Thoracic Myelopathy Secondary to Ligamentum Flavum Ossification

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Abstract

Introduction: Focal ossification of the ligamentum flavum (OLF) is a rare cause of thoracic myelopathy. The lower thoracic spine is most frequently affected and the patients present initially with posterior column disturbances followed by progressively increasing spastic paraparesis. The pathogenesis of OLF has not been conclusively established. Materials and Methods: Five patients with thoracic myelopathy due to OLF underwent decompressive laminectomy and excision of the ligamentum flavum. Magnetic resonance (MR) images consistently revealed a linear or beak-like excrescence, uniformly hypodense on T1- and T2-weighted images, situated posterior to the thecal sac. A comparison of the preoperative neurological status and at follow-up was done using the Japanese Orthopaedic Association and Nurick scores modified for thoracic myelopathy. Results: Decompressive laminectomy with excision of the OLF resulted in significant improvement in motor weakness and gait in 4 (1 excellent, 2 good, and 1 fair) patients who had slow-onset, but progressive compression, OLF. The patient who had acute traumatic spinal injury did not recover despite decompression and rehabilitation. All improved in their gait and spasticity, but 4 patients had persistent sensory deficit. Conclusion: OLF can significantly contribute to a spatial reduction of the thoracic spinal canal, resulting in slowly progressive paraparesis or acute paraplegia after trauma to the back. The T2-weighted sagittal image of MR imaging is the modality of choice for screening the longitudinal extent of OLF, with increased diagnostic accuracy when combined with computed tomographic myelogram. Neurological improvement usually occurs following decompression laminectomy with or without excision of the OLF. However, the persistence of residual numbness or weakness at follow-up may be due to irreversible changes within the cord as a result of severe thecal compression and the delay between the onset of initial symptoms/signs and surgical decompression. Prognosis remains poor for acute myelopathy with pre-existing OLF, despite surgery.

Key words: Compressive myelopathy, Decompression laminectomy, Ligament ossification, Spinal cord compression, Thoracic radiography
8 patients who underwent surgery were found to have an associated OLF. Although the incidence of myelopathy resulting from spinal cord compression by OLF is not known, it is regarded as less common than OPLL; the latter is reported to occur in 19 of 1 million Asians.4 However, the precise pathoanatomy of OLF has only recently begun to be understood. We describe 5 patients with thoracic myelopathy caused by OLF and discussed their clinical manifestations, radiographic findings, pathology and postoperative outcome.

Materials and Methods

We examined 5 Chinese (1 male and 4 females) patients who had thoracic myelopathy caused by OLF-related spinal canal stenosis and who underwent decompression laminectomy. Their mean age was 56 years (range, 45 to 65 years). The duration of follow-up was 1 year in 2. The excised ligamentum flavum in all the patients consistently revealed foci of dystrophic calcification and chondro-osseous metaplasia, all characteristics of OLF.

The motor power and functional disability of all patients were assessed pre- and postoperatively by the Medical Research Council’s (MRC) motor power grading system and the Japanese Orthopaedic Association’s (JOA) scoring system for cervical myelopathy, which was modified for thoracic myelopathy by excluding the motor function score of upper extremity.5,6 The normal score is 11; the lower score is chosen when the neurological disturbance is different between the left and right sides. Postoperative neurological recovery was estimated on the basis of the recovery ratio = (postoperative – preoperative) / (11 – preoperative) x 100%. A score of 75% to 100% is designated excellent, 50% to 74% good, 25% to 49% fair and 0% to 24% poor.7 Functional disability from gait abnormality was graded according to the system proposed by Nurick.8 Grade 1, no difficulty in walking and working; grade 2, gait abnormality not interfering with work; grade 3, gait abnormality preventing work; grade 4, walk only with assistance and grade 5, wheelchair-bound or bedridden.

OLF was detected by MR imaging, which appeared hypodense on T1- and T2-weighted images, situated consistently as a linear or beak-like excrescence, uniformly hypodense on T1- and T2-weighted images, situated posterior to the thecal sac, compressing it (Figs. 1 and 2).

