Idiopathic Spinal Epidural Lipomatosis – Two Cases Report and Review of Literature

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Spinal epidural lipomatosis (SEL), an abnormal localized or tumor-like accumulation of fat in the epidural space, is an infrequent complication of chronic steroid usage and an uncommon cause of spinal cord compression. During the period of 1990 to 2006, we have two cases of medically heath SEL patients without history of steroid administration. Their initial clinical manifestations were low back pain, progressive lower extremities weakness, numbness, followed by rapid deterioration of neurogenic intermittent claudication. They were misdiagnosed and treated as degenerative spinal disease for a long time. Due to prominent neurological deficit, lumbar magnetic resonance image (MRI) was obtained and showed SEL. These 2 patients all underwent laminectomy and removal of epidural fat. Postoperatively, they both showed improvement. We reviewed the literature and discussed the current concept in the management of SEL. *(Chang Gung Med J 2009;32:662-7)*

Key words: lipomatosis, spinal

Spinal epidural lipomatosis (SEL) is a rare condition characterized by an excessive deposition of mature adipose tissue that grew in an infiltrative manner. It usually involved large portions of limb or the trunk, but rarely involved the intraspinal epidural space. Most of these patients are associated with the administration of exogenous steroids or endocrinopathies, such as, Cushing Syndrome and hypothyroidism. Idiopathic, is defined as, medically normal and without the history of administration of steroids. SEL is a very rare disease. We report two cases of idiopathic SEL in a medically health patient and review the literature. Moreover, we include their clinical courses, magnetic resonance image (MRI) features, and the surgical findings.

CASE REPORT

Case 1

A 56-year-old male without any underlying medical disease had suffered from sustained lower back pain in the past 3 years. Prior to his visit to our hospital, he had received treatment for degenerative spinal disease with non-steroid anti-inflammatory drugs. Nevertheless, he denied any exogenous steroid administration. A period of time passed and the symptoms had worsened despite the medicated treatment. Consequently, he came in to the outpatient department with progressive lower back pain associated with left leg weakness and numbness. He also stated that for the past three month he had been

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unable to walk long distances.

On physical examination, he demonstrated a good external physical appearance. The endocrine studies, including serum adrenocorticotropic hormone (ACTH), cortisol, triiodothyronine (T3), thyroxine (T4) and thyroid- stimulating hormone (TSH) levels were within normal limits. He weighed 74 kilograms with a height of 175 centimeters (body mass index = 24.1). The patient had no definite localized tenderness at any point on the back. He showed hyporeflexic bilateral extremities, remarkable left leg weakness but no sensory disturbance was identified. Radiography of the lumbar-sacral spine revealed mild degenerative changed. Due to a definite neurological deficit, MRI was arranged. The MRI revealed moderate bulging intervertebral discs at L3-4 and L4-5 level; abnormal hypertrophy of adipose tissue located at the anterior aspect of spinal canal extending from L3 to S1 level (Fig. 1A). The axial view image revealed overgrowth fat circumferential compression of the thecal sac, presented as typical "Y-sign" or "Stellate-sign" of the thecal sac (Fig. 1B).

An surgical procedure was carried out on the laminectomy from L3 to L5 and debulking of the epidural fat (Fig. 2). A disectomy was not performed on this patient because the spinal canal was relatively patent after debulking of the epidural fat. The patient recovered completely. On a 6 months follow-up, he was symptom-free and no other neurological sequela left.

