A rare neurological presentation due to ossification of the posterior longitudinal ligament of the thoracic spine and ankylosing spondylitis: case report

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Ossification of the posterior longitudinal ligament (OPLL) may be associated with certain rheumatic conditions including ankylosing spondylitis (AS), diffuse idiopathic skeletal hyperostosis (DISH) or spondylosis. More than 95% of all OPLL are localized at the cervical spine. Herein, we report a case of OPLL at the thoracic spine in an HLA-B₂₇-positive female patient with ankylosing spondylitis. The patient was presented to us with spastic paraparesis. The imaging studies included plain roentgenograms, tomograms, myelo-CT and magnetic resonance imaging (MRI). A continuous rod-like ossification along the posterior aspects of the fourth to sixth thoracic vertebrae with spinal cord compression was noted. The patient underwent a laminectomy from T4 to T6. At the second year follow-up examination, residual upper back soreness and mild left thigh pain were noted. However, the patient had resumed a full daily schedule and could walk freely without any support.

Keywords: ossification of posterior longitudinal ligament; ankylosing spondylitis; paraparesis; thoracic spine

Introduction

Ossification of the posterior longitudinal ligament (OPLL) is now a well-recognized disease in Japan with an incidence rate of 2%.^{1,2} It is predominantly found in males.^{1–4} Although most OPLL occurs in the cervical spine, combined or alone, it may also involve the thoracic and/or lumbar spine.^{1–3} In an extensive investigation, it was found that 207 out of 2162 Japanese patients (9.5%) with OPLL had the condition combined with ossification in the thoracic and/or lumbar levels.^{1,2} Trojan *et al.* in a series of 81 non-Japanese, found 10 patients (12%) with OPLL had similar ossifications.³ Interestingly, OPLL at the thoracic level is more common in females than in males, for reasons still unknown.^{1,2} The association of OPLL with ankylosing spondylitis (AS) is rare; however, this might be relatively common in Japan and its incidence is reported to be 2%.^{1,2} On the contrary, the concomitant of OPLL with diffuse idiopathic skeletal hyperostosis (DISH) seen on cervical radiographs is reported to be 50%.^{5,6} To our knowledge, ankylosing spondylitis with OPLL in the thoracic spine, causing spastic paraparesis, has not been previously reported in the English literature. This is a report of a 54-year-old female with such a condition.

Case illustration

A 54-year-old female suffered from increasing upper back pain and unsteady gait for 2 months. She had been confined to bed because of weakness of both legs 2 weeks prior to admission. Her history revealed a complaint of chronic low back pain dating back 10 years. The pain, constant in nature, improved with exercise. She denied having any pain in the peripheral joints and there was no history of urethritis, uveitis, conjunctivitis, diarrhoea or psoriasis. In addition, there was no family history of seronegative spondyloarthropathy.

On examination, the patient had limited chest expansion. A pin-prick test revealed sensory deficits below the xiphoid process, and motor strength of the left iliopsoas, quadriceps, extensor and flexor hallucis longus muscles was grade 3/5. The motor power of the right leg was grade 4/5. There appeared to be no sphincter disturbance. The patient had ankle clonus and increased patellar and Achilles tendon reflexes in both legs. The Babinski reflex was present bilaterally. The range of motion of the cervical spine, hips, knees and shoulders was normal. Laboratory evaluation showed an erythrocyte sedimentation rate (Westergren) 75 mm/h (normal <15), C-reactive protein 62 mg/l (normal <5). The anteroposterior roentgenogram of the lumbosacral spine showed a grade III sacroiliitis, a bamboo spine appearance and apophyseal joint ankylosis of the lumbar and lower thoracic spine (Figure 1).

Figure 1 Anteroposterior view of the lumbosacral spine demonstrating the bridging syndesmophytes (arrowheads) of the lumbar spine (a bamboo spine) and erosive, sclerotic changes of the bilateral sacroiliac joints. Apophyseal joint bony ankylosis was also noted

Plain thoracic roentgenograms showed ossification of the supra-interspinous ligaments and several levels of syndesmophytes anteriorly. The myelo-CT scanning (Figure 2) and plain tomographic studies (Figure 3) revealed a continuous rod-like ossification from T4 to T6 levels, with a significant spinal cord compression anteriorly but more towards the left side. The roentgenograms and CT examinations in the cervical and lumbar regions were also performed and showed an OPLL at the level of C5 vertebral body that did not cause any symptom. There was no OPLL in the lumbar region and no ossification of ligamentum flavum in the cervical or lumbar segments, either. The MRI of the thoracic spine demonstrated an extradural compression of the subarachnoid space, especially at the T5–6 disc level (Figure 4).

Laminectomy was decided on to be the best type of treatment and was performed from T4 to T6 levels. The postoperative course was rather uneventful. At the second year follow-up examination, it was noted that there was residual upper back soreness and mild left thigh pain. However, the patient was able to walk freely and had resumed her normal daily activities.

