Isolated Proximal Thoracic Ossified Ligamentum Flavum Causing Paraparesis in a Caucasian: A Case Report and Review of the Literature

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ABSTRACT
Ossified ligamentum flavum or OLF is a well-known pathology in East Asian countries. Since the first description of the disease by Adams and Davis in a Caucasian, ossified ligamentum flavum is being gradually accepted as a rare but an important cause of thoracic myelopathy among this ethnic group. Ossification of the ligamentum flavum mostly occurs at the lower thoracic segments in multilevel fashion and involvement of the high thoracic region is seen less frequently. A survey of the literature disclosed only three Caucasians with multilevel OLF at proximal thoracic region, and not a single case of isolated type at this region could be found. Herein, a 50-year-old Caucasian female with paraparesis due to proximal thoracic OLF is presented, which is the first example in this ethnic group.

KEY WORDS: Ligamentum flavum, Myelopathy, Ossification, Thoracic spine

INTRODUCTION
Ossification of the ligamentum flavum is a pathologic condition that affects the yellow ligaments mostly in male adults between fifty to sixty years of age. Ossification is more common in the thoracic spine causing slowly progressive myeloradiculopathy. In fact, OLF is so common in East Asian countries that is regarded as a frequent aging pathology in the people of this region (3,10-15,21,22,31,32,36,40,41). However, affection of other ethnic groups is regarded a rare event as it is in Caucasians (1,4,6,8,9,14,23,25,28,30,34,37-39). In the thoracic region, this disorder has a strong predilection for lower segments and is mostly seen in a multilevel form. However, the isolated single type is rare specially at the upper thoracic region even in East Asian reports, meaning that occurrence of isolated OLF at the proximal thoracic area should be regarded as a medical curiosity among the Caucasians. Previously, there have been only three documented case reports in Caucasian with multilevel OLF at this location. Herein, the first Caucasian with an isolated lesion at the proximal thoracic region, who was admitted with severe paraparesis and had a subsequent good outcome after decompressive laminectomy, will be presented.

CASE REPORT
This 50-year-old female was admitted because of difficulty in walking in the last six months. Her neurological exam revealed spastic paraparesis with a sensory level at T4 and position sense impairment with a thoracic JOA score of 5/5.

MRI revealed a hypointense V-Shape, interlaminar mass at T3-T4 level causing severe impingement on the thecal sac and the spinal cord, both in T1 and T2 images (Figures 1, 2). Myelomalacia was not detected. MRI of the rest of the spine was clear from any similar lesion. On CT, a dense triangular mass at the same level corresponding to the hypointense region on MR images was demonstrated (Figure 3). Diagnosis of ossified ligamentum flavum at the T3-T4 interval was made and surgical intervention was decided on. First, partial laminectomy of the cranial
part of T3 and then caudal part of T4 was done in order to expose the intact dura as a guide. Later the remaining parts of the laminas were removed with a high-speed burr. With alternate detaching the ossified OLF from the dura and its thinning with diamond burr, the ossified mass was detached and removed. Reappearance of normal pulsation of the cord pointed to the efficacy of decompression. Postoperatively the patient showed marked recovery. Three months after surgery, thoracic JOA score reached 10 with recovery rate of 86%. The patient's JOA score in the last visit after a year showed further improvement with a JOA score of 11 and recovery rate of 100%.

**DISCUSSION**

Ossification of the yellow ligament with resultant fusion of two adjacent laminas is called OLF or ossified ligamentum flavum, OYL or ossified yellow ligament. It extends from the ventral aspect of upper lamina to the dorsum of lower lamina (3,10-12,36).

OLF is a well-known cause of progressive cord and cauda equina neurology and is reported mainly in East Asian countries. OLF is so common in Japanese people older than 65 years of age that it is regarded as a common feature of aging spine in Japan (3,13,21,31,36,41). This predominance is not confined to these countries and symptomatic cases of OLF have been documented in

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**Figure 1:** T1-weighted MR image showing hyposignal triangular mass at T2-T3, compatible with ossified bone.