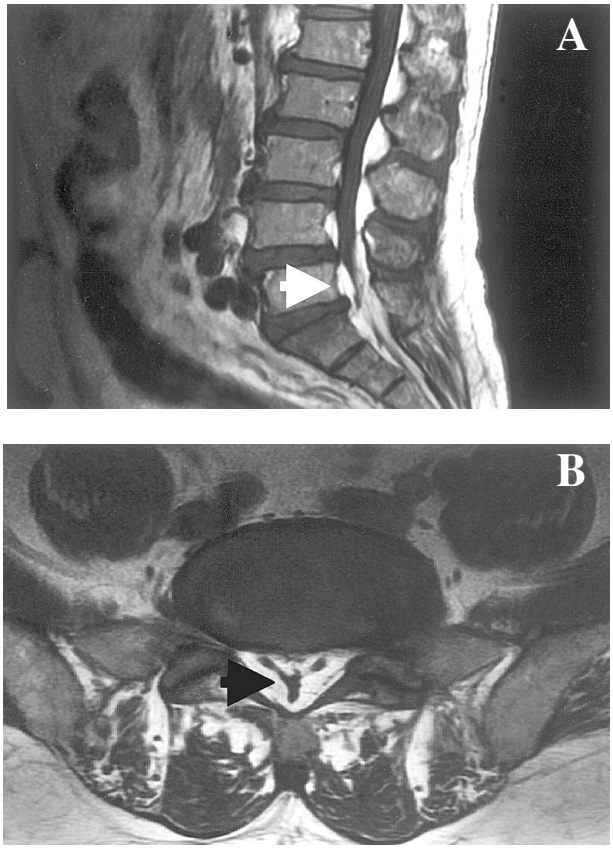

Fig. 1 T1-weighted MRI at lumbar-sacral level (Case 1) showed high intensity adipose tissue compression of the spinal nerve roots, lying at anterior aspect of thecal sac. (A) The T1-weighted sagittal image revealed the hypertrophy of adipose tissue deposition at the anterior aspect of spinal canal extending from L3 to S1 level (white arrow). L3-4, L4-5 buldging intervertebral disc was also demonstrated. (B) The T1-weighted axial image revealed the typical "Y" or "Stellate" configuration of the thecal sac (black arrow).

Case 2

A 44-year-old, otherwise healthy man presented to our out-patient department with the primary complaint of progressive lower back pain and left extremity weakness and numbness for several months. He stated that he had fallen down from a height many years ago but no neurological deficit had resulted. The lower back pain started 2 years to date of the interview with a progressive worsening and an inabiliy to walk for long distances.

He had a fine physique, 70 kilograms in weight and 173 centimeters in height (body mass index = 23.4), the basic endocrine studies, including ACTH, cortisol, T3, T4, and TSH levels were within normal limits. His neurological findings were remarkable for bilateral legs weakness and hyporeflexic bilateral lower extremities.

X-ray of the lumbar-sacral spine revealed a degenerative change of lumbar spine and mild L4-5 spondylolisthesis. An MRI was therefore arranged, which revealed moderate buldging intervertebral discs at L3-4 and L4-5 level. Furthermore, an overgrowth adipose tissue circumferential was found to compress the thecal sac at L3 to L5 level (Fig. 3A). The axial T1 weighted-images demonstrated an epidural fat deposition around the whole thecal sac, although no typical "Y-sign" was shown. The thickness of the fat was 8 milimeters (Fig. 3B).

