

Ossification of The Posterior Longitudinal Ligament As A Rare Cause of Myelopathy Among Turks: Report of A Case

ISMAIL H. TEKKÖK, KEMAL BENLİ

Department of Neurosurgery, Hacettepe University School of Medicine, Sıhhiye, Ankara 06100, Türkiye

Running Title : Ossified posterior longitudinal ligament

Abstract : The case of a 63-year-old man who presented with Brown-Sequard syndrome plus posterior column deficits due to extensive ossification of the posterior longitudinal ligament is reported. Diagnostic evaluation included magnetic resonance imaging in addition to conventional techniques. The patient was surgically treated by corpectomy and fusion using an anterior approach and made a complete recovery. The occurrence of ossified

posterior longitudinal ligament and the diagnostic features are briefly reviewed. With more widespread awareness of this entity and more frequent use of magnetic resonance imaging, we believe more cases will be diagnosed.

Key words : Magnetic resonance imaging, Ossification, Pathologica Posterior longitudinal ligament, Spina cord compression

INTRODUCTION

Compression of the spinal cord by ossification of the posterior longitudinal ligament (OPLL) was originally described by Key as early as 1838 (9). After Tsukimoto's report of an autopsy case in 1960 (19), it was recognized as a clinical entity and similar cases were reported thereafter in Japan, at where 2142 patients with OPLL were registered between 1960 and 1977 (15). Accordingly, it was long regarded as a disease of the Orient, with a prevalence of 2% among the Japanese, almost 1% among Koreans and 0.83% among the Chinese (7,10). OPLL is a very rare condition among Caucasians (2,3,4,5,11,12,22), only 29 cases had been reported, when Hanna and Watt reviewed the literature in 1979 (4). However, with increased use of computed tomography (CT) and recently of magnetic resonance imaging (MR) in the evaluation of cervical myelopathy, this disorder has become more frequently recognizable. Starting with Firroznia et al (3) reporting eleven white patients, series of Caucasians although rather small compared to those of the Japanese, started appearing in the literature

(2,5,8,12) with the total number still not exceeding sixty by 1992. We have recently encountered a case of extensive OPLL presenting with myelopathy which is the first case in the 33-year history of our institution. The purpose of this paper is to present the clinical, radiological and surgical findings of this relatively uncommon disease.

CASE HISTORY

A 63-year-old man presented with a two-month history of rightsided weakness and difficulty in walking. He reported that a urinary catheter had to be inserted a week prior to admission because of difficulty in emptying his bladder. There was no history of trauma.

Neurological examination on admission showed a poorly-nourished man who was unable to stand. Global power was 1-2/5 in the right arm and 2/5 in the right leg. There was marked increase in tone of the right leg. The biceps, triceps and styloradial reflexes could not be elicited on the right. Deep tendon reflexes in the legs were brisk throughout.

Plantar response was upgoing bilaterally. Sensation to pin-prick and light touch was diminished below C5 on the left. Vibration sense was impaired in both lower extremities. There was gross atrophy of the intrinsic muscles of the right hand. Anal tone and anal reflex were diminished.

The clinical diagnosis was lower cervical myelopathy and radiculopathy. Plain x-rays of the spine showed a thick ossification behind the vertebral bodies extending between C4 and C7 as well as some spondylotic changes of the vertebral bodies (Fig. 1). An emergency CT-myelography with iohexol demonstrated compression of the spinal cord over several segments. There was massive posterior displacement of the cord with an anteroposterior dimension of 3 mm at C4-5 level (Fig. 2). MR imaging was performed on a 0.5T superconductive unit (Gyroscan, Philips Medical Systems, Netherlands), which confirmed the findings deduced from the CT-myelogram. The OPLL appeared as a thick hypointense band behind the vertebral bodies C4-C7 (Fig. 3).

Buckling of the yellow ligament at C3-4 and C4-5 contributed to spinal cord compression, creating an "hour glass" appearance.

