

# TETRAPARESIS SECONDARY TO CERVICAL OSSIFICATION OF THE POSTERIOR LONGITUDINAL LIGAMENT

## Case report

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**ABSTRACT** - Ossification of the posterior longitudinal ligament (OPLL) is a rare cause of myelopathy in non-Oriental populations and relatively unrecognized by general practitioners. A case of an Afro-Brazilian 54-years-old woman presenting with tetraparesis due to cervical OPLL is presented. Emphasis is made for the inclusion of OPLL in the differential diagnosis of compressive cervical myelopathy.

**KEY WORDS:** ossification of the posterior longitudinal ligament, spinal cord compression, spinal stenosis.

### **Tetraparesia secundária a ossificação do ligamento longitudinal posterior cervical: relato de caso**

**RESUMO** - Ossificação do ligamento longitudinal posterior (OLLP) é causa rara de mielopatia na população não Oriental e relativamente subdiagnosticada por clínicos gerais. Relata-se um caso de mulher de 54 anos descendência Afro-Brasileira com tetraparesia associada a OLLP cervical. Enfatiza-se a inclusão da OLLP como diagnóstico diferencial de mielopatia cervical compressiva.

**PALAVRAS-CHAVE:** ossificação do ligamento longitudinal posterior, compressão da medula espinhal, estenose espinhal.

Ossification of the posterior longitudinal ligament (OPLL) has been a relatively common cause for myelopathy in Korea and Japan since its first description by Tsukimoto in 1960<sup>1,2</sup>. OPLL occurs in perhaps only 0.2% of the Caucasian population<sup>3</sup>, whereas its prevalence in Japan has been reported as of 1.5-2.4% in adults<sup>4</sup>. The rarity of the condition in non-Oriental populations often leads to delay and/or failure of recognition.

We report an OPLL in a 54-year-old Afro-Brazilian woman who had tetraparesis and a diagnosis based on clinical and radiological investigation. The aim of this report is to discuss the relevant aspects of OPLL, mainly its clinical presentation and diagnosis.

### **CASE**

A 54-year-old hypertensive woman was hospitalized (HU-UFJF-SAME 347836) with approximately four months of gait impairment. On orthopedical evaluation, spastic tetraparesis and trunk instability were observed. In addition to this, on neurologic consultation, increased deep tendon reflexes of both upper and lower extremities were detected. There were no abnormalities on upper-and-lower-limb cerebellar and sensitive examinations. Nor family background or spondylosis, diabetes, trauma, irradiation history were positive.

Radiographs revealed a hyperdense image at the cervical vertebral canal (Fig 1). A computed tomogram (CT) showed a mass of ossification compressing fifty percent of the spinal cord in the posterior aspect, from the 3<sup>rd</sup> to the

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7<sup>th</sup> cervical vertebral bodies. There were neither signs of diffuse idiopathic skeletal hyperostosis (DISH) nor ankylosing spondylitis. Cervical magnetic resonance imaging (MRI) confirmed the CT findings and defined surgery as the main treatment alternative (Fig 2).

A neurosurgical intervention was indicated due to a progressive clinical picture. This case report was performed with the approval of the Ethics Committee of the university hospital (CAAE 0102.0.180.000-06).

## DISCUSSION

OPLL is defined as an abnormal thickening and new bone development of the PLL, causing significant cord compression<sup>4,5</sup>. Ninety-five percent of cases occur within the cervical spine<sup>4,5</sup>, involving 2.7 to 4.0 levels in 70% of cases<sup>6</sup>. Up to 25% of patients presenting with cervical myelopathy have OPLL rather than spondylotic and stenotic myelopathy or disc disease alone<sup>6</sup>. According to Endo et al.<sup>7</sup>, 15 (6.5%) of 231 cervical spinal injuries were associated with cervical OPLL. However, the incidence of OPLL was estimated to be 26 to 38% in subjects sustaining acute cervical cord injury without fracture or dislocation of the spinal column as pointed out by Portha et al.<sup>8</sup> and Katoh et al.<sup>9</sup>.