Results

Motor weakness in the lower limbs and disturbance in gait were the most common initial symptoms in all 5 patients. Two of them had associated numbness of the lower limbs as the initial symptom; the rest developed numbness later. The distribution of numbness was non-dermatomal in nature, and only 3 patients had tract sensory level corresponding to lower thoracic compression. Back pain, stiffness and radicular pain were not prominent complaints. Urinary disturbance was present in 2 patients. Two patients presented with acute weakness, 1 of them with acute paraplegia after a road traffic accident, the other had paraparesis 1 week following total knee replacement. On examination, all had impaired vibration and position sense. Only 1 patient presented with brisk reflexes and spasticity; the rest had diminished reflexes and tone in their lower limbs. All patients had JOA score<6 and all underwent laminectomy. Post-surgery follow-up ranged from 1 to 6 years, with a median of 2 year. All improved in their gait and spasticity, but 4 patients had persistent sensory deficit. Based on the JOA Recovery Ratio by Hirabayashi,7 3 patients had good-to-excellent outcome, 2 had poor-to-fair outcome, although the 1-year follow-up may be too short a duration for any significant neurological improvement in Case 5. The patients who improved after surgery (Cases 1, 2 and 3) had consistent improvement in their ambulation and gait; their JOA scores improvement ranges from 4 to 5 (median of 4). Those who present with slow onset compression (Cases 1 and 2) had better recovery after surgery compared to those with acute myelopathy (Cases 3, 4 and 5). Case 5, who had compression from both OLF and OPLL, only had fair outcome after surgery. Case 3, who had micturition difficulty, reported improved bladder control after surgery, but, Case 4, who developed urinary retention after traffic accident, still required self intermittent urethral catheterisation after 6 years of rehabilitation. Improvement in Nurick and MRC scores ranges from 0 to 2 only (median of 1). Compared to the JOA, both Nurick and MRC scoring systems are less useful in grading postoperative improvement in myelopathy in our series (Table 1).

Discussion

Although first reported by Polgar in 1920 using plain lateral radiographs, most reports of thoracic myelopathy originated from Japan. One of the largest series was described by Miyasaka et al.9 This ligamentous osseous proliferation was reported to occur in up to 20% of Japanese individuals >65 years old and many reports viewed it as a normal feature of the ageing Asian spine.10-12 Iwasaki et al13 reported 4 cases of OLF in the cervical spine. All these cases involved women aged 62 to 70 years and half of them showed ossification of other spinal ligaments. With increasing use of modern imaging modalities, more reports have emerged describing the same disorder in non-Asians.14-16 Okada et al14 found that the thickest OLF was located predominantly in the lower third of the thoracic spine. The most common symptomatic level was T10 to T11 (64%) and T11 to T12 (21%); in our series, it was T10 to T11 (3 out of 5 patients). Smith et al15 had also noted that OLF most commonly occurred at T9 to T12; this region appears to be particularly prone to degenerative processes due to the high tensile force present in the posterior column. There may be
associated kyphosis in the lower thoracic spine and this suggested that altered mechanical stresses at this area led to the development of OLF. Maigne et al reported that OLF occurred most frequently at the thoracolumbar junction and its appearance seemed to correlate with a unique orientation of the zygapophyseal joints that contributed to increased rotatory instability and micromotion. Similarly, Otani et al postulated a localised mechanical aetiology when they noted that 88% of symptomatic thoracic disc herniations were associated with OLF at the same level, and that 87% had marginal syndesmophytes formation at the same level. Later studies demonstrated collagen hyperplasia and hypertrophy in response to localised stress, resulting in thicker ligament and higher collagen content with deposition of both calcium pyrophosphate dihydrate and calcium hydroxyapatite in the ligament; these changes correlated with age and the degree of degeneration present. This fact was further supported by Wang et al who observed that 16.3% of elderly Taiwanese with degenerative cervical spondylosis also had OLF.