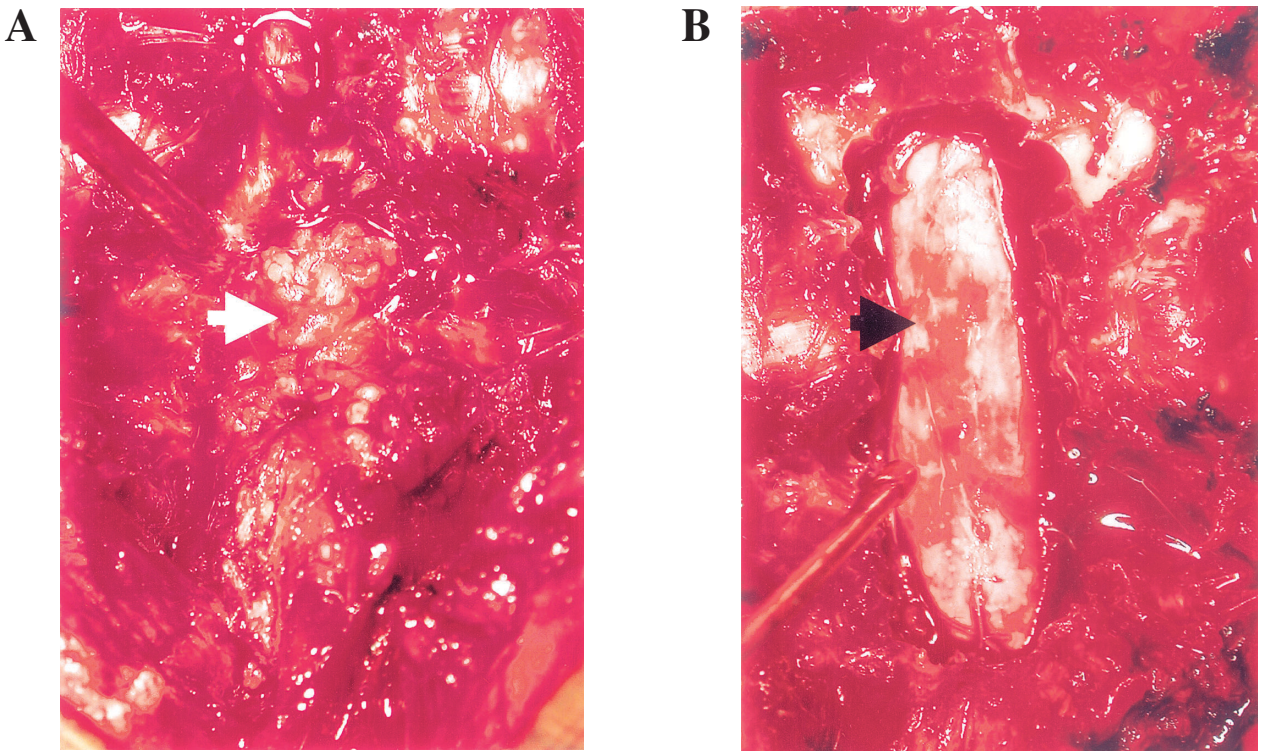

Fig. 2 The intra-operative finding of spinal epidural lipomatosis. (A) After removing the L3-5 laminae and ligamentum flavum, about 2 cm thick lobulated fat tissue accumulated over the epidural space with the dural sac compression (white arrow). (B) The mass was totally removed by suction and curettage easily because of the well demarcation between the mass and dural layer, and the dural sac was decompressed completely (black arrow).

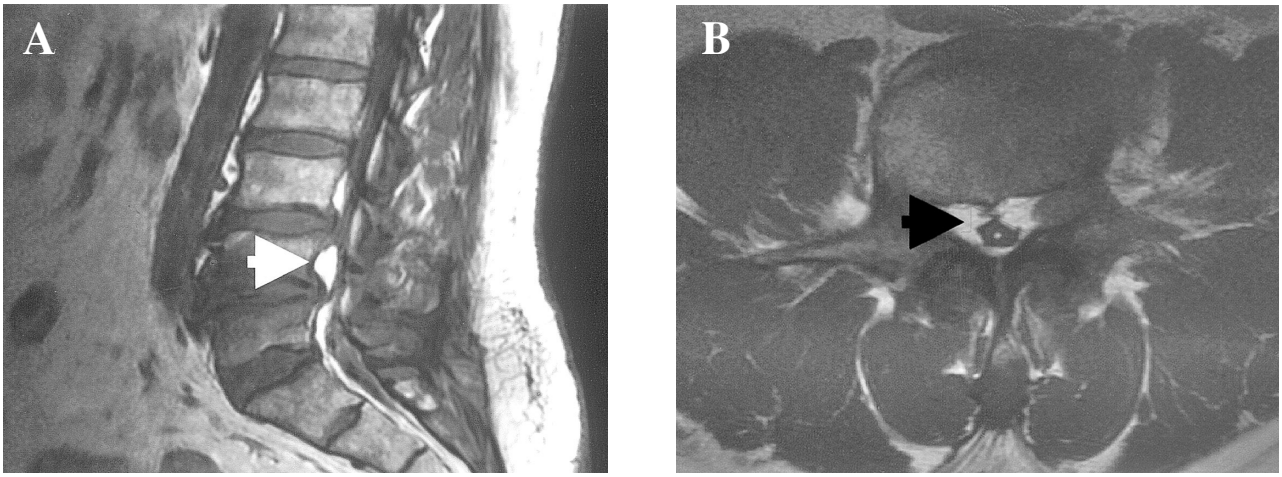

Fig. 3 T1-Weighted MRI showed an extradural high intensity area posterior to the thecal sac at L3-L5 region (Case 2). (A) The T1-weight image revealed moderate buldging intervertebral discs at L3-4 and L4-5 level. The overgrowth adipose tissue circumferential compression of the thecal sac at L3 to L5 level (white arrow). (B) The axial view of MR image showed mark spinal canal compression by overgrowth adipose tissue. The maximal fat thickness measure 8 mm (black arrow).

Consequently a surgical procedure was performed similar to Case 1 and the pathology was confirmed as epidural lipomatosis (Fig. 4). At a 12 month of follow-up, the short-term result was promising, he remained asymptomatic.

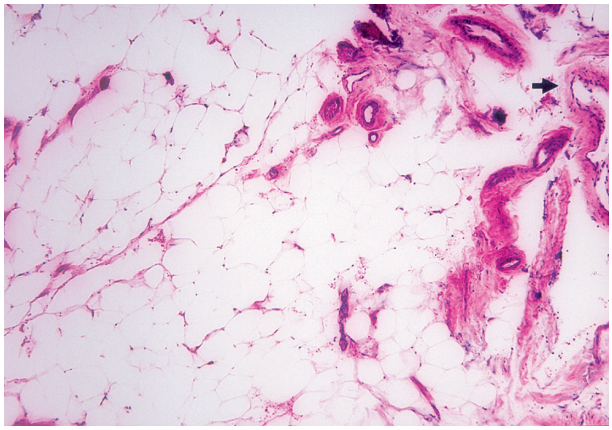

Fig. 4 Photomicrograph revealed the mature adipose tissue that grew in an infiltrative manner (H & E, 100). Epidural venous engorgement was demonstrated in pathological findings (black arrow).

DISCUSSION

SEL is a rare disease. It was first reported by Lee et al. in a post-renal transplant patient in 1975.[1] Subsequent reports on this disorder have been published.

