Case Report

Ossification of the proximal thoracic ligamenta flava causing acute myelopathy in a Caucasian: case report and literature review

DP Tokala^{*,1,2}, KS Lam^{1,3} and HG Prince^{1,4}

¹The Centre for Spinal Studies and Surgery, University Hospital, Queen's Medical Centre, Nottingham, UK

Study design: Case report and literature review.

Objective: To illustrate that ossification of the proximal thoracic ligamenta flava can be a rare cause of acute myelopathy in a Caucasian patient and that timely surgery can lead to a good outcome.

Setting: Nottingham, UK.

Methods: Proximal multiple contiguous ossified thoracic ligamenta flava from T3/T4 to T5/T6 causing acute myelopathy was diagnosed in a Caucasian man based on history and examination followed by magnetic resonance imaging and computed tomography scanning. The literature is reviewed for all reported cases of ossified ligamenta flava causing myelopathy in Caucasians.

Results: Following prompt diagnosis and T3 to T5 laminectomies, our patient made near-complete neurological recovery over a 10-month period. This condition usually affects the lower thoracic spine. Although chronic and subacute myelopathy secondary to this circumstance has been reported in Caucasians, acute myelopathy has not been reported and proximal thoracic involvement has been reported twice.

Conclusion: Ossification of the proximal thoracic ligamenta flava can be a rare cause of acute myelopathy in Caucasians. Prognosis following decompressive surgery is usually good.

Spinal Cord (2007) 45, 310–313. doi:10.1038/sj.sc.3101953; published online 27 June 2006

Keywords: thoracic myelopathy; ossification; ligamenta flava; caucasian

Introduction

Proximal thoracic myelopathy is mainly due to neoplastic disease in Caucasians. A less recognised cause is the ossification of the ligamenta flava (OLF), a condition that is rarely reported in the Western literature. Although thoracic OLF usually occurs in the lower thoracic spine,^{1–10} it has been reported to occur at higher thoracic levels resulting in proximal myelopathy.^{11,12} This is particularly important because, regardless of the level and severity of the disease, OLF is easily treatable by decompressive laminectomy if the diagnosis is made early.

Case report

A 45-year-old Caucasian man presented with a 3-week history of acute progressive lower limb weakness, walking difficulties and loss of sensation below the epigastrium. His upper limbs were unaffected and he

remained doubly continent. Examination revealed a 6 feet 6 inches tall and obese gentleman who walked with a very unsteady broad-based gait. He had a sensory level at T6 with bilateral loss of lower limb proprioception. Both lower limbs were hypertonic with weakness of the proximal hip girdle musculature, but normal power in the distal muscle groups. There were bilateral ankle cloni, lower limb hyperreflexia and up-going plantars. Rectal examination was normal.

Haematological investigations and plain radiographs were normal. Magnetic resonance imaging (MRI) scans showed multiple contiguous hypointense posteriorly based lesions extending from T3/T4 to T5/T6 causing spinal cord compression and myelomalacia at T3/T4 level (Figure 1). Computed tomography (CT) scans showed a right-sided posterolateral hyperdense mass at T3/T4 interlaminar space, causing extradural compression of the spinal cord. Similar but smaller lesions were identified at T4/T5 and T5/T6 (Figure 2).

Bilateral laminectomies at T3, T4 and T5 were performed revealing OLF at all three levels. A generous and careful decompression of the spinal cord was performed. There were no intra- or postoperative

*Correspondence: DP Tokala, 7 Frankland close, Bath BA1 4EJ, UK

²Current address: Royal United Hospital, Combe park, Bath, UK

³Current address: Guy's Hospital, St Thomas' Street, London, UK

⁴Current address: Queen's Medical centre, Nottingham, UK

Figure 1 Sagittal T1- (a) and T2- (b) weighted MRIs showing marked spinal cord compression with myelomalacia at T3/T4 and less severe compressions at T4/T5 and T5/T6 levels caused by posterior low signal intensity lesions

complications. At 10 months follow-up, he was walking unaided within the house, was using one stick for short distances and a wheelchair for long distances. The motor power and sensation, including proprioception in his lower limbs, returned to normal. However, there was persistent bilateral lower limb hyper-reflexia, up-going plantars but no ankle cloni.