Preoperative evaluation revealed obstructive pulmonary disease so surgery had to be postponed for a few days for bronchodilator therapy and chest physiotherapy. Meanwhile dexamethasone 4mg qd was started. A skeletal survey to evaluate possible locations of diffuse idiopathic skeletal hyperostosis (DISH) proved negative. The patient subsequently underwent surgery. With a classical anterior approach the anterior surfaces of the vertebral bodies C4-C7 were exposed. Medial portions of vertebral bodies C4,5,6 and the upper portion of C7 were drilled creating a longitudinal canal 15 mm wide and 55 mm long. As the vertebrectomy proceeded posteriorly, using the operating microscope, the dura was initially exposed at the site of relatively less cord compression, i.e. C7, working upwards with angled microcurettes. The OPLL was like an ivory-hard osteoma, but could easily be dissected from the dura and was completely removed. There was no spread of ossification into the intervertebral foramina. An autogenous graft from the ribs was fitted and secured in the drilled canal, and a rigid cervical collar was applied before the patient recovered from anaesthesia. The postoperative course was unevent-

ful. There was no morbidity associated with the donor site. At 6 months postoperatively the patient is fully continent and able to walk with a cane.

DISCUSSION

The aetiology of OPLL and the reason why there is a high incidence amongst the Japanese is still obscure. Degenerative disc disease (13,14), trauma (19), chronic fluorine poisoning (18), infection (17), dietary factors and disturbance of the calcium metabolism (21) have all been proposed but none has been established as the cause. Cases in association with ankylosing spondylitis have also been reported (16). On the other hand, an association between DISH previously known as Forestier's Disease and OPLL has long been recognized (16) which may suggest a hereditary diathesis of spinal ligament ossification.

Whatever the cause, impairment of spinal cord circulation appears to be most important in the pathophysiology of OPLL. The histopathology of the spinal cord in OPLL patients has been reported by a few Japanese authors (13,19). Macroscopically the cord was found to be anteriorly concave and flattened in these autopsy studies. By light microscopy severe infarction of the grey matter, ascending demyelination in the posterior columns, descending demyelination in the lateral columns and proliferation of hyalinized small blood vessels were found (13). These changes are similar to those found with cervical spondylotic myelopathy suggesting that both result from ischaemia upon reduction of the blood supply in the distal branches of the anterior spinal artery (23).

OPLL occurs predominantly in males with a male to female ratio ranging between 2:1 and 4:1 (5,8,10,13,14,15). When OPLL encroaches upon the spinal canal it may cause myelopathy due to compression or ischaemia and/or radiculopathy due to stretching of the nerve roots. OPLL is more likely to cause myelopathy in a canal already compromised by congenital stenosis, spondylosis or hypertrophy of the yellow ligament (5). Myelopathy occurred in 35-57% of cases in various series (5,8,10,14,15) and corresponds to a more than 60% compromise of the spinal canal. The ligament and in some instances the attached dura becomes calcified and ossified by enchondral calcification. Mean age at diagnosis was 47-55 in various series (5,8,10,20). In 70-95% of the cases ossification occurred in the cervical spine (3,20).

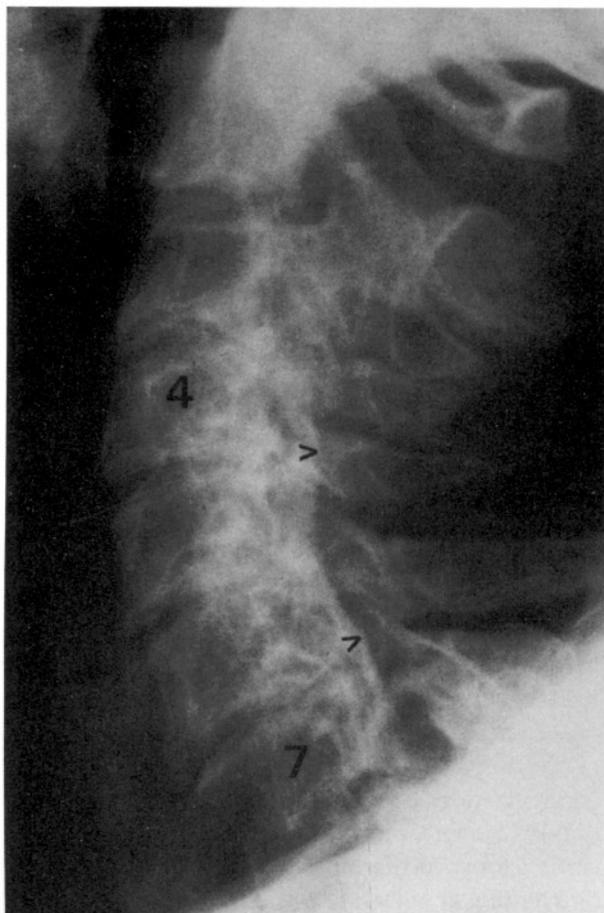


Fig. 1: Lateral plain x-ray of the cervical spine. Spinal canal is markedly narrowed between C4 and C7.