SEL was defined as a pathological overgrowth of adipose tissue in epidural space and often caused dura impingement. In symptomatic SEL, the overgrowth of adipose tissue always led to encroachment on the spinal canal and compression of the spinal cord, conus medullaris, or cauda equine.

It is apparent that symptoms were dependent on the level of canal compromise. Thoracic levels produced myelopathy and lumbar levels resulted in radiculopathy. In most reported cases, including ours, the symptoms were gradual onset and similar to degenerative spinal stenosis. Commonly, the onset of symptoms were lower back pain, gradual with slow progression of lower extremities weakness and numbness. Mono- or polyradicular compression signs had been reported.[2] Both patients in our case also suffered from neurogenic intermittent claudication at the end of the clinical course.

SEL was male-predominance. Seventy five percents of the reported cases were male.[3,4] In Kawai's review,[5] the mean age was 42.3 years and the average body mass index was 29.0 (16.1 to 36.5). Thoracic involvement was most common, a measure of between 58% to 61%. Lumbar involvement was found in 39% to 42% (T6-T8 and L4-L5 were the most commonly involved levels), whereas, cervical SEL had never been reported.[4,6]

Direct compression of the intraspinal dural sac and epidural venous engorgement had been clearly demonstrated in computed tomography (CT) or MRI studies. CT scans and myelography were not specific for SEL and could be misleading. Currently, MRI had replaced myelograms and CT scanning as the study of choice for the diagnosis of SEL. The adipose tissue was characteristic as high – signal intensity on T1-weighted images and intermittent to low-signal intensity on T2-weighted images. Spinal canal compression and subarachnoid spaces obliteration could be clearly demonstrated on the sagittal MRI images. In addition, a typical feature could be found on axial MRI images, where, the overgrowth adipose tissue circumferential compression of the thecal sac, referred to as the "Y-sign" or "Stellate-sign".[7] Epidural adipose tissue with a thickness greater than 7 mm was the diagnostic criteria for SEL.[3]

Exogenous steroid administration was a well-documented etiology,[8,9] however, in some the cause of the disease remained unknown. A number of endocrinopathy cases had been reported to have SEL, including some associated with Cushing's Syndrome,[8,10] Cushing disease, hypothyroidism[11] and pituitary prolactinoma.[12] Obesity had been attributed as a common feature in some of these patients, and in some the disease was considered to be "idiopathic". The popularity of exogenous steroid used, increased of the obese population and the rate of SEL diagnosis appeared to be correlated with the advent of MRI, these factors explained the reasons for an increase of SEL over the last decade.

Two treatment options of SEL had been recommended: conservative therapy and surgical intervention. No clinical trials had been performed to compare outcomes of both modalities because of limited number of cases. Conservative treatment contained weight loss and activity modification was performed on obese patients,[3,13] whereas non-obese patients

with no steroid-related SEL should undergo endocrinological evaluation.

Conservative treatment should be undertaken before surgical intervention, however patients with neurological compression signs tend to be managed aggressively. In our patients, we performed laminectomy and debulking of the deposited fat after SEL was diagnosed. An optimal improvement or resolution of the neurological symptoms after surgical intervention had been reported.[3,14] Both our patients showed good prognosis after surgical intervention.

In summary, SEL had been increased over the last decade due to the popularity of radiological survey. Further studies were necessary to determine the etiology and establish the treatment modalities. Surgical intervention by laminectomy and debulking of the deposited fat had appeared to benefit patients with neurological deficits or conservative treatment failure. The two cases presented should alert clinicians to fact the symptoms of spinal stenosis not only occurred in degenerative diseases and tumors. SEL, as a rare disorder, should be considered in differential diagnosis of patients with persisting back pain or other spinal neural elements compression.

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原發性脊椎硬膜外脂肪過多症：二病例報告和文獻回顧

陳俊賢 張志儒 鄭慶明[1] 黃世鴻[2] 劉元凱 黃金山

脊椎硬膜外脂肪過多症是脂肪在硬膜腔形成類腫瘤似的不正常堆積，它是長期服用類固醇患者不常見的併發症，且是造成脊髓壓迫症不尋常的導因。自 1990 到 2006 年，我們報告兩個未曾使用類固醇治療卻患有脊椎硬膜外脂肪過多症的病例。病人的臨床症狀一開始以下背痛來表現，漸進性下肢無力及麻木感，且快速進展成神經間歇性跛行。因爲誤診，病人被當著退化性脊椎疾病治療了一段很長的時間。終於，經核磁共振獲得診斷，我們爲病人施行椎板切除手術和硬膜外脂肪摘除，經此神經減壓後兩個病人均獲得痊癒。藉此，我們回顧文獻並摘錄目前處理這類案例的臨床趨勢。(長庚醫誌 2009;32:662-7)

關鍵詞：脂肪過多症，脊椎

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