Discussion

Tumour and less commonly bacterial infections are the two major causes of thoracic myelopathy in Caucasians. Myelopathy due to other causes occurs less frequently in the thoracic spine, because it is less prone to degeneration as the ribs restrict movement.³ Thoracic compressive stenotic syndromes are primarily due to the lamina, facet or ligamentum flaval hypertrophy^{13,14} and rarely due to an intervertebral disc herniation.¹⁵ Three conditions causing ossification of the spinal ligaments have been described. Diffuse idiopathic skeletal hyperostosis (DISH) and ossification of posterior longitudinal ligament (OPLL) are comparatively much more common conditions than OLF, the latter being almost exclusively reported from Asian centres.^{4,5,16}

Figure 2 (a and b) Axial CT scans showing an ossified posteriorly located extradural lesion. (c and d) Sagittal reformatted CT scans showing the ossified ligamenta flava worst at T3/T4 than T4/T5 and T5/T6 levels

Table 1 Review of literature: thoracic myelopathy secondary to ossified ligamenta flava in Caucasian patients

<i>Authors and year</i>	<i>Race</i>	<i>Age/sex</i>	<i>Duration of symptoms</i>	<i>Levels involved</i>	<i>Treatment</i>	<i>Follow-up</i>	<i>Outcome</i>
Omojola <i>et al</i> ⁷	? C	53M	> 3years	T10–T12	Laminectomy T10–T12	6 weeks	Marked improvement
Arafat <i>et al</i> ³	C	51F	9 months	T9–T11	Laminectomies T9–T11	10 days	Marked improvement
Parekh <i>et al</i> ⁸	C	34M	2 year (intercostal pain)	T9 and T10 + small lesion at T5	Laminectomy T9, T10 and T5	?	Marked improvement
Shiraishi <i>et al</i> ⁹	C	72F	18 months	T9–T10	Selective excision of ligament	7 months	Marked improvement
Vera <i>et al</i> ¹⁰	C	48M ^a	6 months	T10	Laminectomy T10	5 years	Marked improvement
Van Oostenbrugge <i>et al</i> ¹²	C	57M	18 months	T3–T12	Laminectomy T3–T4	2 months	Slow improvement
Payer <i>et al</i> ²	C	58M	4 years	T10–T12 initially 2 years later T8 and T9	(1) Laminectomy T10–T12 (2) Laminectomy T8–T9	1 year	Slow improvement
Kruse <i>et al</i> ¹¹	? C	(1) 54 (2) 73	?	(1) T9–11 (2) T2–T7	(1) Laminectomy T9–11 (2) Laminectomies T2–7	(1) 3 years (2) 2 years	1. Marked improvement 2. Slow improvement
Celli <i>et al</i> ⁶	C	58 M	?	T10–T11	Laminectomy ? Level	1 year	Marked improvement

^aHad associated X-linked hypophosphataemia + OPLL

Aetiology

The pathoaetiology of OLF remains uncertain. The higher prevalence in Japan implicates genetic and/or environmental factors.¹⁷ Patients with OLF have higher frequency of non-insulin dependent diabetes,¹⁷ obesity, hyperinsulinism, haemochromatosis and calcium metabolism abnormalities.⁶ However, the pathomechanism remains unclear. Our patient suffered from both obesity and non-insulin-dependent diabetes. Thoracic OLF usually occurs at T9–T12,^{1,4,5} this region being particularly prone to degenerative process.¹⁴ Otani *et al*¹⁸ noted a high incidence in cases with thoracic hyperkyphosis and suggested that the altered mechanical stresses as a consequence of this predispose to the development of OLF. Although our patient had a normal thoracic kyphosis, his MRI scan showed multilevel thoracic disc degeneration. Recent studies revealed that fibronectin, bone morphogenic proteins and transforming growth factor beta played an important role in the development of ossification of the spinal ligaments.¹⁷

Clinical course

OLF predominantly affects middle-aged men with a male to female ratio of approximately 2.7:1.⁶ The clinical course is characterised by progressive myelopathy. The most common symptoms are altered gait, weakness and numbness in the lower limbs, and the signs include spastic paraparesis with hyper-reflexia, up-going plantars, clonus; and sensory deficits mainly involving deep sensation and proprioception.^{5,6,19} Sphincter disturbance and radicular symptoms are rare.

Radiological findings

The combination of MRI and CT is required for the precise diagnosis of OLF.¹ MRI accurately delineates the location and extent of spinal cord compression in several planes, but has difficulty differentiating between an ossified or hypertrophied ligament.¹⁶ CT scanning best demonstrates the increased density of the ossific changes. Plain spinal radiographs may show beak-like bone projections from the apophyseal joints into the spinal canal on the lateral views.¹⁷

Histology

Age-related enlargement of the ligamenta flava is common. Ono *et al*¹⁷ reported that ossification of the spinal ligaments have the following characteristics: (1) ectopic bone formation within the ligaments; (2) tissue hyperplasia and cellular proliferation; (3) before ossification, sequential occurrence of fibrocartilaginous cellular proliferation followed by calcification, tissue resorption with vascular ingrowth and (4) specific site of predilection and often occurring in combination with senile ankylosing vertebral hyperostosis or DISH.

