Clinical Case Report

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Spinal cord compression syndrome caused by intraspinal epidural fibrous cord

Three case reports

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Abstract

Rationale: The spinal cord compression caused by intraspinal epidural fibrous cord.

Patient concerns: All patients in this study had spinal cord compression syndrome caused by an intraspinal epidural fibrous cord, manifested as abnormally increased epidural adipose tissue by imaging.

Diagnose: These abnormal fibrous connective tissue strips were not identical to the known pathological tissue such as "meningovertebral ligament." Instead, it might be a novel pathogenic cause for the spinal cord compression.

Interventions: The intraspinal exploratory operation.

Outcomes: the first case has expected effect, the remaining two need further test.

Lessons subsections: The disease could be easily misdiagnosed as spinal epidural lipoma or lipomatosis before the operation. However, the overt intraoperative finding was the indefinite starting and ending points of the epidural adipose mass in addition to the increased amount of adipose tissue. The obvious compression on the spinal cord could be found as the extraordinarily large and broad hypertrophic fibrous connective tissue strips.Further studies are needed to elucidate whether it is different from, or associated with, lipoma and epidural lipomatosis, which is a serious issue to be considered by both clinicians and radiologists. Therefore, early discovery,diagnosis, and treatment should be the prerequisites to achieve a satisfactory effect.

Abbreviations: BMI = body mass index, SEL = spinal epidural lipomatosis.

Keywords: fibrous cord, spinal cord compression, spinal epidural lipomatosis

1. Introduction

It is known that spinal cord compression is caused by intraspinal space-occupying diseases such as tumor, ossification of the posterior longitudinal ligament, and yellow ligament calcification. However, the compression caused by intraspinal epidural fibrous connective tissue has never been reported. The present study described the cases of 3 patients with paraplegia caused by intraspinal epidural fibrous strip, who were admitted to the hospital. Both intraoperative and postoperative pathology indicated that it was the intraspinal epidural fibrous strip that caused spinal cord compression. The origin of the fibrous strip was still unclear. The fibrous strips in the described cases were significantly different from the meningovertebral ligament[1,2] or Hofmann ligament[3,4] regarding length, thickness, width, staring point, or end point. Spinal cord compression is usually caused by spinal epidural lipoma and fat hyperplasia. Most cases have taken the corresponding segment of the whole laminectomy, decompression. There is no report of the existence of fiber cord compressing spinal cord during surgery.

The imaging features of these patients were the large accumulated adipose tissues deposited in the epidural space, accompanied by spinal compression by the fibrous strip. Owing to the lack of awareness on the disease, the fibrous strip could be disguised by accumulating intraspinal epidural adipose tissue, which easily led to miss diagnosis such as spinal epidural lipoma and lipomatosis. The aim behind sharing these cases was to increase the awareness of this type of disease to avoid missdiagnosis.

2. Case reports

This study was approved by Luoyang Orthopaedic Hospital of Henan Province. Informed consent was obtained from all individual participants included in the study.

2.1. Case 1

A 9-year-old girl was transferred to the hospital with the major complaint of "progressive weakness in the lower extremities for 7 months and incontinence for 3 months." The girl was 130 cm in

Editor: Song Liu.

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The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Medicine (2017) 96:33(e7592)

Received: 20 January 2017 / Received in final form: 24 May 2017 / Accepted: 3 July 2017

