Spinal epidural lipomatosis due to adrenal cortisol secreting adenoma in a 14-year-old girl. A case report

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ABSTRACT

Background: Spinal epidural lipomatosis (SEL) is an excessive fat deposition within the epidural space of spinal canal leading to compression of nervous structures and neurological manifestations. Exogenous glucocorticoid administration is the major cause of symptomatic SEL. The reported patient had SEL related to endogenous Cushing syndrome, which is among a few reported cases in the literature.

Case Presentation: A 14-year-old girl with features of Cushing syndrome had severe back pain and the magnetic resonance imaging of the spine showed SEL. Subsequently, computed tomography of the abdomen showed right adrenal adenoma. The patient underwent adrenalectomy which improved her signs of hypercortisolism and SEL.

Conclusion: In view of severe neurological deficits and therapeutic consequences, SEL should be considered in all patients with Cushing syndrome who present with symptoms of spinal cord compression. Correction of underlying endocrinopathy is imperative in the management of SEL.

Keywords: Spinal epidural lipomatosis, cushing syndrome, adrenal adenoma, case report.

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Introduction

Spinal epidural lipomatosis (SEL) is a rare disease, where hypertrophy of the epidural adipose tissue causes narrowing of the spinal canal thereby compressing neural structures. Patients usually present with progressive neurological deficits or radicular pain [1]. The cause behind it may be prolonged exogenous steroid therapy, endogenous overproduction, or idiopathic [2]. Body mass and tissue sensitivity to glucocorticoids are probably important factors in the development of SEL [3]. In adults, the prevalence of overall SEL was 2.5%, but only a few cases in children have been reported with SEL [4,5]. In this case report, a girl with obesity had back pain. While investigating for the cause of obesity with back pain, the underlying pathology is found to be spinal epidural lipomatosis due to adrenal Cushing syndrome.

Case Presentation

A 14-year-old girl from Sindhuli, Nepal came with the complaints of gradual swelling of the whole body for 1 year. Swelling started from the face which progressed to chest, abdomen, then legs. She did not have any urinary complaints, yellowish discoloration of body, palpitation, or shortness of breath. There was no change in appetite but she had been gaining weight. She developed back pain 2 months back which was severe to such an extent that she could not properly stand or walk. She had not attained menarche. There was no history of intake of medicinal drugs, including herbal medications or trauma. There was no significant past or family history. On examination, she was alert with moon face, buffalo hump, hirsutism on the face, maculopapular rash over chest, purplish striae on the abdomen, arms, and thighs. Her weight (40 kg), height (1.55 m), and body mass index (16.6 kg/m2) were on the 25th percentile, 5th percentile, 3–10th percentile, respectively, of National Center for Health Statistics. Her sexual maturation rate was stage 2. Regarding her vitals, pulse rate and respiratory rates were normal but her blood pressure was 160/140 (more than the 99th percentile for height-for-age). Her upper segment by lower segment was 0.78 which is lower than normal. No organomegaly was found on abdominal palpation. Her higher mental function was normal and cranial nerves were intact. Bulk and tone in all limbs were normal but there was tenderness in both hip joints with limitation in range of movement and power. There was no local rise in temperature. Deep tendon reflexes in both upper and lower limbs were brisk. Ophthalmology evaluation showed bilateral entropion with trichiasis and bilateral early posterior subcapsular cataract but no evidence of hypertensive retinopathy. In a laboratory investigation, 24 hours urinary cortisol was normal, 1.144 µg/day in 1.1 litre urine, that is, 104 µg/dl, which