Cervical OPLL can be divided to segmental and continuous types. With the continuous type, an average of three vertebral levels are involved while in the segmental type OPLL, the ossification is interrupted at disc levels. Patients with segmental type OPLL are most often asymptomatic, but recognition of any type is crucial, because even with slight injury, the cord can easily be damaged.

A high index of suspicion is the best aid in the diagnosis of OPLL. It should be considered particularly in male patients over 40 years of age presenting with radiculo- and/or myelopathy. The diagnostic clue for OPLL lies in careful inspection of the lateral plain x-rays of the spine for a dense ossified strip along the posterior margin of the vertebral bodies, however OPLL may not be seen readily on plain x-rays in every patient. Myelography may demonstrate cord compression but fails to differentiate OPLL from disc herniation or spondylosis. CT-myelography with small interslice intervals is invaluable for diagnosis and operative planning (5). So far only three reports dealing with the MR characteristics of OPLL have appeared in the literature (11,20,22). Though MR is accepted to be poor in detecting bony pathologies, longitudinal extension of the ossification is better demonstrated on MR scans than on plain x-rays or CT scans, especially in the lower cervical and upper thoracic spine. On T1-WI, ossification appears as a

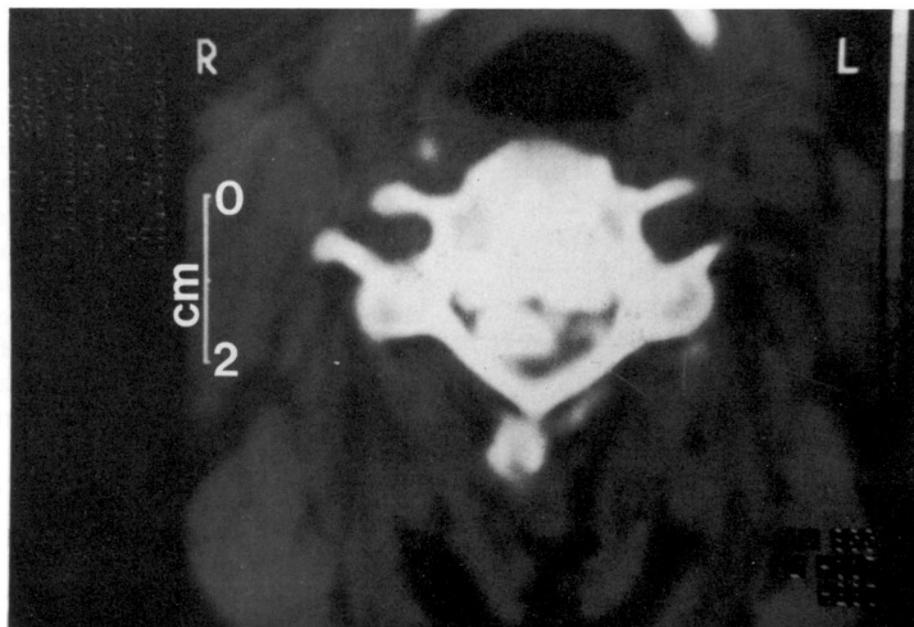


Fig. 2: Axial Ct image after intrathecal iohexol injection. Spinal cord is compressed and displaced by thick ossification of OPLL. Sagittal diameter of the spinal cord is 3 mm.