http://dx.doi.org/10.1097/MD.0000000000007592

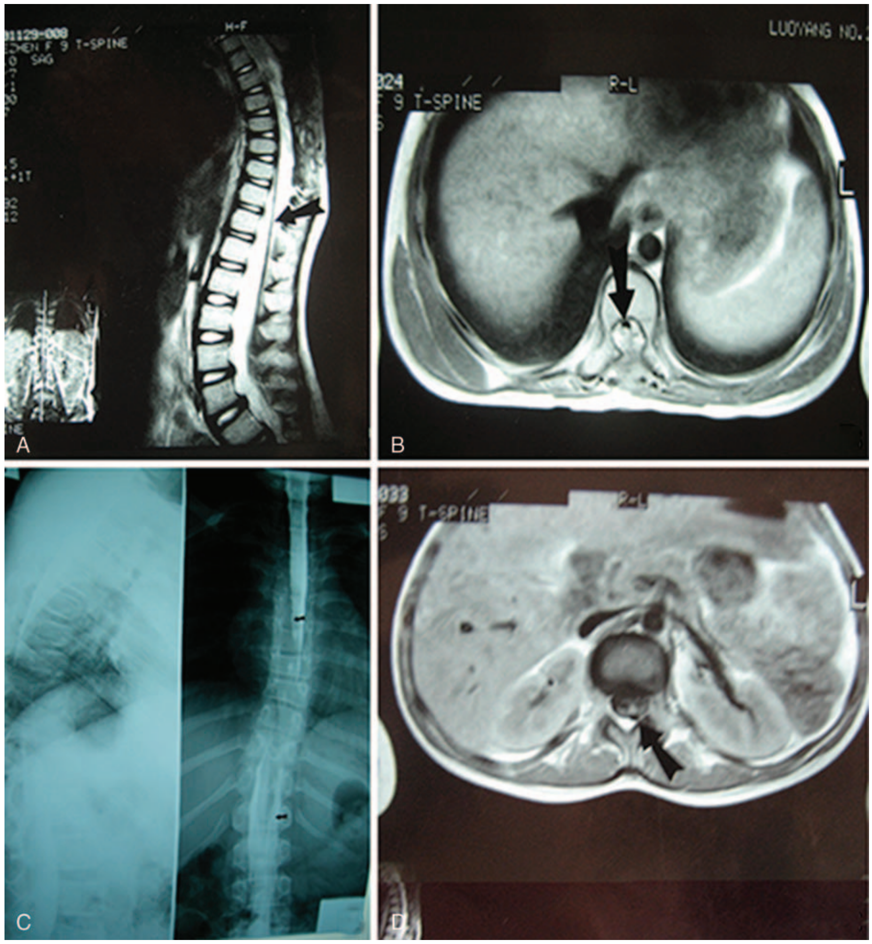

Figure 1. (A) Epidural spindle-like adipose tissue deposition and an abnormal fibrous strip were revealed by the sagittal-plane MRI. (B, C) The abnormal fibrous strip was revealed by the frontal-plane MRI. (D) Intraspinal filling defect displayed by the preoperative x-ray film; the dural sac and spinal cord were pushed away.

height and 26 kg in weight with standard nutritional status. She was diagnosed with "intraspinal epidural lipoma" 2 months ago, and the "T5–T10 vertebral lamina resection and lipoma resection" were performed under general anesthesia. The postoperative pathology suggested lipoma. However, the symptoms did not significantly improve but exaggerated around 1 week later, manifested as a significant movement restriction of 2 lower extremities, inability to walk, and urine or fecal incontinence. The physical examination revealed a 10 cm long incision scar on the back, sensation in 2 lower extremities, muscle strength grade II for the left hip and knee joints, and muscle strength grade 0 for the other muscles. Physiological reflexes of the bilateral abdomen and lower extremities were not induced, with Babinski sign (+) and bilateral ankle shock (+). The epidural spindle-shaped adipose deposition and abnormal fibrous strip shadow could be observed on the sagittal plane of magnetic resonance imaging (MRI), and the abnormal fibrous bundle could be revealed on the different sections of the frontal plane (Figs. 1–4). The cisterna magna and lumbar puncture for bidirectional radiography indicated the intraspinal epidural

