P. N. M. Tyrrell
V. N. Cassar-Pullicino
I. W. McCall

Ossification of the posterior longitudinal ligament in Down's syndrome

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P. N. M. Tyrrell (✉)
V. N. Cassar-Pullicino · I. W. McCall
Department of Diagnostic Imaging,
The Institute of Orthopaedics,
The Robert Jones and Agnes Hunt
Orthopaedic and District Hospital,
Oswestry, Shropshire SY10 7 AG, UK
Fax: +44-1691-404057

Abstract Cervical myelopathy in patients with Down's syndrome is not uncommonly the result of atlanto-axial instability, a condition that is caused by ligamentous laxity and which may be associated with congenital osseous anomalies at the occipito-atlanto-axial axis. Ossification of the posterior longitudinal ligament (OPLL) is well described, particularly in the Japanese population, and

may be associated with diffuse idiopathic skeletal hyperostosis. This is the first case, to our knowledge, of OPLL occurring in a person with Down's syndrome presenting with myelopathy.

Key words Posterior longitudinal ligament · Ossification · Down's syndrome · MRI

Case report

A 47-year-old man with Down's syndrome presented with a 3-month history of progressive lower limb weakness. There were no upper limb symptoms. Bladder and bowel function were normal. He gave a history of periodic severe respiratory tract infections, and he had gout, which was managed with allopurinol. There was no other past medical history of note.

Neurological examination revealed reduced power of both lower limbs (grade 4/5) affecting all muscle groups. There was a diminished sensation to pin prick, reflexes were brisk and there was a positive Babinski sign bilaterally. A mild reduction of power (grade 4+/5) in the upper limbs was detected, but there was no sensory deficit. Clinical examination revealed no other abnormalities apart from the features of Down's syndrome.

Radiography of the cervical spine demonstrated ossification of the posterior longitudinal ligament from C3 to C7 (Fig. 1). Flexion and extension views revealed a minor degree of atlanto-axial instability, with movement between flexion and extension of 1 mm and a maximum atlanto-axial distance of 3 mm on extension. Mild degenerative changes were also present.

An MRI study of the cervical spine showed diffuse low signal in the anterior aspect of the spinal canal, extending from C3 to C7 (Fig. 2). This was associated with marked anterior compression of the spinal cord and compromise of the spinal canal to approximately 65% of its normal calibre (Fig. 3). Posterior encroachment of the spinal canal at the C4/C5 level, and less so at the C5/C6 level, contributed further cord compression. Increased signal intensity within an atrophic cord at the sites of maximal narrowing was also present (Fig. 2b). The atlanto-axial region appeared unre-

markable. Degenerative changes corresponded to those seen on the radiograph, but there was no evidence of disc herniation.

MR imaging of the thoracic and lumbar spine revealed a grade 1 degenerative spondylolisthesis at the L5/S1 level. There was no evidence of disc prolapse. There was no further evidence of ligamentous ossification.

CT of the cervical spine confirmed abundant ossification of the posterior longitudinal ligament, causing marked compression of the spinal cord anteriorly (Fig. 3).

Discussion

Ossification of the posterior longitudinal ligament (OPLL) is well described [1, 5] and is particularly common in the Oriental population, with a reported incidence of 3.2% [5]. The incidence in Caucasians has been quoted as 0.2% [11]. The condition may be entirely asymptomatic and only be discovered incidentally or it may present with a variety of neurological disturbances. These may be non-specific or, more frequently, may indicate a compressive myelopathy [6]. Ono and co-workers also identified a direct correlation between the thickness of the ossified PLL and the presence of neurological manifestations. Cord signs usually occurred when there was a 60% compromise of the spinal canal [6]. Different patterns of OPLL have been described [2]; in this case the continuous

Fig.1 Lateral radiograph of the cervical spine showing ossification of the posterior longitudinal ligament from C3 to C7 (arrow). There are widespread degenerative changes. A small bony projection antero-inferior to C1 is also present

Fig.2 **A** Sagittal T1-weighted MR image of the cervical spine demonstrating diffuse heterogeneous low signal along the posterior aspect of the vertebral bodies from C3 to C7. **B** Sagittal T2-weighted gradient echo MR image of the cervical spine showing diffuse low signal along the posterior aspect of vertebral bodies from C3 to C7. This is causing compression and atrophy of the cord, maximal at the C4/C5 level, with intrinsic high signal of the cord

Fig.3 Axial CT image at the level of C4 confirms the ossified PLL. The canal is narrowed to 65% of its calibre

type was present, probably indicating ossification of both the superficial and deep fibres of the PLL.

Although florid OPLL will be readily visible on radiography, more subtle ossification may be overlooked unless specifically sought. Difficulties in MR diagnosis can arise from localised calcification/ossification and from blood or cerebro-spinal fluid flow artefacts [12, 13]. CT allows the definitive diagnosis on account of the superb bone detail obtainable.

The aetiology of OPLL is unknown, but it has been frequently described in association with diffuse idiopathic skeletal hyperostosis, suggesting a common pathogenesis for these conditions [3, 10]. An association with diabetes and localised intervertebral disc abnormalities has also been proposed [9]. To our knowledge, this is the first reported case of OPLL in a patient with Down's syndrome.

It is possible that the history of gout may also be an independent associated factor, but again no previous known association with gout has been described.

Atlanto-axial instability has been reported in 13.1% of persons with Down's syndrome, with symptoms being present in 1.5% [7]. As a consequence, radiographs are commonly requested in patients with Down's syndrome. Multiple congenital anomalies at the C1/C2 junction are described in these patients, including atlanto-axial subluxation and os odontoideum [4, 8]. Symptoms of myelopathy in patients with Down's syndrome are more usually due to atlanto-axial instability, a condition that may arise from ligamentous laxity at this level. Lateral flexion/extension radiographs will readily identify this instability and also an os odontoideum, but other abnormalities may only be visualised with accuracy on CT and may require

sagittal and coronal reconstructions to identify them precisely. CT will invariably be more accurate than MRI in this regard because of its better definition of the osseous anatomy.

The incidence of OPLL increases with age [5]. The life expectancy of people with Down's syndrome has increased, and if it is true that OPLL is associated with

Down's syndrome, rather than being an incidental finding, one may expect to see more cases in the future.

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