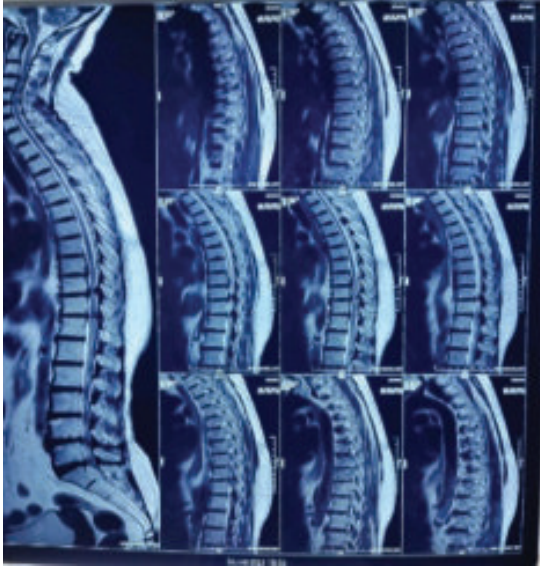
revealed: focal ossification of posterior longitudinal ligament at D9-D11 level with ossification of ligamentum flavum at D10 and D11 level causing severe spinal canal stenosis and compressive myelopathy. There was also disc bulge at C3-C4, C4-C5, C5-C6, L4-L5 and L5-S1. Our final diagnosis was dorsal myelopathy due to OPLL at D9-D11 and ossified ligamentum flavum causing spastic paraparesis.



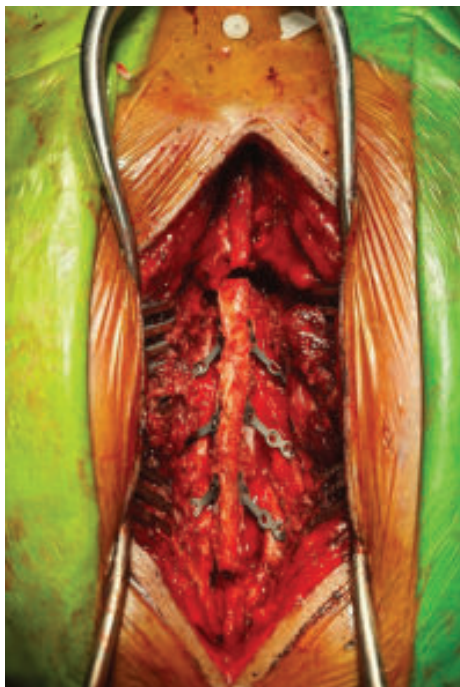
X-ray of dorsolumbar spine (B/V): Degenerative changes in D6 to L5



CT scan: segmental OPLL at C4, OPLL at thoracic spine



MRI of Thoracic Spine with survey of whole spine revealed: focal ossification of posterior longitudinal ligament at D9-D11 level with ossification of ligamentum flavum at D10 and D11 level causing severe spinal canal stenosis and compressive myelopathy. Osteo disc bulge at C3-C4, C4-C5, C5-C6, L4-L5 and L5-S1.



Perioperative view of laminoplasty

After neuro surgical consultation patient underwent laminoplasty with removal of ligamentum flavum and decompression. After surgery patient was able to walk by herself without residual weakness or disability.

Discussion

The posterior longitudinal ligament is essential in the stabilization of the spinal column. When it develops calcifications, it becomes thickened and can lead to spinal cord compression. OPLL is twice as common in men compared with women.⁵ Genetic risk factors contribute significantly to the development of OPLL. In one study of 347 families, OPLL was incidentally discovered in up to 30% of affected patients' siblings. Various genomic studies showed that multiple genetic variations are associated with the occurrence and severity of OPLL; it includes BMP9 and BMP4 haplotypes, COL6A1, and TGF3 gene.⁶ So far, the published literature suggests an association of OPLL with DISH (Diffuse idiopathic skeletal hyperostosis), ankylosing spondylitis and, diabetes mellitus.⁷ OPLL can affect any spinal segment but is most frequently observed in the cervical spine. The frequency is cervical 75%, thoracic 15%, lumbar 10%.⁸ Reasons for this are not entirely clear, however may be related to the narrower caliber of the cervical spinal canal with the increased motion that may predispose this segment to symptom development. Thoracic OPLL is rare, and symptoms of myelopathy are more severe than in cervical OPLL due to the narrow canal, rigidity of the thoracic spine, tenuous blood supply, and inability of the spinal cord to withstand much compression. Surgical intervention is usually indicated due to the severity of clinical presentation.⁹ The most commonly used classification for OPLL is that proposed by the Investigation Committee for Ossification of the Spinal Ligaments, part of the Japanese Ministry of Health, Labor and Welfare-¹⁰