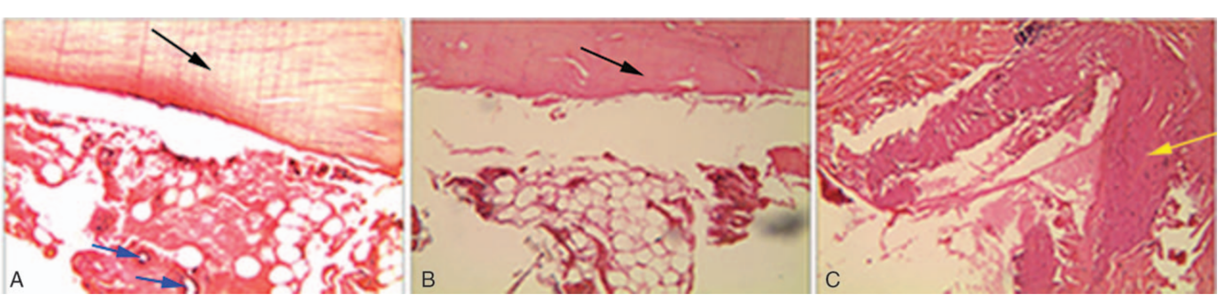

Figure 2. (A–C) Hematoxylin and eosin staining and 100-fold magnification. The condensed fibrous ligament was observed. The proliferation of fibrous connective tissues and thin-wall small blood vessels mixed in the mature adipose cells were also noted.