Figure 2 The myelo-CT examination shows calcification and ossification of the posterior longitudinal ligament at levels T5 and T6. Anterior cord compression, more on the left side, is also noted

Figure 3 Lateral tomograms demonstrate a rod-like ossification along the posterior aspects of the fourth to sixth vertebral bodies (arrowheads)

Discussion

Ossification of the anterior longitudinal ligament can occur in advanced AS^{7–10}; however, the association of OPLL and AS is not commonly seen. In Japan, in fact such a coincidence of OPLL and AS may be relatively

Figure 4 The T2-weighted MR imaging demonstrates a low signal along the posterior aspects of the fourth to sixth vertebral bodies, and significant anterior cord compression was noted at the T5–6 disc level

common, and its incidence is reported to be 2%.^{1,2} In a review of the literature, similar reported cases have been uncommon.^{9–12} The four reported cases all had involvement simultaneously of the cervical spine. Yonenobu *et al.* reported a series of 26 patients with compression myelopathy due to thoracic OPLL or ossification of the ligamentum flavum (OLF).⁶ They found 13 patients (50%) with associated ankylosing spinal hyperostosis, a condition also known as Forestier and Rotes-Querol disease¹³ or DISH.⁵ However, no patient in their series had associated with ankylosing spondylitis.

OPLL in the cervical region is commonly male dominant (2:1) in the sixth decade.^{1–3} However, it has been noted that OPLL at the thoracic level occurs more frequently in females than in males, and the reason behind this is unclear.^{1,2} It has been reported that the highest incidence of OPLL in the cervical spine was found at levels C5, C4 and C6. In addition, the fourth to the sixth thoracic levels have been the more common sites when the thoracic spine is involved.^{1,2,6}

The findings of sacroiliac joint involvement including erosion and sclerosis in this patient are character-

istic of AS. The bridging syndesmophytes in the lumbar and lower thoracic region (a bamboo spine) and apophyseal joint bony ankylosis are also pathognomonic to AS.^{7,19} The patient's history and physical examination provided no evidence of other seronegative arthropathies such as Reiter's syndrome, psoriasis or ulcerative colitis. Resnick *et al.* proposed three strict radiographic features of the spine as a prerequisite for diagnosis of DISH.⁵ These criteria are (1) absence of facet joint bony ankylosis and sacroiliac joint erosion, sclerosis, or intra-articular osseous fusion; (2) absence of extensive radiographic changes of degenerative disc disease; and (3) the flowing calcification and ossification along the anterolateral aspect should involve at least four contiguous vertebral bodies. Indeed, sacroiliac joint abnormalities, like osteophytes, osteoarthritis, do occur with DISH, while erosion or intra-articular bony ankylosis do not. Therefore, the diagnosis of AS in this patient was appropriate.

OPLL was noticeable in lateral views of the spine as an abnormal radiopacity along the posterior borders of the vertebral bodies. The ossification could be one of three types: continuous, segmental, mixed or localized.^{1–3} In this patient, the thoracic OPLL was of the continuous type, with a rod-shaped configuration involving T4–T6 (Figure 3). Generally, as not all OPLL can be seen by plain film without a strict lateral view, the posterior borders of the vertebral bodies might be mistaken to be ossification.^{1,2} In the upper thoracic levels, OPLL can also be masked by the massive shadows of the shoulder girdles. Thus a tomogram or CT scan becomes of great help in diagnosing such a condition. Clinically, recognition of the MRI features of OPLL as a low signal at T1- and T2-weight images has become more important. MRI is also invaluable in evaluating spinal cord compression.⁴ Thoracic compression myelopathy can be caused by OPLL or OLF, and both OPLL and OLF might exist together.^{6,14} A careful imaging evaluation of the spine can identify these two disease entities; however, there was no OLF noted in our patient.

The management of cervical OPLL can be conservative or can involve surgical treatment.^{1–4} Nonsurgical management reports give a 70% success rate. Successful treatment includes alleviating repeated impact or fraction on the spinal cord since OPLL is not just caused by a mechanical problem. Thus, only after conservative treatment fails, is surgery recommended. In the cervical region, the ideal surgical approach is debatable.^{4,15,16} With lesions involving one or two segments, an anterior approach with bone fusion is better. If the cervical OPLL involves more than three segments, a posterior approach with an expansive laminoplasty would be an alternative technique.^{1–4,16} Would this also be the case with the thoracic spine? One essential consideration is that the thoracic spine is in a more kyphotic alignment compared to the cervical spine, and the kyphosis of the thoracic spine will cause the cord to be draped

over the anterior impingement more so than in the cervical spine. If the cord is compressed significantly in the thoracic spine, it was reported that anterior decompression and fusion would be the best approach if the involving segments were less than three.¹⁰ Yonenobu *et al.* found that laminectomy was not always successful on such patients, and favored bilateral anterior surgery to excise the OPLL.⁶ In our patient, with the involvement from T4 to T6 levels and without significant thoracic kyphosis, a laminectomy from T4 to T6 was undertaken. At the second year follow-up examination, the patient showed good recovery from the neurological impairment. She had resumed her normal daily activities and walked freely without any support. The only complaints noted were a mild upper back soreness and occasional pains in the left thigh.

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