Idiopathic Thoracic Spinal Epidural Lipomatosis With Relapsing And Remitting Symptoms

A Karageorgos, A Petsanas, I Gelalis, N Gkantaifis, D Aretha

Citation

Abstract

Background context: Spinal epidural lipomatosis (SEL) is a disease consisting of an excessive deposition of normal adipose tissue in the spinal canal. It is a quite uncommon disease, which can end to severe neurological deterioration. Method: This report presents a patient with idiopathic SEL with relapsing and remitting course. After a 16-month period with clinical improvement, following conservative treatment, the patient was presented with rapid neurological deterioration, consisting in spastic paraparesis. Such a fluctuating presentation is rare in patients with SEL, and mimics a demyelination disease. Multilevel laminectomy from Th6 to Th10, and resection of thick non-capsulated fatty tissue, was performed. Results: Postoperatively the patient regained gradually lower extremity power. Fifteen days postoperatively he developed an epidural haematoma, due to treatment with high doses of anticoagulants for unstable angina. He presented with rapid neurologic deterioration and operated on for haematoma evacuation. The symptoms resolved the next 24 hours and on follow-up six months later he was able to walk and climb stairs without assistance. Conclusions: These patients have high operative risks due to accompanied medical problems. We suggest conservative treatment for patients with minor symptoms, reserving surgical decompression for cases with progressive neurological deterioration.

INTRODUCTION

Spinal epidural lipomatosis (SEL) first described in 1975 (1), is a disease consisting of an excessive deposition of normal adipose tissue in the spinal canal, compressing the spinal cord. Most cases of SEL are associated with long-term use of exogenous steroids, but a number of non-exogenous steroid-related cases have been reported, associated with Cushing disease, hypothyroidism (2) and pituitary prolactinoma (3). Idiopathic SEL, is a rare entity (4), usually observed in obese patients (5).

We report a case of a 68-year-old male patient with SEL presenting with remitting and relapsing neurological symptoms for two years.

CASE REPORT

A 68-year-old man was presented to the outpatient clinic of Neurosurgery Department, complaining of claudication, which was progressively worsened over the last 2 years, with a walking distance of less than 50 meters. His medical history was significant for diabetes mellitus type II, congestive heart disease and peripheral vascular disease. The patient was on antiplatelet treatment with Salospir 325 mg once a day and had no history of steroid use.

On physical examination the patient weighted 91 kilograms and measured a height of 1.65 meters, with a Body Mass Index (BMI) of 33,4. On palpation he complained of mild tenderness throughout his thoracic spine and his range of motion was limited by pain. Examination revealed weakness of both lower extremities, increased deep tendon reflexes with clonus and Babinski sign and sensory level at pinprick below the Th6 dermatome. No bowel and bladder incontinence was reported.

MRI of the thoracic spine confirmed SEL, revealing an overgrowth of epidural fat in the spinal canal compressing dura from th6 to th10 spinal level in a width of 8-10mm.

Initially, conservative treatment was selected due to presented comorbidities. Conservative approach consisted of weight reduction and physical therapy, as progressive improvement was noted in the following months, although weight reduction was only 5 kilograms.

Sixteen months later, he was readmitted with progressively deteriorating spastic paraparesis and inability to walk. A
repeat MRI of the thoracic spine revealed the same findings (fig.1 & 2) and surgical treatment was advised.

**Figure 1**
Fig. 1: Axial T1-weighed image at Th8 level demonstrates an increase in epidural fat, with a sagittal thickness of 10mm.

**Figure 2**
Fig. 2: Sagittal T1-weighted image of the thoracic spine shows an increase in epidural fat from Th6 to Th10 levels.

A multilevel laminectomy of the thoracic spine (Th6 to Th10) was performed. The epidural space was filled by a thick non-capsulated fatty tissue compressing the dura. After complete resection of the fat, the thecal sac became pulsatile (fig.3). The histological examination confirmed normal adipose tissue.
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Figure 3
Fig. 3: Decompression of spinal cord after laminectomy and adipose tissue resection.

Postoperatively, weakness of both legs was remarkably improved. The patient was discharged four days after surgery walking on crutches. Ten days later he was admitted to the hospital for an unstable angina and was treated with high doses of anticoagulants. Two days later he developed an epidural hematoma of the thoracic spine with a progressive neurological deterioration and was operated urgently. The epidural hematoma was evacuated surgically and 24 hours postoperatively patient’s symptoms were resolved. The patient was followed-up for 2 years and was able to walk normally and climb stairs without any assistance.

DISCUSSION
Spinal epidural lipomatosis is an uncommon disease, in which the excess adipose tissue deposited around the thecal sac may cause back pain and neurological signs and symptoms.

The underlying pathological mechanism of SEL is unknown. The literature reveals several underlying conditions associated with SEL: exogenous steroid use, endogenous steroid excess, Cushing disease and an idiopathic group, with or without obesity.

