Idiopathic and glucocorticoid-induced spinal epidural lipomatosis

SOHEIL F. HADDAD, M.D., PATRICK W. HITCHON, M.D., AND JOHN C. GODERSKY, M.D.

Division of Neurosurgery, University of Iowa Hospitals and Veterans Administration Medical Center, Iowa City, Iowa

Pathological overgrowth of the epidural fat in the spine has been described and reported nearly exclusively in patients either with Cushing's syndrome or on chronic intake of glucocorticoids for a variety of clinical disorders. The authors report four patients with documented spinal lipomatosis (three pathologically and one radiologically). Only one of these patients received corticosteroids, and none had an underlying endocrinological abnormality. All four patients were adult males with a mean age at onset of symptoms of 43 years (range from 18 to 60 years). The symptoms ranged from simple neurogenic claudication and radicular pain to frank myelopathy. Myelography followed by computerized tomography were instrumental in the diagnosis of the first three patients; the fourth was diagnosed by magnetic resonance imaging. The thoracic spine was involved in two cases and the lumbosacral area in the other two. The different treatment modalities were tailored according to the symptomatology of the patients. These included weight reduction of an overweight patient with minimal neurological findings in one case and decompressive laminectomy and fat debulking to achieve adequate cord decompression in the remaining three cases. Two patients improved significantly, the condition of one stabilized, and the fourth required a second decompression at other spinal levels. The various modalities of treatment and their potential complications are discussed.

KEY WORDS • epidural lipomatosis • Cushing's syndrome • laminectomy • spinal cord compression

Pathological overgrowth of epidural fat in the spine has been almost exclusively reported in the setting of Cushing's syndrome. It has been seen with increasing frequency in transplant recipients and patients on immunosuppressive doses of glucocorticoids for the control of various diseases such as systemic lupus erythematosus, radiation pneumonitis, rheumatoid arthritis (adult and juvenile forms), Graves' disease, chronic active hepatitis, dermatomyositis, nephrotic syndrome, sarcoidosis, periarteritis nodosa, and asthma.1,5,11,18-21,22,23,26 There exists a group of patients who suffer from symptomatic epidural lipomatosis in the absence of any obvious endocrinopathy, however; these have been rarely reported in the literature.3,18,22 We report here our experience with four consecutive patients suffering from epidural spinal lipomatosis, three of whom did not have a known predisposing factor.

Clinical Material and Methods

The charts, radiological studies, and operative reports (where it applies) of all patients seen by the Neurosurgery Service at the University of Iowa and Veterans Administration Medical Centers with the diagnosis of symptomatic epidural spinal lipomatosis over the last 5 years were reviewed. Four patients were encountered, all were males with a mean age at onset of symptoms of 43 years (range from 18 to 60 years). The mean duration of symptoms before diagnosis was 1.4 years. The following reports of those cases illustrate the various symptomatologies, diagnostic procedures, treatment modalities, and outcomes in this group. Three patients underwent surgery and in these the compressing fat had the appearance of normal adipose tissue on histological examination.

Case Reports

Case 1

This 60-year-old man with known systemic lupus erythematosus was receiving 9 mg of prednisone per day when he started complaining of disabling bilateral leg numbness and pain while walking. He was neurologically intact and did not have any of the stigmata of
Spinal epidural lipomatosis

Cushing's syndrome. A myelogram of the lumbosacral spine followed by a computerized tomography (CT) scan revealed severe stenosis from L-1 to S-1 secondary to epidural lipomatosis. The T₁-weighted sequence of a magnetic resonance (MR) study corroborated the finding of a circumferential hyperintense lesion compressing the lumbar thecal sac (Fig. 1). A myelogram showed diffuse stenosis in the lumbar area (Fig. 2 left). Decompressive surgery was undertaken. For fear of destabilizing the lumbar spine, the laminectomies and fat debulking were limited to L-1, L-2, and L-5 (sites of the worst stenosis). The overgrowth of the nonencapsulated epidural fat was circumferential but worse over the posterior aspect of the thecal sac. The sagittal T₁-weighted MR image obtained postoperatively demonstrated residual stenosis at the nonoperated L-3-4 level, but there was adequate decompression at the previously operated levels, namely L-1, L-2, and L-5 (Fig. 2 right).

The patient did well postoperatively with immediate and significant improvement of his symptoms that lasted for 8 months. Shortly thereafter his symptoms recurred with worsening neurogenic claudication. Investigation revealed persistent stenosis at the L-3 and L-4 levels. Subsequently, he underwent L-3 and L-4 laminectomies and fat debulking with resolution of his symptoms. The radiological studies in Fig. 2 suggest that there was no regrowth of adipose tissue once it had been adequately debulked.