is equivalent to 287.04 nmol/day (reference range 12.9–253 µg/dl). Midnight cortisol level was high, 27.5 µg/dl (reference range < 4.4 µg/dl). There was not adequate suppression of cortisol in overnight dexamethasone suppression test done with a single dose—1 mg given at 11 pm. The test showed cortisol level at 8 am to be 29.3 µgm/dl, which also indicated patient had Cushing syndrome. The two-step dexamethasone suppression test with low and high dose administration of dexamethasone, 30 and 120 µg/kg/24 hour in four divided doses, on consecutive days was also done to differentiate between a central and an adrenal cause of hypercortisolemia. With low and high dose dexamethasone test, serum cortisol level was 29.3 and 29 µg/dl, respectively. As serum cortisol was not suppressed by high dose dexamethasone, adrenocorticotropin hormone ACTH was sent. ACTH level checked with ELISA-EDTA (Enzyme-linked immunosorbent assay-Ethylenediaminetetraacetic acid) came normal, 5.41pg/ml (reference range 0–46pg/ml), so ACTH independent Cushing syndrome was confirmed and the adrenal tumor was suspected. Serum calcium, sodium, and potassium were also normal. Random blood sugar was 190 mg/dl with HbA1c of 6.6%. Twenty-four-hour urinary Vanillylmandelic acid was normal, 1.4 mg (reference range <13.6mg/24hr). Thyroid function test was normal. X-ray of the lumbar spine showed diffuse osteopenia in vertebral bodies, penciling of end plates with biconcave appearance. There was a decreased height of thoraco-lumbar vertebra body giving rise to fish-mouth appearance, probably osteoporotic pathology. X-rays of the pelvis and bilateral hip joints were normal. Magnetic resonance imaging (MRI) of the spine (Figure 1) showed thickened posterior epidural fat extending from T2 to S2 level with effacement of posterior perimedullary cerebrospinal fluid space and mild compression of the spinal cord. Maximum thickness of fat was up to 11 mm, which is suggestive of spinal epidural lipomatosis. Computed tomography (CT) scan of the abdomen (Figure 2) to find the etiology of steroid excess showed a round well-defined homogeneously enhancing soft tissue density lesion of size 2.2 × 2.1 × 2 cm, arising from the superior aspect of the right suprarenal gland with absolute and relative washout consistent with a benign right adrenal adenoma. The lesion was abutting the liver and inferior venacava medially with the maintained fat plane. No fatty component or calcification was seen within the lesion. Left adrenal gland was normal in size. In view of normal ACTH and no suppression of plasma cortisol on high dose dexamethasone, a provisional diagnosis of ACTH-independent Cushing syndrome, probably due to adrenal tumor causing SEL and myelopathy was made. The patient was managed with nifedipine for hypertension and regular monitoring of blood sugar was done. Exploratory laparotomy was carried out. Intraoperatively, a 3 × 3 cm right adrenal adenoma (Figure 3) was detected and right

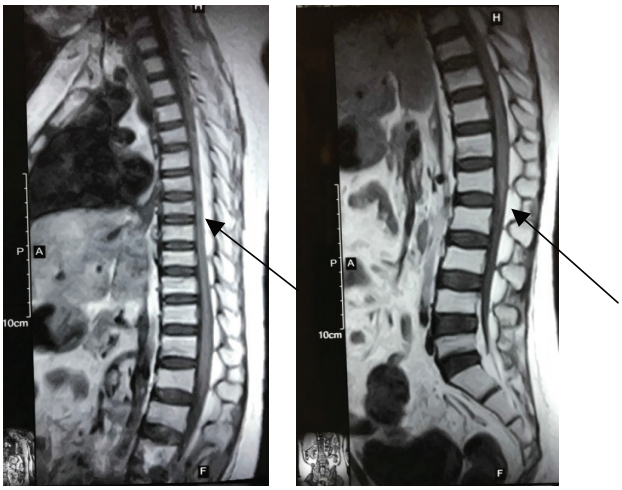

Figure 1. *Sagittal T1-weighted MRI of the spine image revealed the increased amount of epidural fat.*

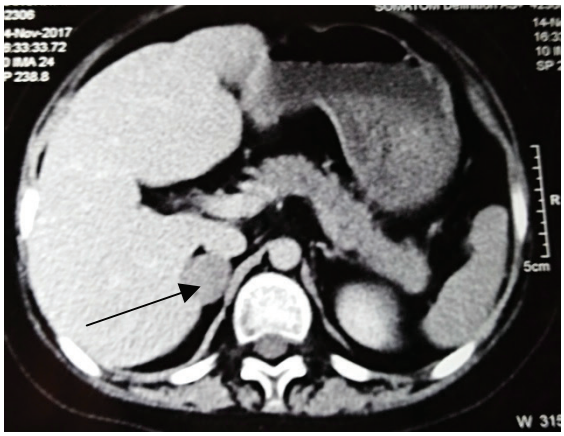

Figure 2. *CT abdomen showing homogeneously soft tissue density lesion in the superior aspect of right suprarenal gland consistent with right adrenal adenoma.*

