

## Case Report

# Idiopathic Thoracic Epidural Lipomatosis with Chest Pain

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Spinal epidural lipomatosis (SEL) is an overgrowth of the normally encapsulated adipose tissue in the epidural space around the spinal cord in the thoracic and lumbar spine causing compression of the neural components. Idiopathic SEL in non-obese patients is exceptional. Idiopathic SEL can result in thoracic myelopathy and lumbar radiculopathy. A thoracic radiculopathy due to idiopathic SEL has not been reported yet. We report a case of idiopathic SEL with intractable chest pain and paresthesia. We suggest that idiopathic SEL should be considered as a cause of chest pain.

**Key Words :** Spinal · Idiopathic · Epidural · Lipomatosis · Thoracic · Chest pain.

## INTRODUCTION

Spinal epidural lipomatosis (SEL) is an overgrowth of the normally encapsulated adipose tissue in the epidural space around the spinal cord in the thoracic and lumbar spine causing compression of the neural components<sup>5,12,13,25,26,32,34</sup>. In cases without definite predisposing factors, the term 'idiopathic SEL' has been used. Idiopathic SEL in non-obese patients is exceptional<sup>3,12,13,26</sup>.

According to previous reports, idiopathic SEL can result in thoracic myelopathy and lumbar radiculopathy<sup>2,4,8,9,13,19</sup>. To our knowledge, a thoracic radiculopathy due to idiopathic SEL has not been reported yet.

We report a case of chest pain caused by idiopathic thoracic SEL in a female patient who did not have any other recognized predisposing factors for this condition.

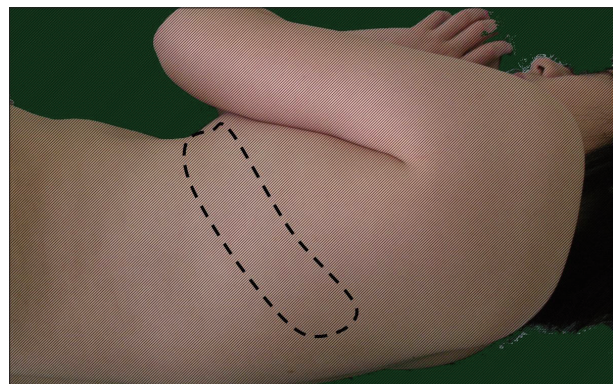
## CASE REPORT

A 16-year-old girl presented with a 2-month history of pain on her left chest. She reported that the pain spontaneously started around 2 months prior to admission and got worse over the two weeks. She could not do her usual daily living activities due to the intractable pain. She had no medical history for diabetes or steroid intake. On physical examination, her body weight was 55 kg and height was 155 cm, her body mass index

was 22.8 kg/m<sup>2</sup> which was within the normal range. Neurologic examination revealed paresthesia on the left side, at the level of the T5 and T6 dermatomes, without myelopathy, motor weakness or claudication (Fig. 1). Hyperesthesia and allodynia were not evident. Laboratory evaluation, including full blood count, electrolytes, creatinine, liver function test, vitamin B12 and folate, urine analysis and culture, thyroid function tests, adrenocorticotropic hormone, and cortisol, were all unremarkable.

Plain radiographs of the thoracic spine showed no abnormalities. Myelography computed tomography (CT) demonstrated a hypodense soft tissue mass within the posterior spinal canal and mainly in left vertebral foramen on T4/5 and 5/6 level (Fig. 2).

Magnetic resonance image (MRI) of the thoracic region presented a posterior compressing mass lesion of increased signal intensity on T1-weighted image (WI) and T2 WI in the epidural space, suggesting an epidural lipomatosis in the thoracic area.



**Fig. 1.** Photograph of patient in a lateral decubitus position showing left T5 and T6 dermatomes (dotted line) where she had the intractable pain with paresthesia.

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An extensive epidural fat deposition was about 6 mm in thickness, extended from T3 to T9 causing anterior displacement of the spinal cord and root compression in the left vertebral foramen on T4/5 and T5/6 levels (Fig. 3).