Case 2

This 40-year-old obese man presented with a 2-year history of right T-5 radicular pain. His medical history was negative for endocrinopathy, and he had not been receiving medications prior to the onset of his symptoms. On examination he had bilateral Babinski signs and lower-extremity hyperreflexia, without evidence of weakness, sensory changes, or bowel or bladder dysfunction. Magnetic resonance imaging of the spine

Fig. 1. Case 1. Axial T₁-weighted magnetic resonance image revealing a hyperintense lesion, consistent with fat, compressing the thecal sac circumferentially (arrow).

Fig. 2. Case 1. Left: Preoperative lumbosacral myelogram showing diffuse stenosis. Right: Sagittal T₁-weighted magnetic resonance image of the lumbosacral area performed after the first operation (L-1, L-2, and L-5 laminectomies and fat debulking) demonstrating persistent stenosis at L-3-4 (arrow) and adequate decompression at L-1, L-2, and L-5.
revealed epidural lipomatosis extending from T-1 to T-10, with evidence of cord compression (Fig. 3). He was placed on a weight-reducing diet that resulted in a 30-lb weight loss over the 1st year. During that time his radicular pain improved significantly and his neurological examination returned to normal. The small but noticeable reduction of the epidural fat seen on his most recent scan was enough for his symptoms to resolve. The posterior subarachnoid space is now open and he remains asymptomatic 2 years following diagnosis.

Case 4

This 54-year-old moderately obese man had a 2-year history of low-back pain radiating down the right leg. His medical history was significant for a chronic right-ankle osteomyelitis followed by fusion of that same joint; he had no endocrinopathy and was not receiving steroids. Examination demonstrated diffuse weakness of the right leg (4/5) accompanied with a long-standing sensory loss below the right knee. Lumbosacral myelography followed by CT revealed an extradural mass of fat density compressing the thecal sac at the L5–S1 level. An L5–S1 decompressive laminectomy and normal-appearing nonencapsulated epidural fat compressing the thecal sac. The fat was debulked and an intradural exploration did not reveal any pathology. The patient’s symptoms improved and he remains neurologically stable 4 years after surgery. However, he had to undergo repair of lumbar pseudomeningoceles at 9 and 28 months postoperatively.

Discussion

Glucocorticoid-Induced Lipomatosis

Symptomatic spinal epidural lipomatosis has been well documented in the literature. The majority of cases have been reported in patients receiving long-term steroid therapy similar to our first case.1,5,11,13,21,23,28 One other case was reported by Toshniwal and Glick29 in a hypothyroid patient without Cushing’s syndrome. Recognition of the real cause of the problem is often delayed. The disease process for which the steroid was prescribed is implicated and the patient is thought to have either a myelopathy or a neuropathy secondary to such disorders as systemic lupus erythematosus, epidural abscess, rheumatoid arthritis, or lymphomatous meningeal spread.2,6,12,13 This entity has also been increasingly recognized in transplant recipients, among
Spinal epidural lipomatosis

TABLE 1
Clinical summary of six cases of idiopathic epidural spinal lipomatosis*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Sex, Age (yrs)</th>
<th>Duration of Symptoms (yrs)</th>
<th>Symptoms</th>
<th>Findings</th>
<th>Wt. (kg)</th>
<th>Ht. (cm)</th>
<th>Treatment</th>
<th>BMI (kg/sq m)</th>
<th>Obesity</th>
<th>Level</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Badami &amp; Hinck, 1982</td>
<td>F, 33</td>
<td>6</td>
<td>LE weakness, bowel &amp; bladder incontinence</td>
<td>T-6 sensory level, sciatric paraesthesia</td>
<td>174</td>
<td>?</td>
<td>laminectomy &amp; fat debulking</td>
<td>++</td>
<td>T1-10</td>
<td>no change, died 75 days postoperatively secondary to pulmonary complications</td>
<td></td>
</tr>
<tr>
<td>Quint, et al., 1988</td>
<td>M, 34</td>
<td>1</td>
<td>T-6 radicular pain</td>
<td>normal</td>
<td>95</td>
<td>167.5</td>
<td>laminectomy &amp; fat debulking</td>
<td>?</td>
<td>T4-8</td>
<td>improved</td>
<td></td>
</tr>
<tr>
<td>Stambough, et al., 1989</td>
<td>F, 54</td>
<td>0.6</td>
<td>LE pain, back pain, neurogenic claudication</td>
<td></td>
<td>89.5</td>
<td>175</td>
<td>laminectomy &amp; fat debulking</td>
<td>3.1</td>
<td>T6-8</td>
<td>improved pain, neurologically normal</td>
<td></td>
</tr>
<tr>
<td>Haddad, et al., 1991</td>
<td>M, 40</td>
<td>2</td>
<td>T-5 radicular pain</td>
<td>LE hyperreflexia, bilateral Babinski signs</td>
<td>94</td>
<td>172</td>
<td>weight reduction</td>
<td>3.1</td>
<td>T1-10</td>
<td>improved pain, neurologically normal</td>
<td></td>
</tr>
<tr>
<td>M, 18</td>
<td>0.6</td>
<td>back pain, LE numbness, gait difficulty</td>
<td>T-6 sensory level, LE weakness</td>
<td></td>
<td>94</td>
<td>167.5</td>
<td>laminectomy &amp; fat debulking</td>
<td>2.9</td>
<td>T6-8</td>
<td>improved pain, neurologically normal</td>
<td></td>
</tr>
<tr>
<td>M, 54</td>
<td>2</td>
<td>rt LE pain &amp; decreased sensation</td>
<td>rt LE weakness</td>
<td></td>
<td>82</td>
<td>172</td>
<td>laminectomy &amp; fat debulking, intra-dural exploration</td>
<td>2.7</td>
<td>L5-S1</td>
<td>improved pain, stable; pseudomeningocele necessitating 2 reoperations</td>
<td></td>
</tr>
</tbody>
</table>

