Ossification of the thoracic ligamentum flavum in an achondroplastic patient: a case report

K Suzuki, M Kanamori, M Nobukiyo
Department of Orthopaedic Surgery, University of Toyama, Toyama, Japan

ABSTRACT

We report a case of spinal stenosis with ossification of the thoracic ligamentum flavum in a 53-year-old man with achondroplasia. Neurological signs indicated flaccid paralysis below L1, and the patient was unable to walk. Ossification of the ligamentum flavum was observed at T4/5 and T9 to T12, compressing the thecal sac. Laminectomy of T9 to L1 was performed. At one-year follow-up, the patient was able to walk with one elbow crutch.

Key words: achondroplasia; laminectomy; ligamentum flavum; ossification, heterotopic; spinal stenosis

INTRODUCTION

Achondroplasia is an autosomal dominant disorder resulting from a characteristic mutation in the gene encoding fibroblast growth factor receptor 3 (FGFR3),1–3 with an incidence of approximately one in 10 000 live-born infants. It is characterised by rhizomelic shortness of the limbs and spinal stenosis, secondary to abnormalities of endochondral ossification. We report a rare case of spinal stenosis with ossification of the thoracic ligamentum flavum in an achondroplastic patient.

CASE REPORT

In March 2003, a 53-year-old achondroplastic man presented with a 15-month history of gradual weakness in both legs and gait disturbance. In 1990, he had undergone a laminectomy of L1 to L5 for weakness of both legs in another hospital.

He was unable to walk. His neurological signs indicated flaccid paralysis below L1. According to the Daniels and Worthingham’s Muscle Testing,4 the motor power of iliopsoas, hamstrings, tibialis anterior, and gastrocnemius were 3, 4, 1, and 0 out
of 5, respectively. Deep tendon reflexes of both legs were absent; hyporeflexia was likely secondary to the lumbar stenosis, complicated by thoracic cord compression. Bilateral hypoesthesia and hypoalgesia were observed below L1.

Anteroposterior and lateral radiographs showed a decrease in the interpedicular distance in the thoracolumbar spine and a wedged deformity and kyphosis, with ossification of the ligamentum flavum at T10 to T12 (Fig. 1). Sagittal T2-weighted magnetic resonance images showed a dorsally compressed thecal sac at T10/11, conus medullaris at L1/2, and stenosis at T10 to L5 (Fig. 2). A myelograph showed severe stenosis at T9 to T12 (Fig. 3). Computed tomographic scans showed ossification of the ligamentum flavum at T4/5, T9 to T12, and marked compression of the thecal sac at T10/11 (Fig. 4), which was considered the major cause of the symptomatic paralysis.

Ossification of the ligamentum flavum was removed using laminectomy of T9 to L1 (Fig. 5). Histology showed the presence of degenerative ligamentum flavum, chondroid tissue–containing chondrocyte-like cells, and mature bone, suggestive of endochondral ossification (Fig. 6).

The patient was able to stand using parallel bars at 3 weeks, to walk with a walker at 8 weeks, and to walk with one cane at 15 weeks. His motor power gradually recovered to 4 or 5, with a considerable decrease in bilateral leg numbness. At one-year follow-up, the patient was able to walk with one elbow crutch.

**DISCUSSION**

In achondroplasia, symptoms of cauda equina usually
manifest in adulthood; about 25% of patients require surgical treatment.\(^5\) Achondroplasia is characterised by spinal stenosis, disc degeneration, exaggerated lumbar lordosis, hypertrophy of the ligamentum flavum and bony spurs, thickened laminae and facet joints, and neurological complications such as hydrocephalus and upper cervical cord compression.\(^6–8\) Abnormalities of endochondral ossification result from mutation of FGFR3 and premature synostosis of the ossification centres of the vertebral body and posterior arch. FGFR3 is a negative regulator and the mutation evokes uncontrolled stimulation of the receptor, leading to bone growth inhibition.

Ossification of the ligamentum flavum occurs

### Table

<table>
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<tr>
<th>Studies</th>
<th>Sex/age (years)</th>
<th>Levels</th>
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<td>Takano et al.(^9) 1987</td>
<td>M/36</td>
<td>T4–L5</td>
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Figure 4  Computed tomographic scans showing ossification of the ligamentum flavum at T4/5, T9/10, T10/11, and T11/12. The thecal sac is markedly compressed at T10/11 (arrow).

Figure 5  Laminectomy of T9 to L1 showing spinal stenosis and ligamentum flavum hypertrophy. Ossification of the ligamentum flavum compresses and adheres to the dura mater at T10/11 and T11/12 (arrows).

Figure 6  Histology of the ligamentum flavum showing the presence of degenerative ligamentum flavum (black arrow), mature bone (arrowhead), and chondral cells (white arrow), suggestive of endochondral ossification (H&E, x200).
through endochondral ossification; the mechanism in achondroplastic patients is unclear and the incidence is rare. Only 6 such cases have been reported (Table9–13; all were Japanese and they were younger (20 to 53 years old) than those with conventional achondroplastic spinal stenosis.

In patients with achondroplasia, congenital spinal stenosis is not severe enough in itself to cause neurological deterioration. Neurological deterioration is due to additional developmental factors, such as thickened laminae, ligamentum, and bony spurs and exaggerated lumbar lordosis. The anteroposterior spinal canal diameter is not predictive of clinical symptoms of lumbar spinal stenosis.14 Patients with anteroposterior spinal canal diameters of less than 13 mm have been reported to score significantly higher on the Pain Disability Index.15

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REFERENCES