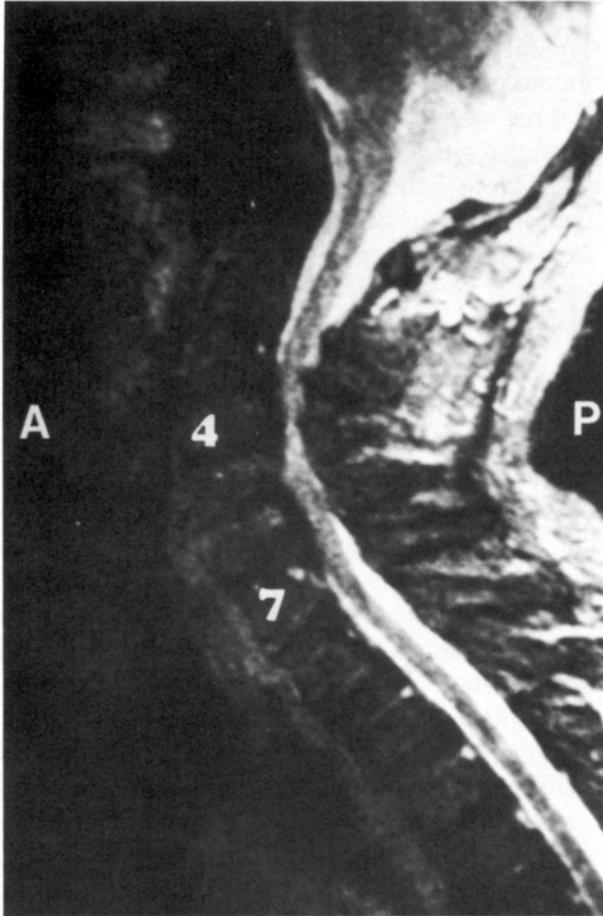


Fig. 3 : Sagittal T2-weighted MR image of the cervical spine demonstrates the marked compression of the cervical spinal cord between levels C4-C6. Ossified ligament appears hypointense.

low signal and it is hard to differentiate slight ossification from cerebrospinal fluid, whereas OPLL may be recognized as an impression on high signal cerebrospinal fluid flow on T2-weighted images (11,20,23). Differential diagnosis of low signal intensity on both T1 and T2-WI include calcified meningioma and arteriovenous malformation (AVM). A meningioma hardly ever spans several segments, while spinal AVM's are usually posterior and rarely cause compression. MR findings of segmental and continuous types of OPLL also differ. Continuous cervical OPLL is more easily recognized at MR imaging than segmental cervical OPLL. An average 6mm thick area of hypointensity is the finding associated with the continuous type (20). According to Yamashita et al, a band of intermediate to high signal intensity within areas of ossification considered to represent bone marrow can be observed in half of the cases with continuous OPLL (20), however we could not

identify such an increased intensity in our patient. In patients with segmental type OPLL, MR shows a thin area of hypointensity, therefore the differential diagnosis between cervical disc herniation, hypertrophy of the posterior longitudinal ligament, spondylosis and segmental OPLL is still difficult with the use of MR imaging alone (20).

The choice of surgical procedure for this entity, including laminectomy, laminoplasty and anterior decompression with fusion, still remains controversial. For almost two decades after Tsukimoto's popularization of OPLL, the preferred treatment was posterior decompression (14,15). With the standard posterior approach, the dura was not opened nor were the dentate ligaments cut; no attempt was made to remove the anterior ossified mass (14). In time, there appeared various concerns about laminectomy because with posterior decompression the root sleeves were often seen to be kinked posteriorly by the ossification making it necessary to remove the medial portion of the facets. The risk of subsequent instability, delayed neurological deterioration and persistence of radiculopathy due to remaining stretch of the nerve roots were the main causes of this dissatisfaction. Laminoplasty appeared as an alternative to overcome the complications arising with laminectomy (6,10), but it too is an indirect operation aiming at enlarging the canal. Posterior decompression remained popular until Abe et al. advocated the anterior approach in 1981 (1). Corpectomy, discectomy and microscopic removal of OPLL followed by interbody fusion is the currently used and most direct approach to the problem (5,8). Somatosensory evoked potential studies are now the established method not only in intraoperative monitoring but also in assessing the clinical course following treatment (8,23). In conclusion, in reporting a case of OPLL evaluated by MR and treated with the contemporary surgical method, our goal was to attract the attention of our colleagues to the disease which is known to be rare among Caucasians. With more widespread awareness of this entity and more frequent use of thin section CT-myelography and magnetic resonance imaging, we believe more cases will be diagnosed.