Although many possible causative factors have been given, such as gender, generalized tendency towards calcification, spondylosis, diabetes, trauma, irradiation, hormonal imbalance and dietary habits, the cause of the disease remains obscure<sup>8-12</sup>. It has been suggested that the increase in prostaglandin I<sub>2</sub> synthase induced by mechanical stress plays a key role in the progression of OPLL, at least in part through the induction of osteogenic differentiation in spinal ligament cells<sup>13</sup>. There are family background as well as strong association with diabetes mellitus, impaired

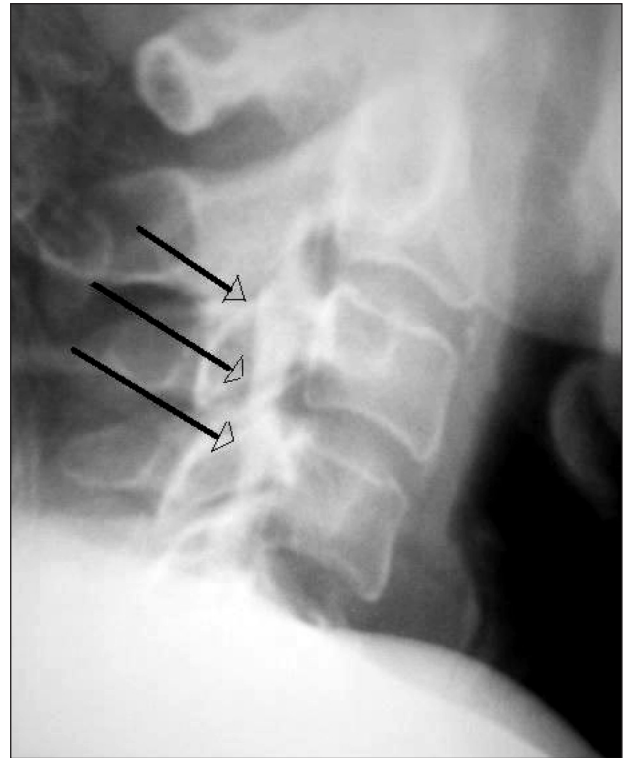


Fig 1. Radiography showing an hiperdense image at the posterior aspect of the 3<sup>rd</sup> to the 7<sup>th</sup> cervical vertebral bodies (long arrow).

glucose tolerance and DISH<sup>9,14</sup>. In white populations, OPLL is closely associated with DISH and it is reported in 43-50% of individuals with DISH<sup>5</sup>.

Patients may develop OPLL in concomitance to ossification of the anterior longitudinal ligament, thoracolumbar OPLL and calcification of the ligamentum flavum in, respectively, 62%, 37% and 37% of cases<sup>15</sup>.

Based on CT scans of the cervical spine performed in whites with cervical myelopathy, Epstein<sup>16</sup> has pro-

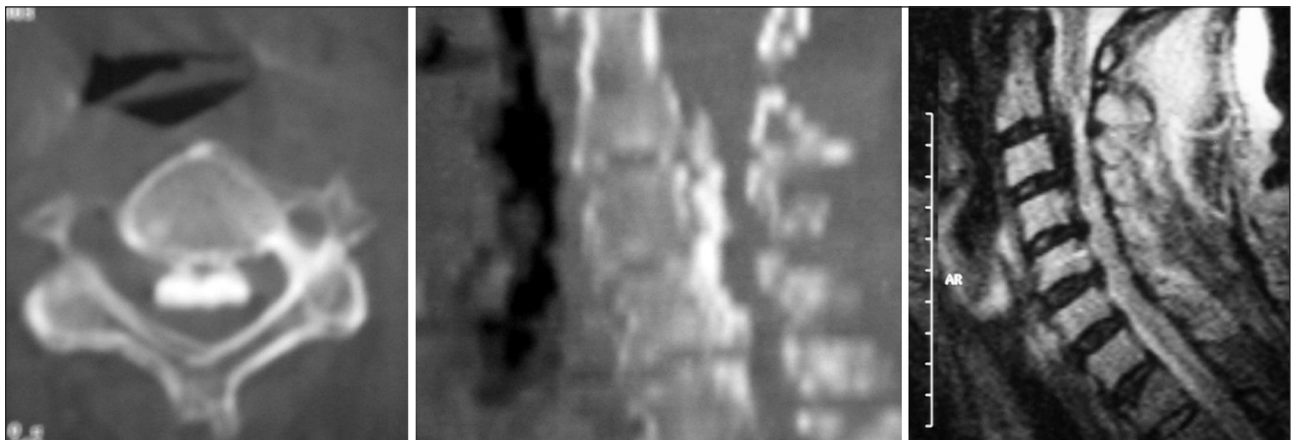


Fig 2. Cervical CT and MRI scans showing spinal cord compression due to OPLL.