1. continuous: a long lesion extending over several vertebral bodies.
2. segmental: one or several separate lesions behind the vertebral bodies.
3. mixed: a combination of continuous and segmental types.
4. localized/focal/circumscribed: mainly located posterior to a disc space.

The incidence of concomitant OPLL in other, remote spinal levels, is high; one study noted that 56.2% of patients with cervical OPLL also had evidence of disease in the thoracolumbar spine.¹¹ If present, symptoms usually manifest in the 4th-6th decades of life.¹² Although 5% of diagnosed patients are asymptomatic, varying degrees of neurologic symptoms can be present including both radiculopathy and myelopathy.¹³ OPLL has a long course of presentation with variable degrees of neurological involvement. This makes it challenging to lucidly understand the natural course of the disease. Traditionally, plain radiographs were used to diagnose and classify OPLL. Unlike the thoracic vertebral segments, cervical spine is not overlapped by shoulders and ribs. Therefore, cervical OPLL are relatively more easily identified on plain radiographs than their thoracic counterparts. Computed tomography (CT) is diagnostically

more accurate in identifying ossified spinal ligaments, especially in the thoracic spine and junctional regions.¹⁴ Surgical intervention has consistently been shown to significantly improve clinical outcomes for most patients with moderate to severe, or progressive, myelopathy attributable to OPLL.¹⁵ Surgical intervention can be undertaken from an anterior, posterior, or combined approach. Posterior approaches, including laminectomy and fusion (LF) and laminoplasty (LAMP), can be used to treat a wide range of OPLL disease severities making it the most popular approach to surgical OPLL correction worldwide.¹⁶ The degree of clinical improvement achieved with surgery has been found to be similar to the surgical benefits achieved with decompression of other forms of degenerative cervical myelopathy.¹⁷ The degree of benefit achieved with surgery is significantly determined by the degree of myelopathy at presentation. Our patient was a female of forty who presented with very slowly progressive spastic paraparesis with bladder involvement over the year. She consulted several physicians, went through multiple radiological investigations including CT, MRI without any conclusive diagnosis and was being treated conservatively as a case of spondylotic degenerative disease. As patient had a long course without any systemic symptom and pain we considered infection like TB as very remote possibility. Absence of sensory involvement excluded inflammatory conditions like Transverse Myelitis. Her features were not consistent with usual presentation of multiple sclerosis Degenerative disease, slowly progressive neoplasm were considered. Radiology excluded neoplasm and findings were not fully consistent with spondylosis. Reevaluation of radiological features ultimately revealed diagnosis of OPLL. Our patient presented with early age without any association or risk factor of disease. Moreover she had mainly dorsal OPLL and only one segmental involvement of cervical spine. Considering whole scenario of presentation though it was uncommon presentation of a rare disease, surgical approach on time resulted in an excellent outcome with recovery.

Conclusion

OPLL is a disease that is a relatively uncommon, but nonetheless important cause of cervical myelopathy and sometimes dorsal myelopathy. Untreated, it may lead to neurologic morbidity and ultimately significant loss of independence. Unfortunately, due to the rarity of OPLL as degenerative myelopathy, it is rarely named on imaging, lumping its radiographic findings under the umbrella of degenerative changes. Compared to nerve specific radiculopathy, the widespread and vague symptoms of myelopathy are not often referred to surgical consultation. However, OPLL should be considered when extensive

degenerative changes are noted on imaging. Though dorsal OPPL is rare, it is essential to make a distinction between OPLL and spondylosis because early recognition of OPLL and surgical intervention can result in cure unlike spondylosis.

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