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CASE REPORT

Lipomatosis of spinal epidural space, peritoneum, and renal sinus: a rare complication of long-term steroid therapy in a child with nephrotic syndrome

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Abstract
Excessive visceral adipose tissue proliferation, resulting in diffuse lipomatosis, is a rare complication of long-term steroid therapy. A 10-year-old boy presented with severe radicular back pain with limitation of lower limb movements. He was diagnosed with steroid-resistant nephrotic syndrome and was on unregulated steroid therapy. Magnetic resonance imaging of the spine showed increased adipose tissue in the epidural space of the lumbo-sacral spine causing clumping of cauda equina nerve roots along with marked proliferation of fat in the renal sinus as well as peritoneum. He was started on pregabalin with tapering of steroids, following which there was a gradual decrease in pain and improvement of activity. Our patient had diffuse lipomatosis involving spinal epidural space, bilateral renal sinus, and peritoneum, secondary to steroid overuse. With the availability of advanced imaging techniques, the condition can be prevented by judicious and proper use of steroids with close follow-up for any untoward complications.

Keywords Diffuse lipomatosis · Steroid toxicity · Adipose tissue · Epidural fat

Introduction

Diffuse lipomatosis is a rare entity, characterized by massive proliferation and accumulation of mature adipose tissues which lack of encapsulation with an extensive infiltrating pattern. Diffuse lipomatosis can involve any part of the body. Abdominal lipomatosis commonly involves the intraperitoneal and the retroperitoneal space with extensive accumulation of non-encapsulated adipose tissues [1]. Spinal epidural lipomatosis (SEL) is a rare disease characterized by overgrowth of adipose tissue in the epidural space of the spinal cord, which slowly causes compression of the spinal cord. Here, we report a child with steroid-resistant nephrotic syndrome who had diffuse lipomatosis.

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Case study

A 10-year-old-boy presented with back pain and painful limitation of movements of the lower limbs for the past 2 months. The pain was slowly progressive and radicular in nature, radiating to his legs, along with painful limitation of movement of lower limbs. There was no evidence of loss of sensation and bowel or bladder involvement. He was diagnosed with steroid-resistant nephrotic syndrome (focal segmental glomerulosclerosis on renal biopsy) since 2 years of age. He was started on steroid-sparing agents (tacrolimus). However, he was lost to follow-up and his parents were treating him with unregulated dosage of oral prednisolone for the interim period (approximate cumulative dose of steroid administered for 3 years, 380–460 mg/kg). On examination, he had cushingoid appearance, hypertrichosis, morbid obesity (BMI, 32.3 kg/m^2), hypertension, bilateral subcapsular cataract, normal muscle power, and deep tendon reflexes. However, he had marked restriction of movements of lower limbs due to pain. Systemic and rest of the neurological examination was unremarkable.

Investigations revealed normal hemogram and renal function tests (urea, 38 mg/dl; creatinine, 0.61 mg/dl). He had low serum vitamin D levels (25 (OH) D3–