Surgery

The surgical procedure most commonly performed is a laminectomy with removal of ossified ligaments (Table 1). Okada *et al*⁴ recommended a laminoplasty. This may provide better stability in patients with extensive lesions and may reduce the incidence of postoperative kyphosis and restenosis owing to the scar tissue. Medial facetectomy with or without supplemental internal fixation can be performed if degenerative hypertrophy of the articular masses is present.²⁰

Prognosis depends not only on the extent of the lesions but also on the time to diagnosis and treatment.¹⁹ The clinical outcome is usually good if early surgical decompression is performed^{1,19} (Table 1). Regular follow-up is necessary as recurrence or development of ossification at other levels has been reported.^{2,20}

Conclusion

Over the past few decades, OLF has been occasionally reported in non-Asian patients. This is the first report of acute proximal thoracic myelopathy caused by OLF in a Caucasian. OLF is an eminently treatable condition if diagnosed early. Hence OLF should be considered in the differential diagnosis of all patients presenting with thoracic myelopathy, even when the upper thoracic spine is involved.

References

- Hanakita J, Suwa H, Ohta F, Nishi S, Sakaida H, Iihara K. Neuroradiological examination of thoracic radiculomyelopathy due to ossification of the ligamentum flavum. *Neuroradiology* 1990; **32**: 38–42.
- Payer M, Bruder E, Fischer JA, Benini A. Thoracic myelopathy due to enlarged ossified yellow ligaments. Case report and review of the literature. *J Neurosurg* 2000; **92**: 105–108.
- Arafat QW, Jackowski A, Chavda SV, West RJ. Case report: ossification of the thoracic ligamenta flava in a Caucasian: a rare cause of myelopathy. *Br J Radiol* 1993; **66**: 1193–1196.
- Okada K, Oka S, Tohge K, Ono K, Yonenobu K, Hosoya T. Thoracic myelopathy caused by ossification of the ligamentum flavum. Clinicopathologic study and surgical treatment. *Spine* 1991; **16**: 280–287.
- Yonenobu K *et al*. Thoracic myelopathy secondary to ossification of the spinal ligament. *J Neurosurg* 1987; **66**: 511–518.
- Celli P, Caroli E, Trillo G, Ferrante L. Ossification of the ligamentum flavum in a Caucasian. Case report. *J Neurosurg Sci* 2002; **46**: 96–99; discussion 9.
- Omojola MF, Cardoso ER, Fox AJ, Drake CG, Durward QJ. Thoracic myelopathy secondary to ossified ligamentum flavum. *J Neurosurg* 1982; **56**: 448–450.
- Parekh HC, Gurusinghe NT, Perera SS, Prabhu SS. Ossification of the ligamentum flavum in a Caucasian: case report. *Br J Neurosurg* 1993; **7**: 687–690.
- Shiraishi T, Crock HV, Lewis P. Thoracic myelopathy due to isolated ossification of the ligamentum flavum. *J Bone Joint Surg Br* 1995; **77**: 131–133.
- Vera CL *et al*. Paraplegia due to ossification of ligamenta flava in X-linked hypophosphatemia. A case report. *Spine* 1997; **22**: 710–715.
- Kruse JJ, Awasthi D, Harris M, Waguespack A. Ossification of the ligamentum flavum as a cause of myelopathy in North America: report of three cases. *J Spinal Disord* 2000; **13**: 22–25.
- van Oostenbrugge RJ, Herpers MJ, de Kruijk JR. Spinal cord compression caused by unusual location and extension of ossified ligamenta flava in a Caucasian male. A case report and literature review. *Spine* 1999; **24**: 486–488.
- Jaspan T, Holland IM, Punt JA. Thoracic spinal canal stenosis. *Neuroradiology* 1987; **29**: 217.
- Smith DE, Godersky JC. Thoracic spondylosis: an unusual cause of myelopathy. *Neurosurgery* 1987; **20**: 589–593.
- Arseni C, Nash F. Thoracic intervertebral disc protrusion: a clinical study. *J Neurosurg* 1960; **17**: 418–430.
- Yamashita Y *et al*. Spinal cord compression due to ossification of ligaments: MR imaging. *Radiology* 1990; **175**: 843–848.
- Ono K, Yonenobu K, Miyamoto S, Okada K. Pathology of ossification of the posterior longitudinal ligament and ligamentum flavum. *Clin Orthop* 1999; **359**: 18–26.
- Otani K, Aihara T, Tanaka A, Shibasaki K. Ossification of the ligamentum flavum of the thoracic spine in adult kyphosis. *Int Orthop* 1986; **10**: 135–139.
- Shiokawa K, Hanakita J, Suwa H, Saiki M, Oda M, Kajiwara M. Clinical analysis and prognostic study of ossified ligamentum flavum of the thoracic spine. *J Neurosurg* 2001; **94**: 221–226.
- Nishiura I, Isozumi T, Nishihara K, Handa H, Koyama T. Surgical approach to ossification of the thoracic yellow ligament. *Surg Neurol* 1999; **51**: 368–372.