The patient's pain was refractory to medication. We did a diagnostic root block at the left vertebral foramina of T4/5 and T5/6 level. Her pain improved for about three hours but recurred. The presumptive diagnosis was a radiculopathy of left T5 and 6 nerve root due to SEL.

An operation for the chest pain with paresthesia was decided. We performed a lateral partial facetectomy and fat debulking on the left side of T4/5 and T5/6 levels (Fig. 4A). During the operation, a fatty mass crowded out of the vertebral foramen after removal of the ligament flavum (Fig. 4B). It appeared as fat tissue severely compressing the dorsal root ganglion (DRG). The compression was more remarkable at the T5/6 than T4/5 level. The fat tissue compressing the nerve root and DRG was removed (Fig. 4C).

Histopathological examination postoperatively demonstrated nodular proliferation of mature fat cells, consistent with lipomatosis (Fig. 5). The patient's intractable chest pain resolved completely and the paresthesia disappeared immediately. The patient was discharged by the 8th postoperative day. The patient had no recurrence of symptoms at 2 years after the operation.

## DISCUSSION

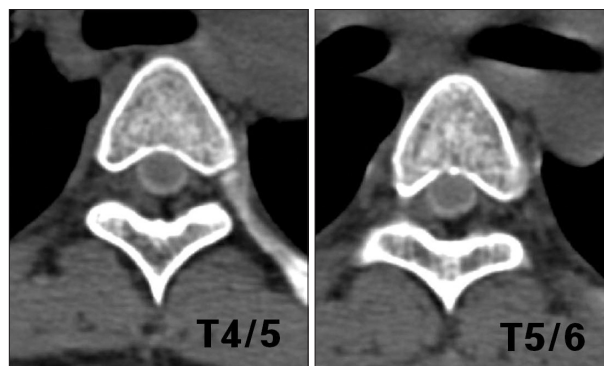
SEL is a condition wherein excess adipose tissue deposits around the thecal sac causing compression of neural structures<sup>2,3,5,7,8,12,17,18,24,31,32</sup>. In 1975, Lee et al.<sup>18</sup> reported the first case of symptomatic epidural cord compression secondary to SEL. It is well known that the main etiological factors of SEL are associated with endocrine dysfunction such as Cushing disease, hypothyroidism, obesity, iatrogenic steroid treatment for immune disorders, COPD and transplantation<sup>9,11,20,27</sup>. In approximately 75% of SEL, this may be secondary to exogenous steroid ad-

ministration for systemic diseases such as rheumatoid arthritis, systemic lupus erythematosus, dermatomyositis, chronic obstructive pulmonary disease, or following organ transplantation<sup>27</sup>. Obesity is the second most common associated category of SEL<sup>5,9,11</sup>.

Idiopathic SEL was first reported by Badami and Hinck<sup>3</sup> in 1982. Haddad, et al.<sup>12</sup> first hypothesized that idiopathic SEL was a byproduct of obesity, with the gradual overgrowth of epidural fat resulting in compression of the spinal cord and nerves. Thereafter, several researchers asserted that the term 'idiopathic' should be used solely to characterize cases of SEL that develop in non-obese patients, who were not receiving corticosteroid therapy, and without any other underlying SEL-related disease such as hypothyroidism or endogenous corticosteroid production<sup>4,6,9,19</sup>. About 16 cases of idiopathic SEL in non-obese patients were reported previously<sup>1,9</sup>. The underlying pathological mechanism of SEL is still unknown.

Our patient was neither receiving steroids nor had any kind of endocrinopathy. Moreover, she was not obese. For this reason, we had difficulty diagnosing idiopathic SEL as the cause of chest pain.

