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Thoracic spinal cord compression caused by hypophosphataemic rickets: a case report and review of the world literature

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Abstract  Vitamin D resistant hypophosphataemic rickets is a rare cause of spinal cord compression. The compression is caused by a combination of thickening of the laminae and calcification of the ligamentum flavum. Modern imaging techniques including CT and MRI provide excellent detail of both the level and degree of compression. MRI is particularly useful for examining the rest of the spinal cord for areas of impending compression and for post-operative follow-up. With careful surgical decompression a full neurological recovery can be achieved.

Key words  Hyperostosis • Rickets • Spinal cord compression

Introduction

Hypophosphataemic vitamin D resistant rickets can lead to spinal stenosis and spinal cord compression. The compression is caused by a combination of thickening of the laminae and calcification of the ligamentum flavum. There have been 11 previously reported cases in the world literature. We present the case of a 49-year-old woman who developed thoracic compression at the T7 level with a motor and sensory deficit. The stenosis was identified using MRI and CT. Decompressive laminectomy was performed and following surgery the motor and sensory functions recovered rapidly and fully.

It has previously been suggested that medical treatment for rickets with vitamin D may have a deleterious effect and help cause the stenosis [2, 4]. Our patient did not receive medical treatment, and we feel that the stenosis is caused by the disease itself, independent of drug therapy.

Case report

A 49-year-old woman presented to the rheumatology out-patient-clinic in August 1994 with a several-year history of pain and stiffness affecting her back, shoulders, hips, knees and ankles. Apart from a slight increase in weight she was well. She had been told that she had suffered from rickets as a child, but could not recall any details or treatment for this. There was no family history of rickets. She smoked 20 cigarettes a day and took Co-proxamol but no other tablets.

On examination, she was short – 4‘8” (1.42 m) – with a weight of 57.1 kg and had bilateral bowed tibiae. She had a reduced range of movement at the cervical, thoracic and lumbar spine, and at the hips and knees.

Radiographs of her pelvis, lumbar spine and shoulders showed widespread changes of metabolic bone disease consistent with hypophosphataemic rickets.

Five months after her initial clinic attendance, she was admitted with increasing right hip pain and numbness of both legs. She had been suffering from increasing symptoms of spinal stenosis, with back and bilateral leg pain after walking 100 yards (91.5 m) which improved on bending forwards or sitting down. She had a sensory deficit in both lower limbs and reduced power of the right hip (MRC grade 4), but had otherwise normal power and reflexes in the legs. Serum calcium was 2.24 mmol/l (normal = 2.20–2.65 mmol/l), serum phosphate was 0.82 mmol/l (normal = 0.80–1.40 mmol/l), alkaline phosphatase was 515 U/l (normal = 70–350 U/l) and C-reactive protein was 6 mg/l (normal = 0–10 mg/l). An MRI scan revealed bony thickening of the laminae and ligamentum flavum, causing cord compression at the T7 level and to a lesser extent at T5, T6 and T9 (Figs. 1, 2). A CT scan confirmed these findings (Fig. 3).

Clinically she deteriorated. She could only walk 20 yards (18.3 m) with a Zimmer frame or stand for 5 min, but sitting time was unlimited. She lost sensation to pinprick, light touch and vibration below the T7 level. She had a sensory deficit in both lower limbs and reduced power of the right hip (MRC grade 4), but had otherwise normal power and reflexes in the legs. Serum calcium was 2.24 mmol/l (normal = 2.20–2.65 mmol/l), serum phosphate was 0.82 mmol/l (normal = 0.80–1.40 mmol/l), alkaline phosphatase was 515 U/l (normal = 70–350 U/l) and C-reactive protein was 6 mg/l (normal = 0–10 mg/l). An MRI scan revealed bony thickening of the laminae and ligamentum flavum, causing cord compression at the T7 level and to a lesser extent at T5, T6 and T9 (Figs. 1, 2). A CT scan confirmed these findings (Fig. 3).

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Decompressive laminectomy was performed from T6-T10. The laminae and calcified ligamentum flavum were grossly thickened to approximately 3 cm and very hard, necessitating the careful use of power burrs. The bone was solid and avascular and there was very little bleeding. The dura was not calcified and was not adherent to the ligamentum flavum. Full decompression was achieved without damaging the dura. Following surgery she experienced a rapid and full recovery of sensory and motor function and was discharged home after 5 days. At review 3 months after surgery, she had no residual neurological deficit and an unlimited walking distance.

**Discussion**

Eleven cases of spinal stenosis leading to cord compression caused by vitamin D resistant hypophosphataemic rickets have been previously reported [1–5, 7]. The compression is caused by a combination of bony thickening due to increased periosteal calcification and calcification of the ligamentum flavum. It has previously been noted that 69% of patients with hypophosphataemic rickets develop exuberant calcification of tendon and ligament insertions and of joint capsules [6]. Fortunately, this has not often caused spinal stenosis.

Including our patient, there have been eight men and four women affected. The age at which cord compression developed ranged from 24 to 60 years (mean age 47.9 years). The compression affected the thoracic region in nine patients and the cervical region in five. Compression occurred at both the cervical and thoracic levels in two patients. The cases date back to 1966 and the early cases were diagnosed by myelography, which demonstrated complete arrest of the contrast medium. More recently, both CT and MR imaging techniques have been used. Bussière et al. [2] suggests that MRI is the investigation modality of choice as it explores the whole spine and can pinpoint areas of stenosis, which may be multiple.

The only treatment available is decompressive laminectomy, which was performed in 11 of the 12 patients. The 12th patient became depressed and attempted suicide before surgery could be arranged. Although her suicide attempt failed, she later died from massive bleeding from a gastric ulcer (it is not known whether this was drug related). Four of the 11 patients who underwent surgery, including our patient, made full recoveries, 3 made partial recoveries, 1 remained unchanged and 3 patients neurologically deteriorated after surgery.

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