Initially, the ligament hypertrophies and calcifies before it ossifies. The ossification process starts at the base of the ligament with endochondral ossification of the hypertrophied, vascularised fibrocartilaginous tissue. Early

Table 1. Ossified Ligamentum Flavum Case Series

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Preoperative symptoms</th>
<th>Preoperative neurological examination</th>
<th>Level of compression</th>
<th>Postoperative follow-up</th>
<th>Recovery Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>45</td>
<td>M</td>
<td>Gait difficulty “giving-way” for 1 year. Weakness and numbness in both legs. Motor JOA 2. Nurick 2.</td>
<td>Bilateral hyperreflexia and hypertonia at knees and ankles. Babinski absent. Decreased sensibility in both legs. MRC 4. JOA 7.</td>
<td>T10-11 (Figs. 1 and 2)</td>
<td>At 15 months’ follow-up, no weakness in climbing stairs, good symmetrical strength bilaterally, no numbness, no hyperreflexia nor hypertonia. MRC 5. JOA 11. Nurick 1.</td>
<td>100% Excellent</td>
</tr>
<tr>
<td>3</td>
<td>65</td>
<td>F</td>
<td>Acute weakness and numbness in both legs 1 week following left total knee replacement, unable to walk. Motor JOA 0. Nurick 5.</td>
<td>Knee and ankle jerks absent bilaterally. Babinski positive bilaterally. T12 sensory level with impaired vibration and position sense. MRC 3. JOA 3.</td>
<td>T12-L1</td>
<td>At 2 years’ follow-up, improvement in ambulation and able to walk with walking frame. Numbness still present in both feet but improved. MRC 4. JOA 7. Nurick 4.</td>
<td>50% Good</td>
</tr>
<tr>
<td>4</td>
<td>56</td>
<td>F</td>
<td>Acute paraplegia after being knocked down by a car. Motor JOA 0. Nurick 5.</td>
<td>Bilateral loss of knee and ankle reflexes and complete flaccid paralysis with sensory level at T12 and acute urinary retention and lax anal tone. No fracture dislocation on X-ray and CT (Figs. 3 to 6).</td>
<td>T8-9 and T10-11 with almost complete spinal canal obliteration and spinal cord oedema.</td>
<td>After 6 years of rehabilitation, she was still wheelchair-bound with little motor nor sensory recovery, and requiring intermittent urethral catheterisation. MRC 1. JOA 0. Nurick 5.</td>
<td>0% Poor</td>
</tr>
<tr>
<td>5</td>
<td>53</td>
<td>F</td>
<td>Progressive inability to walk with bilateral lower limb weakness and numbness. Motor JOA 0. Nurick 5.</td>
<td>Bilateral lower limb loss of reflexes and tone. MRC 3. JOA 5. Anterior T7-8 discectomy with rib strut graft was performed for thoracic disc herniation. Postoperative recovery was complicated by Aspergillus osteomyelitis with epidural abscess, requiring anterior drainage, corpectomy and fusion of T6-8. Motor and sensory deficits did not recover after surgery.</td>
<td>T10-11 compression of spinal cord by OLF and OPLL below the level of original discectomy of T7-8 discs</td>
<td>At 1 years’ follow-up, improvement in ambulation and able to walk with walking frame. Numbness still present in both feet but improved, other than persistent bilateral footdrop. MRC 4. JOA 7. Nurick 4.</td>
<td>33% Fair</td>
</tr>
</tbody>
</table>
ossification is usually found on the capsular side of the ligament in front of the facet joints. It expands on the outer surface of the ligament anteriorly towards the spinal cord, gradually giving rise to 2 paramedian nodules within the spinal canal that compress the posterolateral portion of the spinal cord. This lateral or capsular outgrowth rarely impinges on an exiting segmental nerve root, resulting in foraminal stenosis and radiculopathy. Both masses are usually connected by a film of elastic fibres which subsequently also ossifies on its epidural surface and occupies the spinal canal in the form of a central nodular mass. This further contributes to the cord compression as it thickens and extends in a caudocranial direction along the posterior part of the spinal canal in a linear fashion as a “beak-like” outgrowth. The ossified nodular masses were reported to also expand outwards and backwards onto both surfaces of the lamina: posterolaterally on the medial surface of the apophyseal joint and anteromedially on the surface of the lamina. High expression of transforming growth factor beta-1 (TGF-β1) by fibroblasts was found in the ossified matrix within ossified ligaments and in chondrocytes within cartilaginous areas adjacent to the ossified ligaments; TGF-β1 could have played a role in chondroid metaplasia and ectopic ossification in OLF. Under chronic compression, the spinal cord exhibits destructive changes believed to cause motor paresis. These include infarction, ischaemic necrosis and neuronal loss or
chromatolysis in the grey matter, as well as demyelination in the white matter in the posterior and lateral white columns. Yamaura et al. found that apoptosis in acute spinal cord injury induced both secondary degeneration at the site of injury, and chronic demyelination of tracts away from the site of injury. They observed apoptotic cells in the chronically compressed spinal cord in a human autopsy and mouse model. In mice with spinal cord compression, descending degeneration in the anterior and lateral columns and ascending degeneration in the posterior column were observed. Spinal cord cell apoptosis produced destructive changes in the spinal cord under chronic compression, resulting in profound and irreversible neuronal degeneration in the posterolateral long tracts. This might explain the persistence of residual spasticity at follow-up due to irreversible changes within the cord caused by significant thecal compression. Delay between the onset of initial symptoms and surgical decompression is believed to worsen these changes.