posed a new entity, OPLL in evolution. She noted relatively frequent hypertrophy of the PLL with punctuate calcifications in the ligament substance. Kondo et al.<sup>17</sup> support the hypothesis that hypertrophy of the PLL is a prodrom condition to OPLL. This may compress the spinal cord or nerve roots. Motegi et al.<sup>18</sup> reported that hypertrophy of the PLL is often considered an early stage of OPLL because of the apparent clinic-pathological similarities.

The disorder is often asymptomatic<sup>16</sup>. It is known that the presence of prominent OPLL does not always indicate the presence of cervical myelopathy<sup>19</sup>. The clinical symptoms of OPLL appear mostly above 40 years of age. Patients complain of pain in the neck and in the occipital area, and this is followed by paresthesia and weakness in the upper and lower extremities. Paresthesia is a very common symptom and it is found in up to 70% of patients. Patients with OPLL sometimes present with acute cervical cord injury after only minor trauma, such as a fall to the ground<sup>19</sup>.

The CT appearance of OPLL, with ossified material corresponding to the levels of the vertebral bodies as well as the discs, helps to distinguish it from spondylosis<sup>18</sup>. The calcification may be continuous or segmental, the latter making the condition more difficult to diagnose. On T1 and T2-weighted MRI, a continuous low signal band is shown between the vertebral body and theca. Bone marrow is presented as an increased or intermediate signal within the ossification in over 50% of cases<sup>19</sup>. The ossification is best recognized on T2-weighted sequences because of the contrast with the high signal cerebrospinal fluid. The low signal corresponding to the ossified ligament on MRI is not entirely specific. According to Koyanagi et al.<sup>19</sup>, MRI is useful to understand the level and mechanism of injury, as well as the true extent in height of the ossified ligament.

It is essential to make a distinction between OPLL and spondylosis because failure to recognize OPLL resulting in inappropriate surgery can exacerbate the situation.

Conservative treatment is indicated for patients with mild paresthesia and no evidence of muscular weakness. Treatment options include the use of non-steroidal or steroidal agents and occasional immobilization in cervical orthoses<sup>6</sup>.

Early decompressive surgery should be recommended to patients with progressive neurological deficits in spite of conservative treatment or when there is obvious compression of the spinal cord. Op-

erative alternatives include anterior surgery alone, anterior surgery combined with posterior stabilization or posterior procedures comprised of laminectomy with or without fusion, or laminoplasty. Surgical alternatives are based on the severity of myelopathy, the presence or absence of lordosis or kyphosis and the coexistence of disc disease, congenital stenosis and spondylosis along with the recognized OPLL<sup>6</sup>. Kamizono et al.<sup>20</sup> pointed out that surgical treatment (open-door type laminoplasty) of OPLL successfully contributed to patients returning to their occupations and productive activity. A total of 160 (53%) of 301 patients were able to return to work.

This case vignette is interesting because the patient was a 54-year-old Afro-Brazilian woman who had presented with OPLL manifested by tetraparesis and trunk instability without any sensitivity deficits. OPLL had involved 5 levels of the cervical spine, which is rather rare (30% of cases), and it had not been associated to ossification of the anterior longitudinal ligament, thoracolumbar OPLL and calcification of the ligamentum flavum. Etiology remained unknown since the patient had presented with no co-morbidities or risk factors. She had neither family background, nor diabetes mellitus, impaired glucose tolerance and DISH. Clinical picture of tetraparesis and gait impairment, associated with initial cervical radiogram findings were suggestive of OPLL. CT and MRI further investigations confirmed the suspicion, demonstrating the lesion extension and compression of the cervical spinal cord which is described in 25% of cases<sup>6</sup>. In our patient, conservative and surgical treatments were impossible due to, respectively, her progressively deteriorating clinical picture and pulmonary embolism with subsequent death before the surgical procedure. Our interest is to emphasize the inclusion of OPLL in the differential diagnosis of cervical myelopathy.

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