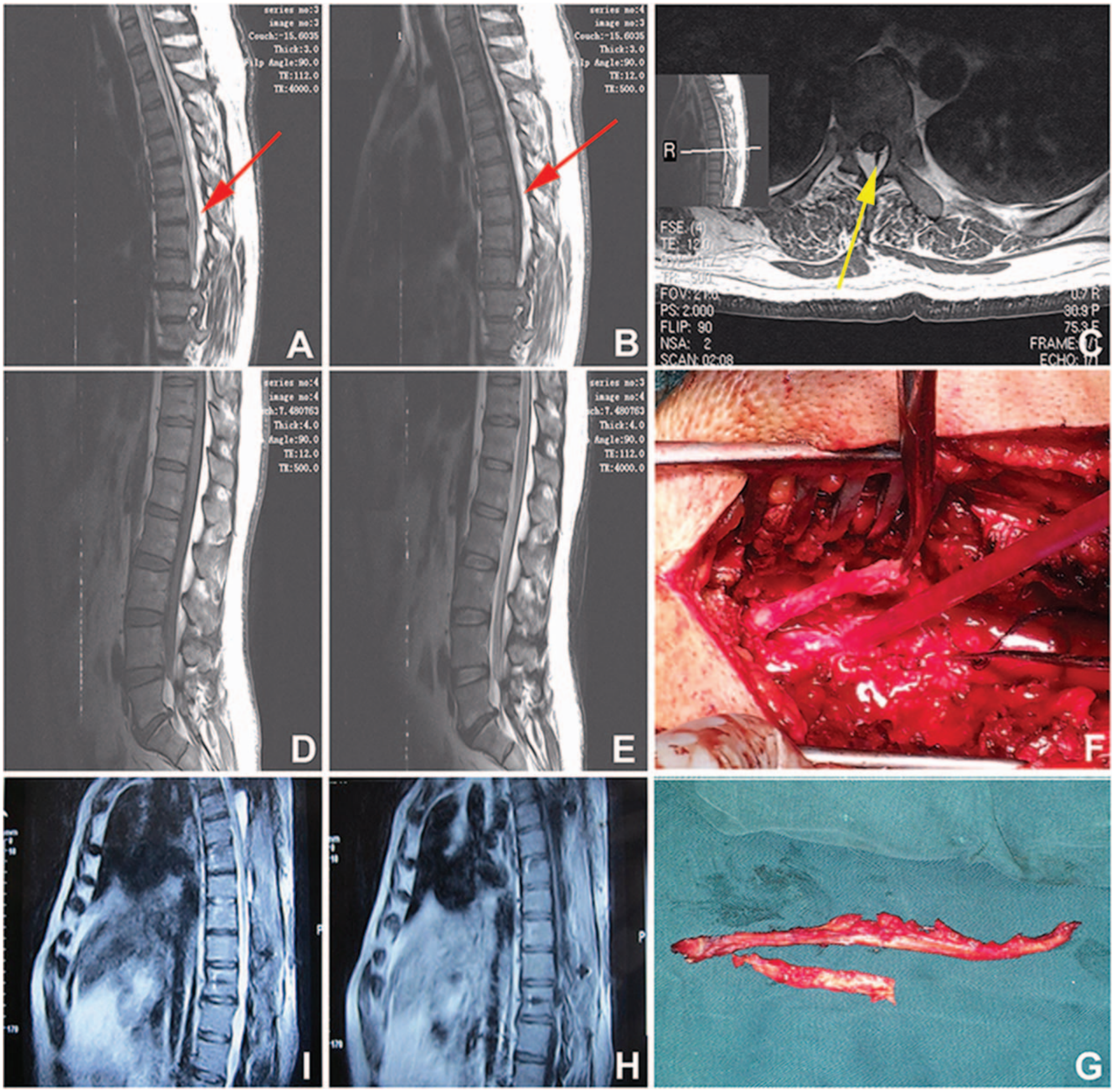

Figure 3. (A–C) Sagittal-plane MRI of the thoracic vertebral body suggested the spinal cord atrophy from C7 to T5, epidural lipoma or angiolipoma observed from T1 to T10, and fibrous strip signal from the left dural sac to the dorsal space. (D, E) Fibrous strip extended to L4 revealed by lumbar vertebra MRI. (F, G) The fibrous strip was raised and resected. (H, I) MRI revealed that dural sac and spinal cord compression disappeared 10 months after the operation.

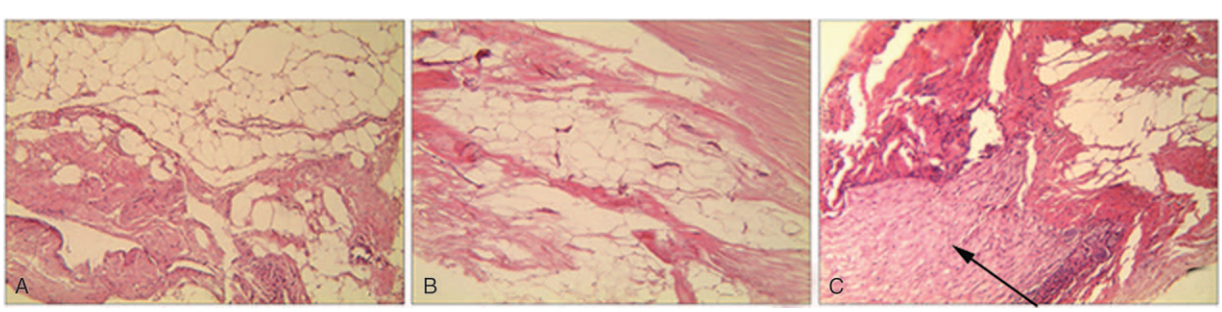

Figure 4. (A–C) Hematoxylin and eosin staining and 100-fold magnification. The nerve fibers were wrapped in the fibrous tissue.

spacing-occupying lesion from T2 to T11, manifested as the contrast agent filling defect and interruption. The dural sac and spinal cord were pushed to the left anterior.