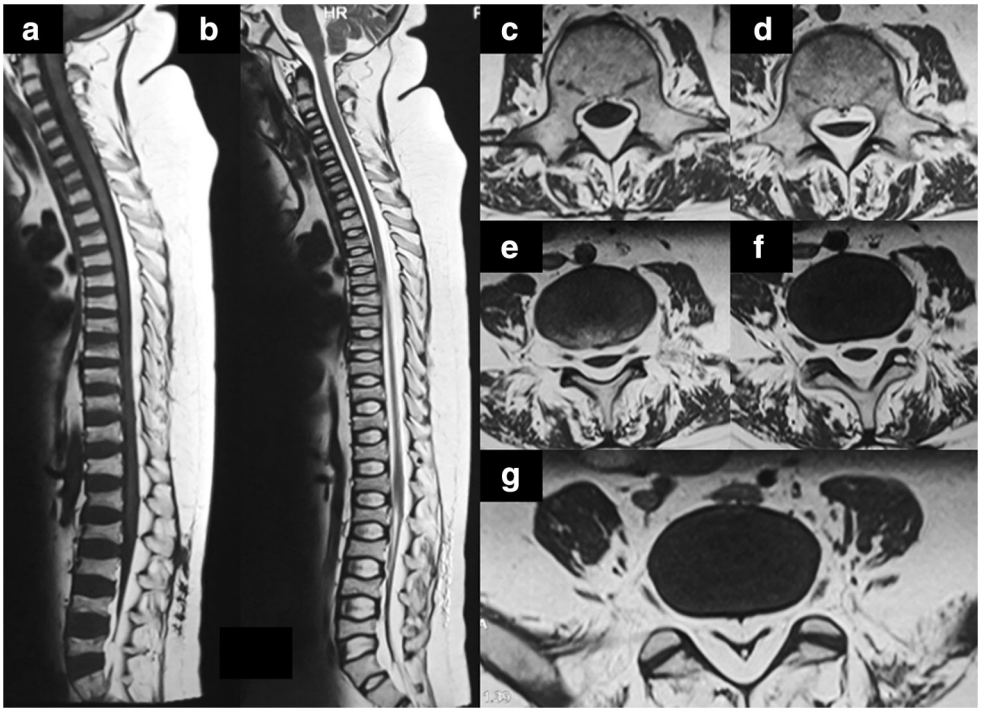

Fig. 1 MRI of the spine. T1-weighted (**a**) and T2-weighted (**b**) sagittal sections showed diffuse scalloping of the vertebral bodies with mild vertical height reduction with extensive epidural fat in lumbo-sacral spine causing marked effacement of the thecal sac and centrally placed clumped cauda equina nerve roots, most marked at L5–S1 level. The severity of fat deposition progressively increases (short-TI inversion recovery sequences, axial sections, **c**–**g**) from L2 to S1 levels, making an inverted Y appearance (g)

12.09 ng/ml), elevated alkaline phosphatase (362 IU/l), normal serum calcium (8.6 mg/dl), phosphorus (4.4 mg/dl), and parathyroid hormone levels (43.30 pg/ml). Urinalysis showed non-nephrotic range proteinuria (19.6 mg/m^2/h). Skeletal X-rays showed marked osteopenia in long bones of the upper and lower extremities, as well as of the vertebrae without any evidence of fracture. Dual-energy X-ray absorptiometry scan showed osteoporosis (− 3.6 *z* score), with an increase in body fat content. His nerve conduction study was normal. Magnetic resonance imaging (MRI) of the spine (Fig. 1) and abdomen (Fig. 2) revealed increased adipose tissue in the epidural space of lumbo-sacral spine and marked proliferation of fat in bilateral renal sinus, suggestive of renal sinus lipomatosis, as well as in the peritoneum. He was started on oral amlodipine and enalapril for hypertension. Steroid-sparing agent Tacrolimus was restarted in view of the disease process and prednisolone was tapered to alternate day regime (40 mg alternate day for 4 weeks followed by tapering at 5 mg every 2 weeks till a minimum dose of 10 mg alternate day). Calcium and vitamin D supplementation was initiated, along with pregabalin for the neuropathic pain. Over a period of 1 month during the hospital stay, his clinical symptoms improved. On follow-up at 2 months, he lost 10 kg of weight and is able to walk without any support with the absence of pain symptoms. Follow-up imaging of the spine was not performed due to significant improvement in his symptoms.

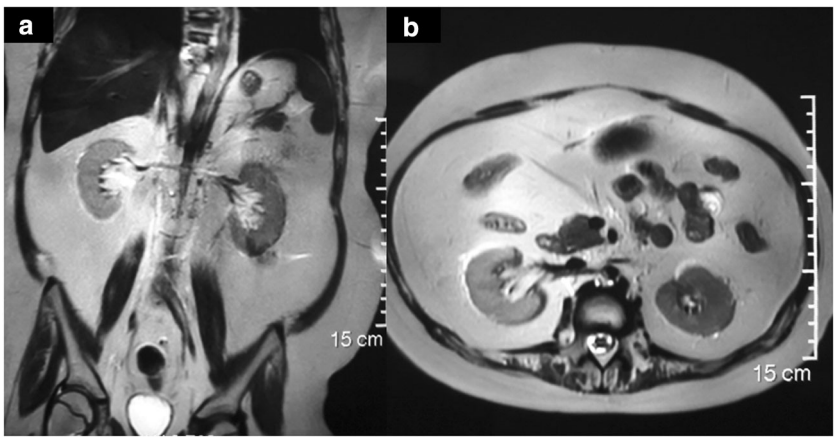

Fig. 2 MRI of the abdomen. T2-weighted coronal (**a**) and transverse section (**b**) showed diffuse fat deposition in bilateral renal hilum and peritoneum