OLF may present as 2 distinct syndromes. The first involves chronic spinal cord compression over a long period of time and presents with unsteady gait, difficulty with balance and climbing stairs, with or without unilateral/bilateral neurogenic claudication. This is followed by progressively increasing spastic paraparesis during walking on level ground and requiring walking aid or support. Sensory, bladder and bowel involvement are not uncommon in the later stage. Physical examination of the lower extremities reveals both long tract signs (increased tone and reflexes, motor and sensory deficits) and posterior column signs (ataxia, dysdiadochokinesia, loss of vibration, proprioception, and 2-point discrimination senses). Rarely, there may be radicular pain with nerve tension signs (straight leg-raising test) if there is foraminal stenosis. In the second syndrome, OLF may present as acute myelopathy after minor trauma. There is a sudden compromise in an asymptomatic, but narrowed, spinal canal by haematoma and oedema, with or without bony/soft tissue impingement secondary to the trauma. The motor and sensory deficits are usually more severe than the first syndrome and recovery is usually poor (as in Case 4). Differential diagnoses, including high myelomeningocele, haemangioma, arteriovenous malformation, neurosyphilis, multiple sclerosis, spinal tumour and infection, must be excluded.

Epidural-evoked potentials often show disappearance of the third negative component and positive-going waves in the most compressed area. Large nodule of OLF is sometimes visible on plain radiographs’ especially in the cervical spine, but can be difficult to visualise in the thoracic spine due to overlap from the shoulder and liver (Fig. 3). The definite diagnosis and nature of the compressing OLF are established by spiral CT with multi-axial reconstruction, although it is best combined with intrathecal myelogram demonstrating the lateral extension, size, length of OLF and any nerve root compression (Fig. 4). This necessitates an extended bony window to ensure discrimination of an ossified ligament from thecal metrizamide. Thickened ligamentum flavum are often found with degenerative disease and spinal stenosis at multiple levels, and should not be ignored as a possible major contributing factor to the patient’s symptoms when detected on CT myelogram. In addition, patients with a secondary block from a thickened ligamentum flavum should be studied from above with C1-C2 puncture to exclude other clinically significant diseases. Attention to findings of thickened ligamentum flavum is important since surgical intervention involves removal of the entire ligament(s); otherwise, the clinical symptoms may not be relieved. Review of 1021 CT studies prospectively and found 6080 ossified ligamentum flava. In >96% of cases, ossification was confined to the lateral articular portion of the ligament and central ossification was rare. With the advent of MR imaging, more cases of OLF were detected. This appears as a linear or beak-like excrescence, uniformly hypodense on T1- and T2-weighted images, situated posterior to the thecal sac. T2-weighted MR imaging is particularly useful in showing the degree of spinal cord injury (Figs. 2 and 5) and the longitudinal extent and multiple sites of spinal cord compression (Fig. 6). As such, the use of intrathecal contrast can be avoided because MR imaging provides good visualisation of the entire length of the spinal cord and thecal sac. It also detects high-signal intramedullary intensity within the spinal cord on T2-weighted images that might indicate poor prognosis. On the other hand, cases of OLF “missed” on MR imaging and later diagnosed by CT imaging had also been reported. This is because the null signal of ossification can be difficult to detect on MR imaging, unlike the hyperdense ossified lesion detected on CT imaging. It is, therefore, useful to combine CT and MR imaging: the former diagnoses the aetiology of the compression, while the latter indicates the compression level and cord changes.