The secondary intraspinal exploratory operation was performed on August 20, 1998. The vertebrae T3–T8 were routinely exposed, and it was found that the T5–T7 lamina had been resected. Then, the epidural scar was excised. A large amount of adipose tissue was observed once the T4 lamina was removed. The adipose had no envelope, with the color and texture similar to that of the normal adipose tissue. A fibrous strip with the thickness of 3 mm and width of 3.5 mm was exposed from the right posterior of the capsule, extending to the right anterior when the adipose tissue was removed. The tension of the fibrous strip was great without adhesion to the dural sac, and the dural sac was severely compressed by the fibrous strip such that the pulse could not be detected and the dural sac itself became flat. The fibrous strip was cut at the middle point, and contracted around 1 cm in cephalic and caudal directions. The starting point was not revealed even though it was explored till T3, and the end point was not found either, although it was explored till T8. Therefore, around 6 cm long strip was resected. Then, the dural sac bulged again and the pulse was recovered. The muscle strength of the left lower and right lower extremities was recovered to grades III and II, respectively, 2 days after the operation. The autonomous urination was also recovered. The muscle strength of the left lower extremity returned to normal, and the muscle strength of the right lower extremity was grade IV. Urination and defecation were nearly normal when the patient was discharged around 2 weeks after the operation. The dense fibrous ligament, fibrous collagen tissue proliferation, and mature fat cells mixed with small thin-wall vessels were observed under a microscope during the postoperative pathological examination. The sensation and muscle strength of the lower extremities were completely recovered, and urination and defecation returned to normal 1 year after the operation when the patient underwent follow-up examination.

2.2. Case 2

The patient was a 40-year-old man, 180 cm height and 95 kg weight; body mass index (BMI) was 29.32 kg/m^2. The patient had no history of intrathecal injection of glucocorticoids, and was admitted to the hospital with the major complaint of "progressive weakness in the lower extremities for more than 6 years." The patient had perceptive weakness of the left ankle back extension around 6 years ago and underwent tendon transfer in the County People's Hospital (detailed information not available). The treatment effect was unsatisfactory, and the weakness in the lower extremities progressively exaggerated. After admission, MRI revealed a long segmented spindle-like short-T1 and long-T2 signal, and the dural sac was pushed forward. The C7-T5 segment of the spinal cord was significantly thinned with a small patch of medium-T1 and slightly long T2 signal, and the boundary was blurred, suggesting "(1) C7–T5 vertebral segment spinal cord atrophy and (2) T1–10 epidural angiolipoma." The physical examination indicated no obvious abnormality or tenderness, and the sensation of the lower extremities diminished below the groin. The muscle strength of the bilateral quadriceps femoris was grade IV. The muscle strength of right ankle dorsiflexors and plantar flexors was grade IV. Other findings were left ankle joint prolapse with inversion deformity, no dorsal flexion movement, hyperreflexia of bilateral knee and heel tendon, bilateral Babinski sign positive, and ankle and patellar clonus positive. The sensation around the saddle area was normal; however, the anal reflex was weakened. The intraspinal exploratory operation was performed on March 2, 2012, under general anesthesia. A tenacious strip-like tissue was observed intraoperatively posterior to the dural sac. A large amount of adipose tissue was attached to the surface and led to the significant compression on the dural sac. The dural sac floated backward and bulged when the tenacious strip was raised manually. The epidural strip was thinner when it was explored to the T11 vertebral segment level. The dural sac bulged with good pulsation when around 23 cm epidural strip was removed from T5 to T11. The nerve fiber was found to be wrapped in the fibrous tissue. The pain and temperature sensation was partially recovered, despite no significant improvement in the muscle strength 15 months after the operation.

