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Case Report

Challenging diagnosis of spinal epidural lipomatosis: Case report and review of the literature.

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Abstract

Spinal Epidural Lipomatosis (SEL) is a rare disorder characterized by an overgrowth of the spinal epidural fat in the extradural space. It commonly affects male patients. The pathogenesis of SEL may involve exogenous steroid use, obesity, overproduction of endogenous steroids (like in Cushing's disease), spine surgery, hypothyroidism, and pituitary prolactinoma. However, idiopathic cases have been identified. The best modality of SEL diagnosis is Spine MRI with and without Gadolinium contrast to assess the fatty tissue along the extradural space. Treatment of such rare entities depends on the severity of the clinical presentation. Surgical and non-surgical treatment plans may be considered in these cases. We herein report a case of spinal epidural lipomatosis associated with kyphosis in a 17 -year- old male patient, who was successfully treated with kyphosis dorsal fixation and hemilaminectomy for the spinal cord decompression.

Keywords

Epidural Lipomatosis; Spinal Decompression; Steroid; Obesity; Kyphosis; Paralysis.

Introduction

Spinal Epidural Lipomatosis (SEL) is condition that results from excessive accumulation of unencapsulated adipose tissue in the extradural space of the spinal cord [1]. It mostly occurs in the thoracic and lumbosacral regions. It commonly presents in males. Exogenous steroids use, hormonal diseases with increased endogenous steroid levels, obesity, and spinal surgeries are reported to be the major causes of SEL [2]. However, idiopathic SEL have been widely reported [3]. Based on a twenty years Pubmed literature review, we tried to highlight the characteristics and the diagnostic difficulties of this rare entity.

Observation

A 17-year-old male patient presented to our hospital with a chief complaint of paraplegia of nine months duration. The patient had no chronic diseases and underwent laparoscopic appendectomy 2 years ago. The symptoms started with right knee pain, imbalance while walking, difficulty of swallowing and incontinence for stools and urine. No history of trauma and no regular use of any kind of medications were noted.

Whole-body Computed Tomography (CT) scan was done with no remarkable abnormalities. Due to the persistence and aggravation of the symptoms, transverse myelitis was suspected. Solumedrol and intravenous Immunoglobulin (IV-IG) were prescribed with no improvement. Bone marrow analysis showed hypocellularity and T-cell clonality. Plasmapheresis was indicated. Six sessions were performed with objective sensational and motor improvement. Few months later, a sudden relapse was observed. Neurological examination revealed spastic paraplegic lower limbs, bilaterally positive Babinski reflex and decreased sensation from the level of L2 and below. Spine MRI and CT scan (figure 1) were done and revealed Spinal Epidural Lipomatosis (SEL) with progressed kyphosis in the thoracic spinal region. The patient underwent D6-D7-D8-D9-D10 transpedicular fixation for correction of the kyphosis and left hemi-laminectomy for decompression of the spinal cord. The postoperative Course was uneventful. Postoperative spine CT and MRI follow up showed satisfactory dorsal epidural dorsal fat resection and optimal dorsal screws placement (figure3). The patient was referred to the physiotherapy and rehabilitation department. On the 2 months follow up, the patient was able to walk without assistance and no incontinence was noted.

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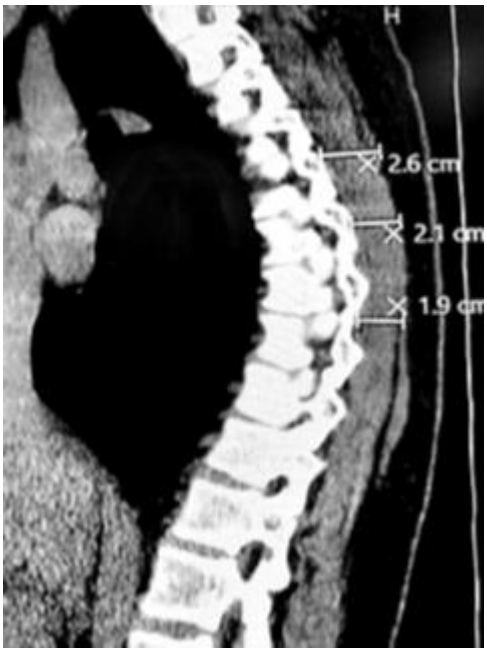

Figure 1: A sagittal view of a dorsal spine CT scan without contrast showing a thickening of the epidural fat and dorsal kyphosis at levels D6-D10.

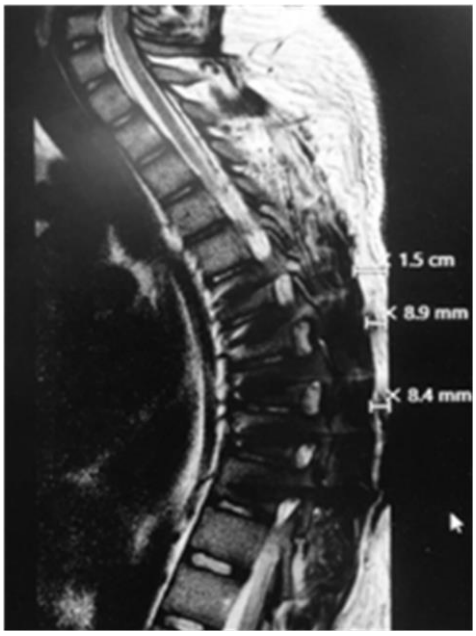

Figure 2: Figure 2: Sagittal view of postoperative dorsal spine MRI scan showing considerable resection of the epidural fat.