Prompt surgical intervention and appropriate rehabilitation management play key roles in improving the functional outcome of myelopathy caused by OLF. The prognosis after decompressive surgery can be good, especially if intramedullary hyper intensities are absent on preoperative T2-weighted MR images. Posterior decompressive laminectomy (with or without medial facetectomy) and removal of the enlarged ossified yellow ligament are the most common surgical procedures performed in patients with compressive thoracic myelopathy due to OLF. Okada et al., however, recommended performing laminoplasty with preservation of the posterior element after decompression. This is because of the unsatisfactory results observed after laminectomy alone,
with late neurological deterioration due to the recurrence of OLF at the same site or increased kyphotic deformity of the spine (especially with no fusion after more than 1-level decompression). Trivedi et al.\(^4\) observed that none of their patients retained the ability to run briskly and all of them had residual spasticity despite decompression. In our series, 1 patient had excellent result, 2 good and 1 fair; the patient with spinal trauma had a poor outcome. The poor outcome of laminectomy alone was also reported by other investigators.\(^3\) They suggested that the spinal cord in the OLF-related thoracic narrow spinal canal, being ischaemic and oedematous due to prolonged cord compression, was more prone to the slightest cord damage (such as, intraoperative hypotension). This may be worsened by a concurrent OPLL ventral epidural encroachment (rare but not uncommon), giving rise to marked posterior expansion of the cord and persistence of neurological deficit after posterior laminectomy alone. Posterior circumferential or combined anterior and posterior decompression with fusion is recommended in stenosis involving OLF and OPLL (as in Case 5).\(^3\) Furthermore, late neurological regression or local pain might be related to worsening post-laminectomy kyphotic deformity, resulting in local recurrence of OLF or the development of OLF at upper levels.\(^6\) Ascending narrowing at other thoracic levels, with spontaneous progression after laminectomy at a lower level, was also observed by separately Van Oostenbrugge et al and Yonenobu et al.\(^6\) They postulated that although patients with spondylosis might have a stable spine, the membrane at and above the laminectomised area might be stretched and subsequently ossified due to increased local mechanical stress. Excision of OLF also has its risk. Long-standing ossified ligament adheres to the dura and its removal risks major dural disruption. Occasionally, thick OLF had to be excised with the adherent outer layer of the dura.\(^1\)This rarely leads to cerebrospinal fluid leakage and fistula formation.\(^1\) Shiokawa et al.\(^2\) found that thoracic OLF frequently developed in the lower thoracic spine in middle-aged men and was often complicated by various spinal lesions in many cases (such as OPLL and disc protrusion). The preoperative duration of symptoms and extent of compression on CT were significantly shorter in patients with excellent outcomes than in those with fair outcomes. No significant difference was observed in the correlation between other factors (age, sex, morbidity, initial symptoms, chief complaint, duration of symptoms, patellar reflex, Achilles reflex, intramedullary change determined by MR imaging and coexisting spinal lesions). They, therefore, concluded that a high index of suspicion, early diagnosis and treatment were important in OLF as their findings have suggested that a delay in diagnosis and treatment correlated with poor functional prognosis postoperatively.

In conclusion, OLF can significantly contribute to a spatial reduction of the thoracic spinal canal, resulting in slowly progressive paraparesis or acute paraplegia after trauma to the back. The T2-weighted sagittal image of MR imaging is the modality of choice for screening the longitudinal extent of OLF; its diagnostic accuracy is increased when combined with CT myelogram. Neurological improvement can be expected following decompressive laminectomy, with or without excision of the OLF. However, the persistence of residual numbness or weakness at follow-up might be due to irreversible changes in the cord, which are attributed to significant thecal compression and delay between the onset of initial symptoms/signs and surgical decompression. Prognosis remains poor for acute myelopathy with preexisting OLF, despite surgery.

REFERENCES