2.3. Case 3

The patient was a 62-year-old man, 170 cm height and 50 kg weight; BMI was 17.3 kg/m^2. The patient had no history of intrathecal injection of glucocorticoids, and was admitted to the hospital with the major complaint of "numbness and weakness of the lower limbs for more than 20 years, which aggravated for 5 years." The numbness and weakness of the left lower extremities were found around 20 years ago. However, no obvious abnormality was found by the local hospital. The symptoms aggravated in 5 years. The MRI examination at the outpatient department revealed the posterior epidural strip with nonuniform hyperintensity signals from the inferior rim of T2 vertebral body to the superior rim of T10 vertebral body. The strip of hypointensity signals was detected inside, with the fat suppression signal uniformly reduced. The enhancement scanning indicated the enhanced strip shadow and no abnormal signal within the spinal cord. Therefore, the patient was admitted with the diagnosis of "epidural angiolipoma." The physical examination showed numbness below the mastoid process and obvious atrophy of 2 lower extremities. The muscle strength of the left lower and right lower extremities was grade III and grade IV, respectively. The other positive findings included numbness of saddle area, bilateral knee and heel tendon hyperreflexia, bilateral Babinski sign positive, and left patellar and ankle clonus (right was negative). The intraspinal exploratory operation was performed on May 20, 2015, under general anesthesia. A posterior median incision was made to expose the tissue of each layer and resect the lamina of T4–T10. A fibrous strip gray white in color was observed in the dural dorsal space, covered by a large amount of vascular-rich adipose tissue. Some parts of the segments adhered to the dural sac and were difficult to separate. Obvious hemorrhage was observed when the adipose tissue was removed. The fibrous strip was peeled off slowly from inferior to superior, and around 11 cm strip was eventually resected and sent for pathological examination. The bleeding from the lateral wall venous plexus was stopped by the gelatin sponge compression hemostasis. The drainage was placed before the incision was closed sequentially. The fibrous connective tissue was found by postoperative pathological examination, and a large number of nerve fibers were distributed into it. Many thick-walled and dilated blood vessels could be found in the mature adipose tissue. The perception of numbness was similar to that before the operation when the patient was followed up for 10 months and no improvement in the walking ability of bilateral extremities was observed (Figs. 5 and 6).

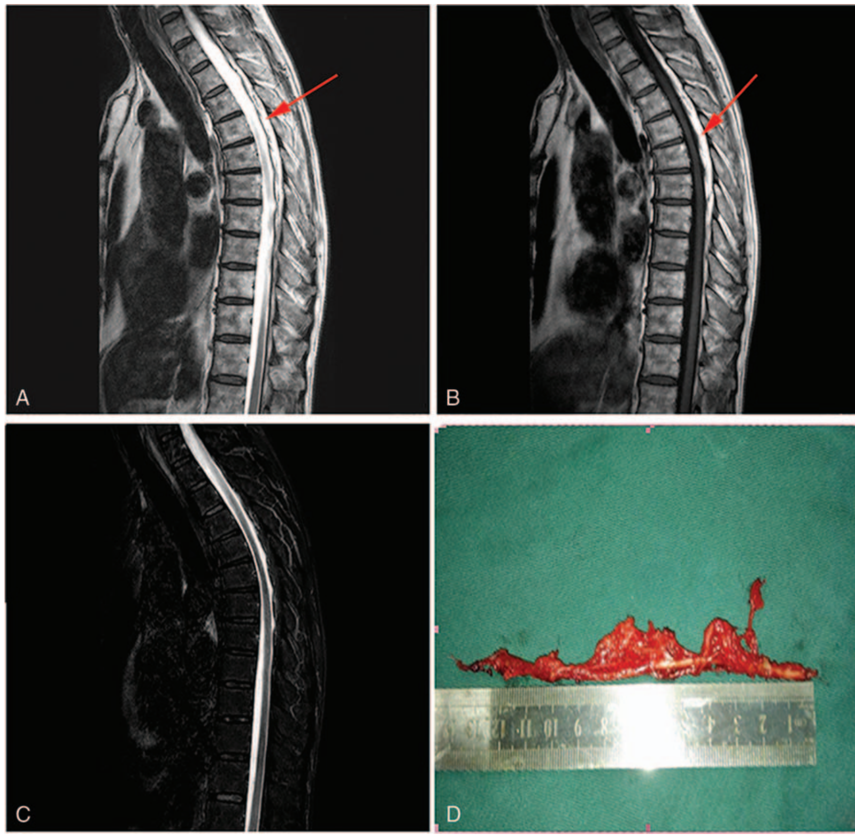

Figure 5. (A–C) Posterior epidural strip with nonuniform hyperintensity signals was detected from the inferior rim of T2 vertebral body to the superior rim of T10 vertebral body. The strip of hypointensity signals was detected inside, with the fat suppression signal uniformly reduced. (D) The fibrous strip was intraoperatively removed.