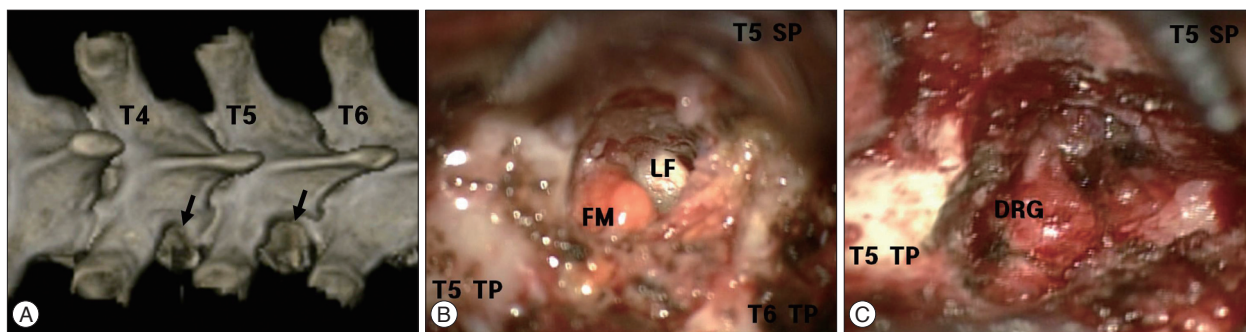
The majority of idiopathic SEL involve the mid-thoracic and



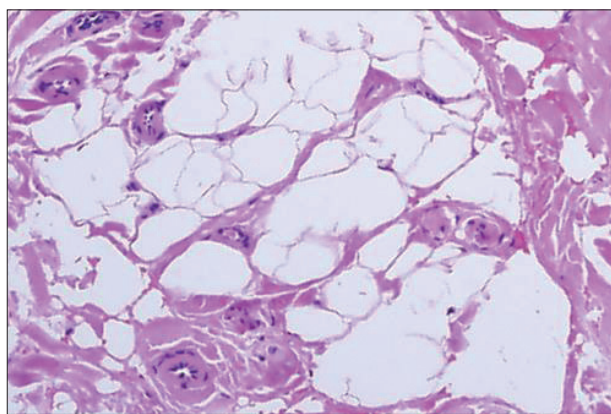
**Fig. 2.** Myelography computerized tomography (CT) demonstrating a hypodense soft tissue mass in the posterior spinal canal and mainly in the left vertebral foramen on T4/5 and T5/6 level.



**Fig. 3.** Thoracic spinal magnetic resonance imaging; Sagittal T2-WI (A), sagittal T1-WI (B), T4/5 axial T2-WI (C) and T5/6 axial T2-WI (D), showing a posterior epidural mass with mild compression of the dura extending from T3 to T9 segments with high signal intensity at T2-WI (A) and T1-WI (B) similar to that of subcutaneous adipose tissues. This epidural mass is extending into the left vertebral foramen on thoracic 4/5 (C), and T5/6 (D) levels. WI : weighted image.



**Fig. 4.** Postoperative 3D computerized tomography (A) and intra-operative photos (B and C). A : Lateral facetectomy was done at the left side of T4/5 and T5/6 levels. B : A fatty mass bulging out of the vertebral foramen after removal of LF. C : DRG has been decompressed after removal of surrounding fatty tissue. T5 TP : T5 transverse process, T6 TP : T6 transverse process, T6 SP : T6 spinous process, LF : ligament flavum, DRG : dorsal root ganglion, T4 : T4 vertebra, T5 : T5 vertebra, T6 : T6 vertebra.



**Fig. 5.** Histopathological examination demonstrates nodular proliferation of mature fat cells, consistent with lipomatosis (H&E x200).

lower lumbar vertebral levels, representing two peaks<sup>9,15</sup>. SEL causing neurological deficits occurs more frequently in the thoracic than the lumbar spine<sup>17,23</sup>. Patients may present with progressive and long standing complaints of pain, weakness, numbness, incontinence, ataxia, abnormal reflexes and rarely paralysis. According to Al-Khawaja et al.<sup>2</sup>, patients with idiopathic SEL involving the thoracic segments had myelopathy in 70% and paraplegia in 5% of patients. All of these symptoms were induced by compression of the spinal cord. In cases of lumbar SEL, symptoms associated with lumbar radiculopathy were reported but thoracic radiculopathy has not been reported<sup>1,4,8,9,30,34</sup>.