**Figure 2A,B:** T2-weighted MR image, demonstrating hyposignal triangular mass with cord compression.

**Figure 3:** Reconstructed CT showing marked canal compromise by a beak-type ossification. Note that the ossifications never unite.
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other ethnic groups. A few series from India, the Middle East, North Africa, and Caribbean with sporadic cases in Caucasians have been reported (2,5,26,27,29,35). Xu et al could find only 22 Caucasians with thoracic OLF reported in the past in a review of the literature in 2008 (1,4,6,8,9,14,23,25,28,30,34,37,38,39). All except one were located in the lower segments. 18 out of these 22 were multilevel and the remaining four were isolated, all in the lower thoracic region, indicating the rarity of isolated upper thoracic OLF in their review.

The etiology of OLF remains uncertain. Both systemic and local mechanisms are hypothesized (8,20,24,36). Currently a couple of factors including mechanical, traumatic, metabolic, environmental, chronic degenerative, biological and hereditary are proposed (18,24,33,36). The higher prevalence in East Asian countries, particularly in Japan, China and Korea implicates genetic and environmental factors (3,10-12). Recent studies revealed that fibronectin, bone morphogenic proteins and transforming growth factor beta play an important role in the development of ossification of the spinal ligaments (20). Patients with OLF have higher frequency of non-insulin dependent diabetes, obesity, hyperinsulinism, haemochromatosis and calcium-phosphate metabolism abnormalities (1,20,36,38).

With respect to high prevalence of thoracic OLF compared to other regions of the spine, Otani et al attribute this higher incidence of ossification of thoracic spine to thoracic hyperkyphosis and suggest that the altered mechanical stress and its consequence predispose the thoracic spine to the development of ossification of the yellow ligaments (24). Magnie et al believe that higher prevalence of OLF at the thoracolumbar region to be due to unique orientation of facet joints and their relative hypermobility (18).

Affection of the thoracic spine might be isolated and confined to one segment or it can more commonly affect multiple levels (30). In the multilevel or multifocal type, the affection might occur in the adjacent segments which is called the continuous form or the affection can appear in a random alternate pattern called the non-continuous form (7,10,21,32).

Thoracic OLF is generally demonstrated in the lower thoracic region mostly at Th9 thorough Th12 (15). Affection of higher levels especially at the proximal third of the thoracic spine are notably less seen in all ethnic groups.

A comparison of the different types of OLF affecting the proximal thoracic spine reveals that the isolated type is apparently less commonly encountered than the multilevel type among East Asian people (11-13,15,30). Far more rarely is OLF of this special location among the Caucasians and after its first description by Van Oostenburgge, only two more cases that were of multilevel type have been published so far in the English literature (14,34,37). The present case is therefore the first isolated proximal thoracic OLF in a Caucasian in the literature.

OLF of the thoracic spine is predominantly seen in the middle-aged subjects with the mean age at the time of surgery ranging from 50 to 60 years according to different series (3,10-13,21,31,41). Males outnumber females with a male to female ratio ranging from 2:1 to 4:2 (10, 41).

The diagnosis of this disorder is usually delayed due to its insidious onset and very slow progression. However, once the condition becomes symptomatic, its major clinical picture is characterized by progressive myelopathy in more than 95% of subjects. In such instances, the cardinal symptoms are altered gait, ataxia, weakness as well as numbness of the lower limbs, presented in neurological exam as spastic paraparesis with hyperreflexia, clonus, up-going planters. Sensory deficits are mainly superficial, deep and proprioceptive impairment (3,4,10,29,31).

Radiculopathy that indicates outgrowth and lateral extension of the lesion toward the foramen is the least common symptom seen in only 4 % of the cases. Sphincter

<table>
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disturbances presenting with voiding difficulty is seen in about one fourth to one third of the patients (11,12,31). Neurological assessment of the patients affected by thoracic OLF can be best achieved through application of modified thoracic JOA scoring (21).

On some occasions thoracic spine traumas of moderate intensity in OLF patients with an already compromised thoracic spinal cord can result in catastrophic central cord syndrome that has a wide spectrum from dysesthetic pain of the lower extremities to severe neural impairment and even paraplegia.