Correspondence : Dr. Ismail H. Tekkok,
Hacettepe Universitesi
Tıp Fakultesi
Beyin ve Sinir Cerrahisi
Ana Bilim Dalı Sıhhiye
Ankara 06100 Türkiye

REFERENCES

1. Abe H, Tsuru M, Ito T, Iwasaki Y, Koiwa M: Anterior decompression for ossification of the posterior longitudinal ligament of the cervical spine. *J Neurosurg* 55:108-116, 1981
2. Dietemann JL, Dirheimer Y, Babin E, Edel L, Dosch JC, Hirsch E, Wackenheim A: Ossification of the posterior longitudinal ligament (Japanese disease). A radiological study in 12 cases. *J Neuroradiol* 12:212-222, 1985
3. Firooznia H, Benjamin VM, Pinto RS, Golemb C, Rafii M, Leitman BS, McCauly OI: Calcification and ossification of posterior longitudinal ligament of spine. Its role in secondary narrowing of spinal canal and cord compression. *NY State J Med* 82:1193-1198, 1982
4. Hanna M, Watt I: Posterior longitudinal ligament calcification of the cervical spine. *Br J Radiol* 52:901-905, 1979
5. Harsh IV GR, Syper GW, Weinstein PP, Ross DA, Wilson CB: Cervical spine stenosis secondary to ossification of the posterior longitudinal ligament. *J Neurosurg* 67:349-357, 1987
6. Itoh T, Tsuji H: Technical improvements and results of laminoplasty for compressive myelopathy in the cervical spine. *Spine* 10:729-735, 1985
7. Izawa K: Comparative roentgenographic study on the incidence of ossification of the posterior longitudinal ligament and other degenerative changes of the cervical spine among Japanese, Koreans, Americans and Germans. *Nippon Seikeigeka Gakkai Zasshi* 54:461-474, 1980 (in Japanese)
8. Klara PM, McDonnell DE: Ossification of the Posterior Longitudinal ligament in Caucasians. Diagnosis and surgical intervention. *Neurosurgery* 19:212-217, 1986
9. Key CA: On paraplegia depending on disease of the ligaments of the spine. *Guys Hosp Rep (Series 1)* 3:17-34, 1838
10. Lee T, Chacha PB, Khoo J: Ossification of posterior longitudinal ligament of the cervical spine in non-Japanese Asians. *Surg Neurol* 35:40-44, 1991
11. Luetkehans TJ, Coughlin BF, Weinstein MA: Ossification of the posterior longitudinal ligament diagnosed by MR. *AJNR* 8:924-925, 1987
12. McAfee PC, Regan JJ, Bohlman HH: Cervical cord compression from ossification of the posterior longitudinal ligament in non-Orientals. *J Bone Joint Surg (Am)* 69:569-575, 1987
13. Murakami N, Muroga T, Sobue I: Cervical myelopathy due to ossification of the posterior longitudinal ligament. A clinico-pathologic study. *Arch Neurol* 35:33-36, 1978
14. Nagashima C: Cervical myelopathy due to ossification of the posterior longitudinal ligament. *J Neurosurg* 37:653-660, 1972
15. Ono K, Ota H, Tada K, Hamad H, Takaoka K: Ossified posterior longitudinal ligament. A clinico-pathologic study. *Spine* 2:126-138, 1977
16. Resnick D, Guerra J, Robinson CA, Vint VC: Association of diffuse idiopathic skeletal hyperostosis (DISH) and calcification and ossification of the posterior longitudinal ligament. *AJR* 131:1049-1053, 1978
17. Rozario RA, Levine H, Stein BM: Cervical myelopathy and radiculopathy secondary to ossification of the posterior longitudinal ligament. *Surg Neurol* 10:17-20, 1978
18. Singh AS, Dass R, Hayreh SS, Jolly SS: Skeletal changes in endemic fluorosis. *J Bone Joint Surg (Br)* 44:806-815, 1962
19. Tsukimoto H: A case report: autopsy of syndrome of compression of spinal cord owing to ossification within spinal canal of cervical spine. *Nippon Geka Hokan* 29:1003-1007, 1960 (in Japanese)
20. Yamashita Y, Takahashi M, Matsuno Y, Sakamoto Y, Yoshizumi K, Oguni T, Kojima R: Spinal cord compression due to ossification of ligaments. MR imaging. *Radiology* 175:843-848, 1990
21. Yoshikawa S, Shiba M, Suzuki A: Spinal cord compression in untreated adult cases of vitamin-D resistant rickets. *J Bone Joint Surg (Am)* 50:743-752, 1968
22. Yoshino MT, Seeger JF, Carmody RF: MRI diagnosis of thoracic ossification of posterior longitudinal ligament with concomitant disc herniation. *Neuroradiology* 33:455-457, 1991
23. Yu YL, Leong JCY, Fang D, Woo E, Huang CY, Lau HK: Cervical myelopathy due to ossification of the posterior longitudinal ligament. *Brain* 111:769-783, 1988