* LE = lower extremity; BMI = body mass index in kg/sq m; ? = no available data. Obesity: ++ = obese; + = moderately obese.

whom case reports of patients with progressive spinal dysfunction secondary to long-term steroid usage abound. It is thought that the spinal epidural fat responds to steroids by progressive hypertrophy in a manner similar to the centripetal fat deposition seen with Cushing's syndrome, affecting among other locations the mediastinum and pelvis. This was clearly shown by Buthiau, et al., who were able to document an increase in the spinal epidural fat during steroid therapy in an elderly lady with dermatomyositis. Steroid reduction and surgical decompression have been the mainstay of therapy either in combination or separately, with variable but generally favorable outcomes.

Idiopathic Epidural Lipomatosis

Symptomatic epidural spinal lipomatosis has rarely been reported without concomitant endocrinopathy. There are only six cases of well-documented idiopathic symptomatic epidural spinal lipomatosis, including our three cases (Table 1). Four of the patients were males, with a mean age at diagnosis of 38.8 years (range 18 to 54 years); the mean duration of symptoms before diagnosis was 2 years. The lipomatosis consistently involved two or more spinal levels (mean of 5.3 levels), affecting most commonly the thoracic spine and (in two cases) the lumbosacral spine, but sparing the cervical segments. Five patients were obese, and there were no available data on the habitus of the last patient. The four patients who had both weight and height measurements available had a mean body mass index (weight in kilograms divided by the square of the height in meters) of 30.7 kg/sq m with a range of 27.7 to 34.1 kg/sq m; this was greater than the 27.5 kg/sq m which is the National Institutes of Health's definition of obesity. The fifth patient, who weighed 174 kg, was described as being "morbidly obese;" her height was not available. Symptoms included simple radiculopathy and neurogenic claudication in five patients, a combination of weakness and gait difficulty in three, and frank paraplegia in one.

Two modalities of treatment were employed; the first and most common consisted of laminectomy and fat debulking and the second was weight reduction. The former gave excellent results in four patients experiencing mild lower-extremity weakness and pain; however, the patient who was paraplegic did not benefit from the operation and died 75 days postoperatively secondary to pulmonary complications. The patient treated with a weight-reduction diet lost 30 lbs in a year and had complete resolution of his T-5 radicular pain and myelopathic signs.

The only common etiological factor in all of these cases is their moderate to severe obesity (except for the patient report by Quint, et al., whose weight and
height were not mentioned) causing hypertrophy of the epidural fat and resulting in compressive symptoms. The fact that one patient improved both symptomatically and neurologically upon losing weight without other therapy gives credibility to this hypothesis and opens new avenues in the treatment of this recently recognized complication of obesity. Dietary control may be attempted as the first line of management of the patient with mild stable symptoms. Laminectomy and fat debulking is an effective treatment.

Clinical judgment should be utilized to determine whether surgery is appropriate, as in neurologically unstable patients or those in whom weight reduction cannot be achieved or fails to resolve incapacitating symptoms. The diagnosis of these cases has been made accurately with both CT and MR imaging. On CT scans fat is hypodense with values ranging from −80 to −120 Hounsfield units, and on MR images it is hyperintense on both T1- and T2-weighted images, obviating the need for operations for diagnosis only. The knowledge that this entity exists should help to decrease the incidence of intradural exploration and its accompanying potential complications in obese patients.

References

Manuscript received March 30, 1990. Accepted in final form July 9, 1990. Address reprint requests to: Souheil F. Haddad, M.D., Division of Neurosurgery, Department of Surgery, University of Iowa Hospitals and Clinics, Iowa City, Iowa 52242.