3. Discussion

The common features among the aforementioned three patients were as follows: spastic paralysis of the lower extremities with varied severity was manifested by all patients; the abnormal increase in epidural adipose tissues was revealed by imaging examination; and a hypointensity signal of the epidural fibrous strip was detected. The spinal cord atrophy was found in the 2 adult patients.

All patients were preoperatively misdiagnosed with intraspinal lipoma or angiolipoma. The intraoperative observation confirmed that the spinal cord was compressed by an abnormal fibrous strip, which was a new pathogenic feature. In Case 1, the partial adipose tissue was removed by the primary operation, but no obvious therapeutic effect was observed. In the secondary operation, a large amount of epidural adipose tissue was found again but was not removed. Only the fibrous strip was cut off, and the symptoms of the lower extremities were improved and gradually recovered. The unexpected satisfactory outcome indicated that the spinal cord compression was caused by the fibrous strip. The boundary between the epidural adipose tissue and the fibrous strip was clear in the first patient, and the strip was easily separated. However, the adipose tissues of the 2 adult

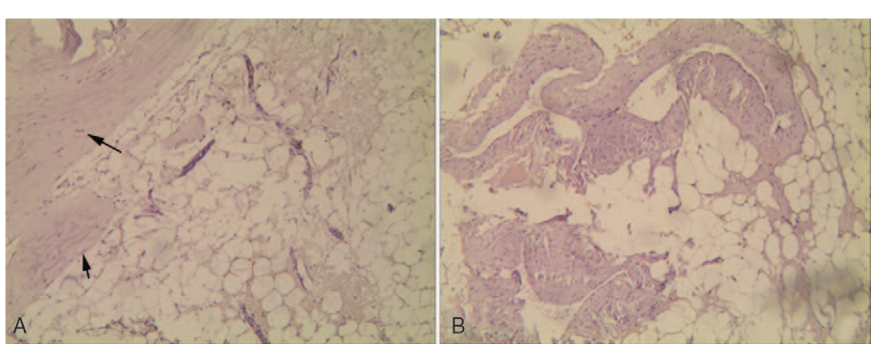

Figure 6. (A, B) Hematoxylin and eosin staining and 100-fold magnification. A large number of nerve fibers were distributed in the fibrous connective tissue, as observed under the microscope. Many thick-walled and dilated blood vessels were noted in the mature adipose tissue.

patients were attached to the fibrous strip. Especially in Case 3, the fibrous strip was tightly adhered to the dural sac and hence difficult to separate. Therefore, it was speculated that these 3 patients might have the same disease but at different stages. The 2 adult patients had spinal cord atrophy suggested by preoperative MRI due to long-term missed diagnosis, misdiagnosis, and mistreatment, and hence the motor function had no significant improvement after the operation. Therefore, early discovery, diagnosis, and treatment should be the prerequisites to achieve a satisfactory effect.

The diagnosis should be differentiated from the spinal epidural lipomatosis (SEL). SEL demonstrated a typical "Y" or "star" shape on the transverse sectional MRI.[5,6] Al-Khawaja et al [7] reported that around 90% patients underwent surgeries, and most of them had full laminectomy decompression and fat resection. No fibrous strip or similar observations were reported, which was significantly different from the findings in the aforementioned cases. The origin of the fibrous strip was still unclear.

In summary, the aforementioned cases were different from angiolipoma or SEL, and the abnormal fibrous strip compressing the spinal cord was one of the novel pathogenic factors. As the clinicians lacked awareness of this type of disease, the increased amount of epidural adipose tissue revealed by MRI, which covered the fibrous strip and led to misdiagnosis and mistreatment, was the common feature in the cases that required the attention of clinicians and radiologists.

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