The most common association is exogenous steroid use. It is well established that hypercorticolism leads to accumulation of adipose tissue in atypical distribution, as in face, neck, trunk and mediastinum (Cushing syndrome) (6). Hypertrophy of adipose tissue already present in the spinal canal is theorized to be the cause of SEL in certain cases of exogenous steroid use (7). It has been suggested that a dose of 30 mg/d, taken orally over several months, is required for the development of spinal lipomatosis (8).

Cushing disease from endogenous sources and other medical entities is another important group of underlying conditions for SEL.

Idiopathic SEL, usually observed in obese patients (5), is a third entity (4). Obesity is the second most common, after steroids, underlying condition of SEL, although some investigators question whether obesity plays a causal role in SEL or is merely a predisposing factor (9).

We believe that we must classify the entity of SEL in two major categories, according to underlying conditions, steroid related and non-steroid related (Table).

Figure 4
Table 1

<table>
<thead>
<tr>
<th>Steroid related</th>
<th>Non steroid related</th>
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</thead>
<tbody>
<tr>
<td>Exogenous steroid use</td>
<td>Endogenous steroid excess (Cushing syndrome)</td>
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Our patient had BMI of 33.4, but no other causative factor for epidural adipose tissue hypertrophy was identified.

There seems to be a strong male preponderance in idiopathic spinal epidural lipomatosis (10). The condition is most commonly located at Th6-Th8 levels of the thoracic spine and L4-L5 levels of the lumbosacral region. Thoracic involvement is observed in 58 - 61% of the patients and lumbar involvement in 39 - 42% (4). Patients with thoracic spinal epidural lipomatosis generally present at an earlier age than the lumbosacral group (average age 38.1 years versus 51.4 years). This might be explained by the narrower space of the thoracic spinal canal (11).

In our case the adipose tissue was located between Th6 and Th10. The patient was 68 year-old and his first symptoms appeared 2 years prior to the first examination.
Back pain is the most frequently reported symptom associated with SEL and often presents long before the neurologic symptoms. Lower extremity weakness is also a common complaint and appears to be slowly progressive in most cases. Sensory changes with numbness, paresthesias, or radicular symptoms are also common. Bowel and bladder incontinence are reported, but appear to be rare and late complaint. On physical examination, lower extremity weakness is the most common finding with decreased pin prick sensation and altered deep tendon reflexes also frequently occurring (5). Obviously, the symptoms are somewhat depended on the level of the canal compromise (spinal cord, conus medullaris, or cauda equina). Pressure of the spinal cord in the thoracic spine produces myelopathy, while pressure in the lumbar spine radicular effects.

Our patient suffered mainly from myelopathic claudication and lower extremity weakness. His presentation was unusual, thus he seemed to recover for 16 months after conservative treatment, although he had not sufficient weight reduction. This similar fluctuating course has been seen in 1 case in SEL (12), and in some cases of spinal epidural angiolipomatosis (13). The pathogenesis is probably multifactorial. Venous stasis with thrombosis has been suggested as precipitin factor. Such a clinical presentation may mimic a demyelization disease.

Spinal MRI is the diagnostic imaging modality of choice. High-signal intensity on T1-weighted images and an intermediate signal on T2-weighted images are characteristic of adipose tissue. Circumferential compression of the thecal sac, referred to as the ‘‘Y-sing’’, is pathognomic in lumbar axial imaging (14). Epidural adipose tissue that has a thickness greater than 7mm has been reported to be the diagnostic criterion for SEL (5). In our case MRI of the thoracic spine revealed a thickness of 8 to 10 mm of the epidural adipose tissue, from Th6 to Th10, which was diagnostic of SEL.

There are two treatment options for idiopathic SEL: conservative management with weight reduction, and surgical treatment by decompressive laminectomy and fat debulking (10). For weight reduction to be effective, a loss of 15kg or more is necessary (5). The indication for surgical decompression is determined by the patient’s neurological status. After debulking of the excess epidural fat, it is possible for the thecal sac to resume its normal configuration (5). Despite the good surgical results, it has been reported a 22% mortality rate in these patients within 1 year after surgical decompression, due to severe concomitant medical problems and comorbidities (15). Due to major health comorbidities of our patient, we first treated him conservatively. We operated him on, after the progressive neurological deterioration and the failure of conservative treatment, despite the known operative risks due to his medical problems. Post-operatively the patient presented immediate improvement in his neurological status. Although he developed an epidural haematoma that required evacuation, 2 years later at the follow up his neurological status was remarkably improved.

CONCLUSION

Fluctuating clinical course is extremely rare in SEL. Such pathology should be considered in the differential diagnosis of demyelinating diseases, in younger patients. In patients with minor symptoms and medical comorbidities, we suggest conservative treatment, reserving surgical decompression for cases with progressive neurological deterioration.

References

4. Stern JD, Quint DJ, Saweasy TA, Hoff JT. Spinal epidural lipomatosis: two new idiopathic cases and a review of the literature. Stere JD, Quint DJ, Saweasy TA, Hoff JT.
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