Table 1 Characteristics of the children with corticosteroid-induced spinal epidural lipomatosis

S. No	Author, year	Age (years)	Sex	Diagnosis	Duration of steroid therapy (months)	Dosage of steroid	Symptoms	Imaging	Extent of involvement	Therapy	Clinical outcome
In English literature											
1.	Lee (1975) [6]	17	M	Renal transplant	12	40 mg/day	LL weakness	Myelogram	T1–T11	Surgery	Improvement
2.	George (1983) [7]	13	M	Renal transplant	18	45 mg/day	LL weakness, hip and back pain	Myelogram	T1–L5	Steroid reduction	Resolution
3.	Perling (1988) [8]	6	M	juvenile idiopathic arthritis	48	40 mg/day	LL weakness and pain, bowel and bladder dysfunction	Myelogram	T2–T6	Surgery steroid reduction	Improvement
4.	Quint (1988) [9]	11	M	Pinealoblastoma	12	20 mg/day	LL weakness, bowel and bladder dysfunction	MRI	T3–T9	Surgery	Progression
5.	Arroyo (1988) [10]	6	M	Systemic onset juvenile idiopathic arthritis	12	10–40 mg/day	LL weakness, back pain	CT	T6–T7	Surgery	Resolution
6.	Vazquez (1988) [11]	16	M	Renal transplant	36	0.4/kg/day	LL weakness, back pain	CT	T1–T12	Steroid reduction	Resolution
7.	Munoz (2002) [3]	8	F	Crohn's disease	78	10–60 mg/day	Back pain	MRI		Steroid reduction	Died
8.	Miller (2002) [12]	14	M	Systemic lupus erythematosus	10	60 mg/day	Back pain	MRI	T1–L3	Steroid reduction	Improvement
9.	Kano (2004) [5]	5	F	Nephrotic syndrome	17	5–60 mg/day	Back pain	MRI	T4–S1	Steroid reduction	Resolution
10.	Kano (2005) [4]	10	F	Nephrotic syndrome	5	20–60 mg/day	Back pain	MRI	T7–T9	Steroid reduction	Resolution
11.	Caruba (2010) [13]	12	F	Lung transplant	1	25 mg/day	LL weakness, dysuria	MRI	T2–T11	Steroid reduction	Resolution
12.	Moller (2011) [14]	10	F	Relapsing polychondritis	5	30 mg/day	Back pain	MRI	T1–S5	Steroid reduction	Resolution
13.	Index case	10	M	Nephrotic syndrome	36	380–460 mg/kg (cummulative dose)	Back pain	MRI	L5–S1	Steroid reduction	Improvement
In other languages											
14	Moller, (2010) [15]	14	F	Systemic lupus erythematosus	6	0.2/kg/day	Back pain, bladder and bowel dysfunction	MRI	T2–L5	Steroid reduction	Resolution
15	Moller, (2010) [15]	11	F	Sjogren syndrome	7	0.5/kg/day	Back pain	MRI	L4–L5	Steroid reduction	Resolution
16	Moller, (2010) [15]	7	F	Systemic onset juvenile idiopathic arthritis	18	40 mg/day	Back pain	MRI	T2–S5	Steroid reduction	Resolution
17	Shiraj (1990) [16]	10	M	Nephrotic syndrome	9	60 mg/day	LL weakness, back pain, bladder and bowel involvement	Myelogram MRI	T11–L3	Steroid reduction	Improvement
18	Kano (1996) [17]	11	M	Nephrotic syndrome	10	12–60 mg/day	LL weakness, back pain	Myelogram	T1–L2	Surgery	Improvement
19	Kano (1996) [17]	14	F	Nephrotic syndrome	3	24–80 mg/day	Back pain	MRI	L3–S1	Steroid reduction	Resolution
20	Kano (1996) [17]	14	M	Nephrotic syndrome	5	48-80 mg/day	Back pain	MRI	T4–T8 L4–S1	Steroid reduction	Resolution
21	Kano (1996) [17]	10	M	Henoch Schonlein purpura	10	36–72 mg/day	Numbness	MRI	T2–T6	Steroid reduction	Resolution

CT, computerized tomography; *MRI*, magnetic resonance imaging; *M*, male; *F*, female

Discussion

Diffuse lipomatosis in the pediatric age group is rarely seen and majority of the cases reported in the literature are from the adult population. The spectrum of presentation varies from involvement of the neck, trunk, and extremities to the abdomen including pelvis, intestine, and peritoneum [2].

