An Unusual Cause of Paraparesis in a Patient on Chronic Steroid Therapy

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Abstract

Background/Objective: Spinal epidural lipomatosis is the excessive deposition of unencapsulated fat in the epidural space. This is a rare disorder often associated with high levels of endogenous steroids or the administration of exogenous steroids.

Case Description: A 32-year-old man with congenital kyphosis treated with prednisolone daily for 5 months for interstitial lung disease developed compressive myelopathy.

Findings: Magnetic resonance imaging showed congenital kyphosis along with epidural lipomatosis compressing the cord. Cessation of steroid therapy was associated with improvement in the symptoms.

Conclusions: Spinal epidural lipomatosis is a rare side effect of chronic steroid therapy that may occur with relatively short-term, low-dose regimens. In patients with congenital vertebral anomalies, spinal fat deposition may worsen the neurological status in an already compromised cord. Discontinuation of steroid therapy is beneficial; some patients may require surgical intervention for decompression.

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INTRODUCTION

Spinal epidural lipomatosis (SEL) is a state of pathological fat tissue overgrowth in the vertebral canal. It is a rare and dangerous complication of chronic steroid therapy that may lead to back pain, radiculopathy, or paraparesis. Cases have been reported in patients with morbid obesity, Cushing syndrome, and hypothyroidism. This condition should be suspected in any patients presenting with back pain and neurological symptoms in a clinical setting of chronic steroid therapy (1,2). This is a report of such a case of SEL.

CASE REPORT

A 32-year-old man presented with a history of progressive increase in weight gain, back pain, and numbness and weakness of both lower limbs for 4 weeks. He also complained of hesitancy in passing urine for 1

week. His symptoms started gradually, and initially he ignored the symptoms until he developed difficulty in walking and passing urine. He denies any history of trauma, back pain, or fever, but he has had dorsal kyphosis since birth. He was diagnosed with idiopathic interstitial lung disease 5 months ago and was started on 40 mg prednisolone/d, reduced gradually to 10 mg/d over the past 5 months on improvement of chest symptoms.

On examination, he had features of Cushing syndrome, with moon facies, truncal obesity, and thoracic kyphoscoliosis (present since birth). Chest examination revealed late inspiratory crepitations, and abdominal examination was unremarkable except for obesity and purple striae. Neurological examination revealed increased tone in both lower limbs along with preserved muscle mass. Power was grade 3 (Medical Research Council grading) in all the groups of muscle in both lower limbs along with hyperreflexia at the knee and ankle. Babinski sign was positive in both lower limbs. Upper limb examinations were normal. Sensory examination revealed impairment of all sensory modalities below T11.

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Figure 1. (A) Sagittal T₂-weighted MR image shows wedge-shaped T9 vertebrae resulting in kyphotic angulation of the spine. The cord is stretched over the vertebrae and compressed because of prominent epidural fat. (B) Sagittal T₁-weighted image shows cord hyperintensity at the T9–T10 level.

This patient was diagnosed clinically with compressive myelopathy and studied further. Investigation revealed normal hematological and biochemical parameters including serum creatine phosphokinase. Radiograph of dorsal spine revealed severe dorsal kyphosis because of a congenital vertebral deformity but no evidence of obvious vertebral fracture. Magnetic resonance imaging (MRI) of the spine (Figures 1A and B and 2) revealed high-intensity fat within the spinal epidural canal compressing and indenting the thecal sac (T8–T11), consistent with SEL with vertebral deformity. Prednisolone was tapered and stopped gradually, and the patient was started on azathioprine 100 mg/d as a steroid-sparing measure and for pulmonary fibrosis. An option of surgical decompression was also considered, but the patient refused any surgical intervention. After 6 weeks, the patient was able to walk but was still having weakness and a spastic gait, but no urinary symptoms.

DISCUSSION

Centripetal fat deposition in the region of trunk, neck, and faces are characteristic of Cushing syndrome. Less commonly deposition occurs in the mediastinum and popliteal fossae, and more rarely, in epidural spaces, causing local pain and neurological deficits (3). SEL has been reported in a number of non-steroid-related cases such as hypothyroidism and obesity and also in some idiopathic cases. It seems to occur more commonly in men than women, with 75% of reported cases involving male patients (4,5). Although it is seen mostly in patients taking high doses of glucocorticoids for various reasons, it has also been reported on steroid doses as low as 15 mg prednisolone/d for as short a duration as 4 months (6,7).

Figure 2. Axial T₁-weighted image at T9 level reveals absence of posterior elements, confirming the congenital nature of the deformity. The cord is compressed because of prominent epidural fat that is contiguous with hypertrophied fatty tissue in the paraspinal region.

Thoracic involvement is the most common, followed by lumbar involvement. Cervical SEL has never been reported. MRI is the investigation of choice. Epidural adipose tissue that has a thickness greater than 7 mm has been reported to be the diagnostic criterion for SEL (5,8).

In our patient, SEL developed after 5 months of steroid therapy and on a dose of 10 mg/d. This early development and with low-dose therapy could be attributed to preexisting compromise of the spinal canal by congenital kyphosis caused by vertebral deformity. Why there is a preferential deposition of fat at this site is not clear. Conservative therapy, including weaning of patients from steroids and weight loss, has been reported to be very successful in patients with SEL (5,9). Weight loss has been reported to be very successful in patients with SEL in whom obesity is thought to be the cause of the adipose hypertrophy. Surgical intervention, with decompressive laminectomy and resection of epidural adipose tissue, has reportedly been successful in a large percentage of patients (10). It seems that a majority of these patients experience improvement or resolution of their neurological symptoms after surgical intervention (6).

In our case, the congenital anomaly resulted in kyphotic angulation of the spine, because the dural sac was stretched in the anterior portion of the cord with relative prominent epidural fat in the posterior portion of the cord, which was continuous with paraspinal fatty

tissue because of the absence of posterior elements. The steroid treatment resulted in hypertrophy of the fatty tissue and resulted in compression of cord. This was probably precipitated because of prominent posterior epidural fat and preexisting stretching of the cord caused by kyphosis.

This case report exemplifies the importance of SEL in chronic steroid therapy in patients with congenital vertebral anomalies, which may worsen neurological status in an already compromised cord.

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