Figure 3. *Postoperative image of the excised adrenal adenoma.*

adrenalectomy done. Histopathology report of resected right adrenal gland showed tumor composed of small nests of tumor cells with eosinophilic and clear cytoplasm and round nuclei and inconspicuous nucleoli, no capsular invasion suggestive of right adrenal adrenocortical adenoma. Postoperative period was uneventful. Intravenous steroid was given in tapering dose for total 5 days and

stopped following adrenalectomy. Intravenous hydrocortisone 100 mg was given before and after adrenalectomy on the day of surgery and then subsequently 200 mg on the first, 100 mg, 75, 50, and 25 mg twice a day on second, third, fourth, and fifth day, respectively, then equivalent oral prednisolone was given in tapering dose. As oral hydrocortisone is not available in Nepal, maintenance dose with prednisolone was continued [6]. Analgesic was continued for her back pain. Antihypertensive drug was titrated and stopped as her blood pressure gradually normalized after a week of surgery. Her blood sugar also normalized on the 14th day of post-surgery. She could walk with no back pain on the 20th postoperative day. She was then discharged with a weight loss diet plan with follow-up plans, including investigation such as morning cortisol level to see adrenal recovery.

Discussion

SEL is rare in children with few reported cases as young as in 5-year-old girl [5]. The underlying etiology for SEL may be exogenous glucocorticosteroids, epidural glucocorticosteroid injections, endogenous Cushing syndrome, hypophysal tumors, obesity, or may be idiopathic [2]. The adolescent girl described in a report presented with signs of hypercortisolism as centripetal weight gain, striae, hypertension, bilateral early posterior subcapsular cataract, hyperglycemia, and back pain with exaggerated reflexes in lower limbs. Back pain and exaggerated lower limb reflexes are due to SEL extending from thoracic to sacral region. Features of compressive myelopathy such as back pain, followed by lower extremity weakness, sensory changes as radicular pain, numbness, and paresthesias are the common presenting features of spinal epidural lipomatosis depending on the site of the lesion [1]. In our patient, X-ray of the lumbar spine was done which showed diffuse osteopenia in vertebral bodies. On further investigation, the MRI of the spine revealed thoracosacral epidural lipomatosis. The preferred imaging modality for SEL is MRI of the affected spinal segments [7]. The cause behind SEL was increased adrenocortical steroid production by right adrenal adrenocortical adenoma, which was found in CT abdomen. Adrenocortical tumors are rare in children. It represents <0.5% of all childhood neoplasm. The first year of life and between the ages of 9 and 16 years are two peaks of incidence of adrenal cortical neoplasm. The frequent manifestation of the adrenal tumor is Cushing syndrome but feminizing and masculinizing syndromes are less common [8]. Hypertension is due to either aldosteronism or hyper-reninemia. Refractory hypertension requiring more than three antihypertensive agents and hypokalemia are common hallmarks of primary aldosteronism attributable to an aldosteronoma [9]. The girl described had spontaneous normalization of blood pressure after right adrenalectomy. In our patient, adrenal adenoma was the underlying etiology for clinical manifestations and investigatory findings of steroid excess and epidural fat deposition causing SEL. The management of the SEL is removing the underlying cause of excess steroid production, weight loss and if not resolved, opt for surgical intervention as decompressive laminectomy and resection of epidural adipose tissue [1]. The reported girl had symptomatic relief of back pain and was able to walk. There was normalization of hyperglycemia and blood pressure without medication.

Conclusion

Only a very few cases describe SEL related to endogenous Cushing syndrome. However, while treating a patient with SEL, such cause should be excluded. The management involves surgical intervention as removal of adrenal tumors followed by supportive care. If not improved or when neurological symptoms worsen, resection of epidural fat may be necessary.

List of Abbreviations

SEL	Spinal epidural lipomatosis
ACTH	Adrenocorticotropin hormone
MRI	Magnetic resonance imaging
CT	Computed tomography
EDTA	Ethylenediaminetetraacetic acid
ELISA	Enzyme-linked immunosorbent assay

Consent for publication

Yes, informed written consent was taken from the guardian of the patient on patient's behalf.

Ethical approval

Not required.

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Summary of the case

Patient (gender, age)	1	Female, 14 years
Final diagnosis	2	Spinal epidural lipomatosis due to adrenal cortisol secreting adenoma
Symptoms	3	Gradual swelling of whole body with back pain
Investigations	4	Biochemical, hormonal, radiological, and histopathology
Medications	5	Symptomatic treatment
Clinical Procedure	6	Surgery to remove adrenal tumor
Specialty	7	Pediatrics