Steroid-induced spinal epidural lipomatosis (SSEL) in children is a rare complication due to prolonged use of corticosteroids [3]. The frequency of SSEL was found to be 4% in children with renal diseases where corticosteroids are the mainstay of therapy [4]. Steroid-sparing agents whenever indicated can help in the reduction and discontinuation of steroid use and hence further reduce the serious adverse effects [5]. SSEL in the pediatric population is extremely rare, and only 12 cases have been described worldwide in English literature [3–14] and eight cases in different languages [15–17](Table 1). It was first reported by Lee et al. [6] in a renal transplant patient who was on oral prednisolone and presented with neurological symptoms secondary to epidural fat deposition and underwent laminectomy. Exogenous glucocorticoid therapy is the commonest cause, accounting for 75% of the cases [18]. Nephrotic syndrome is one common renal disease in children where corticosteroids are the mainstay of therapy [19]. Other causes include Cushing disease, hypothyroidism, prolactinoma, and obesity. SSEL affects both genders. The median age of reported case is 11 years (range, 5–17 years). The median duration of steroid used is 10 months (range, 1–78) and median dose of steroids is 40 mg/day. The thoracic spine is the commonest site of involvement (90%) followed by the lumbo-sacral spine. Overgrowth of adipose tissue in the epidural space of the spinal cord causes slow progressive compression of the cord. Back pain is the commonest symptom, followed by progressive weakness of the lower limbs. Radicular pain and paresthesia are also reported; however, bowel and bladder involvement is rare. SEL in the thoracic spine presents as myelopathy but manifests as radiculopathy if it affects the lumbar segments [20]. Although X-ray of the dorso-lumbar spine cannot detect SEL, it is a good screening tool to pick up degenerative diseases and fractures. Magnetic resonance imaging of the spine is the modality of choice and demonstrates fat in the epidural space, which is typically hyperintense on T1-weighted images, with the typical "inverted Y" appearance of the dural sac on the axial sections [4]. Treatment is mainly conservative, including tapering of exogenous steroids and weight loss. In case of endogenous hormonal excess, specific therapy is indicated. Surgical therapy includes decompressive laminectomy and excision of adipose tissue and is usually reserved for patients with cord compression and cauda equina syndrome [21]. Majority of the patients have good outcome [22]. Our child showed good improvement on conservative management and laminectomy was not performed.

Renal sinus lipomatosis is caused by the similar pathology where there is abnormal proliferation of adipose tissue in the renal sinus but is asymptomatic and does not affect renal function. It can be secondary to aging, infection, obesity, calculus, renal transplantation, and even with steroid excess and can mimic lipoid neoplasms of the kidney like angiomyolipoma, lipoma, and liposarcoma [23]. Patients are usually asymptomatic, but may have non-specific complaints like malaise, fever, or flank pain. Ultrasound is usually suggestive of a hyperechoic mass in the renal fossa, suggestive of fatty tissue. Computed tomography and MRI can identify fatty nature of the mass and define its extent as well as detect associated complications. Treatment of this condition usually depends upon the underlying etiology.

Peritoneal lipomatosis is extremely rare, with around 50 cases described, among which only four belong to the pediatric group. It is characterized by diffuse proliferation of intra and retroperitoneal fat and may present with abdominal distension, pedal swelling, ureteral obstruction leading to renal failure, and features of bowel and bladder dysfunction [24].

Conclusions

Exogenous steroid use has often been associated with spinal epidural lipomatosis. However, diffuse lipomatosis has rarely been reported. Steroid-induced diffuse lipomatosis is an uncommon condition of long-term steroid therapy. The cases diagnosed late usually requires surgical intervention whereas if diagnosed early can be managed conservatively with good clinical outcome. In our patient, the symptoms improved with conservative management. With the increasing awareness of the condition and the availability of advanced imaging techniques, the condition can be prevented by judicious and proper use of steroids with close follow-up for any untoward complications. A high index of suspicion is necessary for clinicians to be able to diagnose the condition.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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