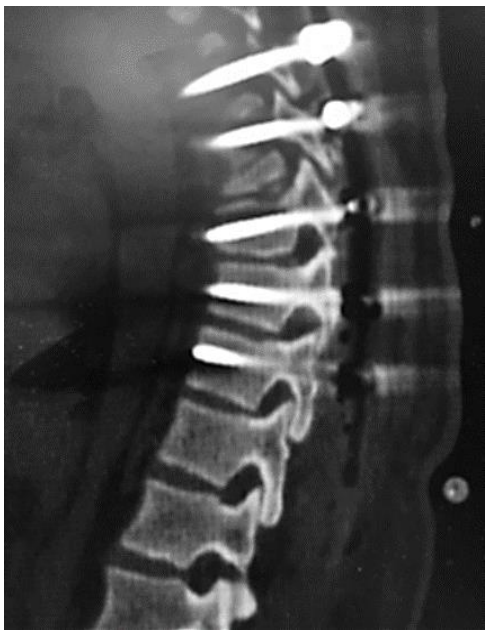

Figure 3: Sagittal view of dorsal spine CT scan without contrast showing satisfactory correction of the kyphotic spine D6-D10.

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Discussion

Spinal Epidural Lipomatosis (SEL) is a rare condition characterized by an overgrowth of unencapsulated adipose tissue in the extradural space. Excessive tissue accumulation in the thoracic and lumbar regions may lead to spinal cord compression and neurological complications [4]. More common in males, the incidence of this entity varies from 0.6 to 2.5%. However, some of the epidemiologic characteristics are still to be assessed on larger scale studies [5-7]. The incidence may be higher in obese patients and in case of corticosteroid therapy [8]. SEL has been reported in children with chronic renal disease, who underwent methylprednisolone pulse treatment [9-11]. SEL present usually in the thoracic spine in more than 50% of cases. Lumbar spine SEL is seen in 40% of cases most commonly at L5-S1 level [12,13]. SEL is a multifactorial entity. The most common leading factor is the long corticosteroid therapy. It was reported in about 70% of SEL cases mostly located in the thoracic spine [14]. Obesity and its related syndromes may be involved in more 24% of cases [15-16]. Endogenous steroid hormonal diseases such as Cushing syndrome, hypothyroidism, and prolactinoma are noted in about 3.2% of cases [17]. Other causes such as history of spinal surgery and idiopathic SEL are reported in 17% of cases. These cases are mostly located in the lumbar spine [18].

SEL pathogenesis involves the activation of adipocytes in the spinal epidural space and steroids-induced corticosteroids receptors stimulation. That would explain the excessive fat accumulation in thoracic and lumbar spine regions [19-21]. SEL was reported in several other pathologic conditions in which the spinal adipose tissue overgrowth was directly or indirectly enhanced. This would be the case of some AIDS patients undergoing regular highly active antiretroviral therapy, Scheuermann's disease (Congenital kyphosis) patients, and some of prostate cancer patients following androgen antagonist treatment [22,23].

SEL presentation is usually paucisymptomatic. Symptoms are with non-specific. Patients are usually diagnosed with transverse myelitis and treated accordingly. SEL can present with radiculopathy, myelopathy, claudication, cauda equina syndrome (CES), or paraplegia. These spine cord compression related symptoms depend on the location and degree of adipose tissue overgrowth. Acute-onset paraplegia reported in steroid-induced SEL may be due to thoracic compression fracture due to osteoporosis in the setting of long-term exposure to steroids[24].

MRI is the most accurate for SEL diagnosis. It provides a higher sensitivity in the fat accumulation measurement and subsequent dural sac deformation [25,26]. Appropriate MRI request timing would reduce misdiagnosis rate and enhance the prognosis of this disease via early targeted management [26].

Management of SEL can be rarely conservative. More than 90% of patients are undergo surgery either directly or after failure of the conservative option [27]. Conservative management is mainly based on the discontinuation of the exogenous steroid in SEL related cases and reducing the patients BMI.

Surgery should be considered immediately in severe cases. Patients usually report gradual recovery after laminectomy and show significant improvement in pain and quality of life. Surgical decompression and removal of excess fatty tissue is a reasonable option in patients with acute cord compression. However, the conservative management is still recommended first, and the surgery is indicated almost only on an emergency basis. The prevention and appropriate monitoring of high-risk patients specially with predisposing hormonal status should improve the diagnosis and the management of this entity [28-30].

Literature Review

Using the following key: spinal, epidural, lipomatosis, characteristics, management, an electronic database research was conducted in Pubmed, Medline and Embase. Two hundred thirty-one articles were reviewed. After exclusion of non-relevant, thirty paper were collected in this cased based literature review.

Conflict of Interest: None

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