In classical OLF of the thoracic spine, the interval between the appearance of the first symptoms to diagnosis is quite variable and is usually from several months to a few years (3,11,12,31). Undoubtedly, the duration depends on the individual canal size, association of thoracic kyphosis, the number of the ossified levels, type of OLF and preexisting OPLL. Association of OPLL with thoracic OLF is infrequent but will apparently contribute to myelopathy (12, 15).

The combination of MRI and CT is required for the precise diagnosis of OLF (7,15,16,19,21). MRI accurately delineates the location and extent of spinal cord compression in several planes. OLF shows itself as a hypointense triangular or round or beak-like mass both in T1 and T2 images compressing the cord from the posterior aspect in sagittal plane (7,21,32). The appearance of myelomalacia which express itself as a hyperintense intermedullary signals in T2 images correlates with the degree and duration of cord compression although it has a controversial predictive value in assessing the outcome after surgery. Moreover, MRI is the best diagnostic tool to detect coexisting lesions such as OPLL, intervertebral disc herniation and congenital narrow canal (7,10,21,24,32).

Once the diagnosis of OLF is established on MRI, a CT Scan is necessary for the assessment of the extent of the ossification in the transverse and sagittal planes. With regard to CT features, different classifications of OLF have been proposed. One of these has classified OLF as unilateral, bilateral and bridged types (15). According to another classification, five types including, lateral, extended, enlarged, fused and tuberous exist (21). Lateral, diffuse and nodular are other valid subtypes described with regard to axial CT images (16). However, the cranial and caudal parts of OLF never unite even in the most thickened ones. Muthukumar has noticed comma-shape holes in CT slices of OLF indicating dural affection between the layers of ossification. He believes that it has a predictive value with possibility of dural tear during surgery (19).

Conservative treatment is not expected to improve thoracic myelopathy, thus surgery remains a solution. The surgical procedure most commonly performed for decompression is laminectomy (12). However, special care should be used in surgery in order to avoid dural tear. Dural tears are inevitable in more than 30% of bridged, nodular and tuberous forms (16,17,21).

Recently en block laminectomy as well as laminoplasty have been proposed and employed as an alternative for the treatment of OLF. However, because of limited decompression afforded by laminoplasty and with regard to some evidences and reports of deterioration of the neurological status by this method, laminectomy has remained the most acceptable surgical mode in OLF patients with the most satisfactory results (19,40). Instrumentation subsequent to laminectomy has been advocated by a few authors for prevention as well as management of kyphosis (39).

Histological examination of OLF typically shows mature lamellar bone associated with endochondral ossification of ligamentum flavum that begins near the facet joint. The proliferation of cartilaginous tissue triggers the ossification. Inside the lamellar bone, there are poorly developed bone marrow civilities.

Postoperative recovery should be expected with excellent outcome if early surgical decompression is performed. Existence of preoperative neurological symptoms over a long period presenting with OLF-induced myelomalacia may result in incomplete recovery and even poor prognosis (13,21,31).

Affection with more than two interlaminar segments, old age and coexistence of OPLL have worse outcome than a single segment OLF without coexisting pathologies in a younger subject (3,10-13,21,31). Notably, cord compression by OYL in the lower and middle third of the thoracic spine has a better prognosis than proximal third lesions (13). Among these, the duration of the symptoms before surgery is the most important variable (21). Postoperative degree of recovery can be easily estimated and graded with application of recovery rate formula.
based on preoperative and postoperative thoracic JOA scores (21).

Regular follow-up is necessary as recurrence at the same level or development of ossification at other levels has been reported.

In conclusion, symptomatic spinal cord compression secondary to isolated proximal thoracic OLF is rare but it should be included in the differential diagnosis of thoracic myelopathy in all ethnic groups. Characteristic features of MRI can provide sufficient clue for proper diagnosis and surgical planning. Whole spine MR is advocated for demonstration of similar pathology in the other regions of the spine as well as coexisting ossified lesions such as OPLL. Prompt decompressive surgery should be planned once the diagnosis is made. Early removal of the offending ossifying lesion is known to result in resolution of the deficits and good outcome.

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