The mechanism of SEL induced neurologic symptom is mainly by direct compression of the adjacent nervous structures and epidural blood vessels resulting in venous engorgement which then compromise contribute to the evolution of myelopathic and radicular complaints<sup>1,30</sup>. We identified the direct compression by the lipomatous tissue and with venous engorgement around the DRG during the operation and believed that these findings induced the radiculopathy with intractable chest pain and paresthesia. The DRG is exquisitely sensitive to direct pressure and can generate prolonged neural discharges even after brief compression. The nerve root itself responds to this same stimulus only after it has become inflamed and sensitized<sup>14,33</sup>. These features of the DRG are thought to be the cause of intrac-

table pain in our case.

Symptoms and signs associated with SEL can be caused by a variety of diseases. Disc disease, thoracic intradural extramedullary tumor like meningioma and intradural lesions, like ependymoma, astrocytoma, or a syrinx were considered likely differentials prior to MRI<sup>30</sup>. Diagnosis is best based on MRI<sup>6,10,15</sup>. T1-WI differentiate epidural fat from dural content with a high degree of specificity and allow for measurement of adipose tissue thickness. Definite MRI finding of SEL is the pathognomonic appearance of fat on MRI; high signal intensity on T1-WI, intermediate signal intensity on T2-WI<sup>6,9,10,15</sup>. On CT scans, SEL presents as a homogenous hypodense epidural mass with a density between -80 and -160 HU<sup>24</sup>. Several grading system by MRI were reported but could not show correlation with clinical symptoms<sup>6,10,15,16,21</sup>. The mean sagittal thickness of the epidural fat in these studies was 4.6 mm with a normal range of 3 to 6 mm. In contrast, imaging in 6 patients with SEL revealed a mean thickness of 8 mm<sup>25</sup>.

MRI findings of our patient showed that the spinal cord was displaced anteriorly by a lipomatous tissue which was 6 mm in thickness and extended into the vertebral foramen around the DRG. We believe that the epidural fat with a 6 mm thickness was not enough to induced myelopathy.

The treatment of SEL ranges from conservative management to surgical excision. The treatment of SEL is a matter of debate, because many patients have been treated by simple weight reduction with good results<sup>7,22,24</sup>. However, this conservative treatment remains restricted to patients with mild neurological symptoms. Surgery is often necessary in cases wherein weight reduction failed to alleviate the symptoms or in severely symptomatic patients although the clinical suspicion for the presence of this particular neoplasia was not high<sup>4,15,19,26,28</sup>. A decompressive surgery with fat debulking should be considered when diet therapy proves unsuccessful or when the patient suffers acute neurological deterioration<sup>15</sup>. The prognosis of patients with idiopathic SEL after surgical management is favorable and no case of recurrence has been reported<sup>9</sup>.

Although our patient did not present with myelopathy, she was not able to do her usual activities and could not sleep well

because of intractable chest pain which was not controlled by medications. We chose the operation for symptomatic relief and decompressed just lateral part of facet joint at symptomatic level minimally. The outcome of the operation was good and there was no recurrence for 2 years.

Whether the epidural fat shows histological alterations compared with normal fat remains unclear. Further studies should focus on the histological entity of epidural fat, given that hypervascularization and bleeding within the epidural fat could be a risk factor for symptomatic manifestation of extradural lipomatous tissue<sup>28,29,31</sup>. Our patient demonstrated irregular fibroadipose tissue, nodules of mature fat cells with venous engorgement on histopathological examination and no hypervascularization and bleeding. Also, we suggested that the main cause of our patient's symptom was the direct compression of the DRG by the lipomatous tissue.

## CONCLUSION

We report a case of idiopathic SEL with intractable chest pain and paresthesia. We suggest that idiopathic SEL should be considered as a cause of chest pain.

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