Leptomyelolipoma: analysis of 20 cases

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Leptomyelolipoma (lumbosacral lipoma) is a common form of spinal dysraphism. The deficits produced include sensory, motor, bowel, and bladder dysfunction, and vary in incidence between the pediatric and adult populations. Twenty patients treated surgically at the Mount Sinai Hospital between 1972 and 1988 are reviewed. Fifty percent were 12 years of age or less and 50% were older than 18 years of age. The surgical approach was designed to accomplish untethering of the conus medullaris, debulking of the lipomatous mass compressing the cord, reconstruction of the dural canal, and reapproximation of the paraspinal muscles and lumbosacral fascia to prevent future trauma. Postoperatively, no patient experienced deterioration of neurological function. Of the symptomatic patients, 67% displayed dramatic improvement or became asymptomatic and 33% experienced stabilization of their deficits. The symptoms most resistant to surgical correction were orthopedic foot deformities and bowel dysfunction, whereas bladder dysfunction, motor weakness, and radiculopathies were most amenable to surgical therapy. Early surgical repair is recommended in these cases to forestall irreversible neurological damage.

KEY WORDS • leptomyelolipoma • lumbosacral lipoma • pathophysiology

LEPTOMYELOLIPOMA is the most common form of spinal dysraphism reported in many series. Although this entity was first reported more than a century ago, in the majority of cases data regarding leptomyelolipomas have been grouped with the category of spinal lipomas, which often do not demonstrate a dural defect. Some of this surgical literature was tabulated prior to the microsurgical era. Composing between 1% and 5% of all intramedullary spinal cord tumors, leptomyelolipomas usually involve an abnormally low-lying conus which is tethered by a lipomatous mass traversing a dural defect. Compression and tethering of the conus leads to progressive neurological deficits. Based on the excellent outcome reported in the recent surgical literature, early decompression and untethering of the spinal cord with reconstruction of the dural canal is advocated. This study is a retrospective review of 20 surgically treated patients with leptomyelolipoma.

Summary of Cases

Case Material

Between July, 1972, and December, 1988, 25 patients underwent microsurgical correction of spinal cord lipomas at the Mount Sinai Hospital. Two lipomas were thoracic and thus excluded, and an additional three cases could not be analyzed due to insufficient data. Of the remaining 20 patients, there were 13 males (65%) and seven females (35%), ranging in age from 7 months to 57 years (Table 1).

Clinical Presentation

All but one of the patients had cutaneous stigmata of spinal dysraphism (Table 2), accompanied by a low-lying conus medullaris with tethering of the cord by an intramedullary lipoma in all cases. Spina bifida was seen in 17 (85%) of the patients. At the time of presentation, 13 patients (65%) had gradually progressive deficits, five (25%) were neurologically intact, and two (10%) had stable fixed deficits (Table 3). Both of the latter patients were 1 year of age or less. The majority of patients were neurologically intact as infants. In the age range under 6 years old, three children (50%) were neurologically intact preoperatively, while only one child (25%) in the 6- to 12-year-old age range and one (10%) in the category older than 18 years were without deficit.

Four patients aged 12 years or less presented with cutaneous malformations. Motor weakness and sphincter abnormalities were equally common among the six
Leptomyelolipoma

TABLE 1
Age and sex distribution in 20 patients

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Total</th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt; 6</td>
<td>6-12</td>
<td>&gt; 18</td>
<td>No.</td>
<td>Percent</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>male</td>
<td>5</td>
<td>2</td>
<td>6</td>
<td>13</td>
<td>65</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>female</td>
<td>1</td>
<td>2</td>
<td>4</td>
<td>7</td>
<td>35</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>totals</td>
<td>6 (30%)</td>
<td>4 (20%)</td>
<td>10 (50%)</td>
<td>20</td>
<td></td>
<td></td>
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</tbody>
</table>

patients in this age group who had neurological deficits. Only one of the six symptomatic children complained of paresthesias. Two of the six patients had orthopedic foot deformities (talipes cavovalgus).

Of the nine symptomatic patients in the adult group (> 18 years old), six presented with sphincter abnormalities and four with radiculopathy or paresthesias. Although weakness and diminished sensation were universally present, these complaints were minor. Only one of nine symptomatic adults had an orthopedic deformity: namely, a talipes cavovalgus deformity which had been present since childhood.

A subcutaneous lipoma with an accompanying dimple, hypopigmentation, and hypertrichosis had been present in one neurologically intact adult patient since birth. At the age of 25 years, she sought medical advice when a blow to the back elicited transient paraplegia. Two other adult patients experienced several similar episodes dating back to childhood; however, they presented only after persistent neurological deficits appeared.

Abnormal reflexes were frequently found. Absent or hypoactive reflexes were five times as common as hyperactive reflexes (Table 4).

Surgical Results

Postoperatively, our patients were classified as improved, stabilized, or deteriorated. None of these cases deteriorated after surgery, consistent with the results of most other series.\cite{1,2,18,32,48} A mean postoperative fol-

TABLE 2
Dermal stigmata among 20 patients

<table>
<thead>
<tr>
<th>Cutaneous Stigma</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>lipoma</td>
<td>18</td>
</tr>
<tr>
<td>hyperpigmentation*</td>
<td>7</td>
</tr>
<tr>
<td>hypopigmentation</td>
<td>2</td>
</tr>
<tr>
<td>dimple or sinus</td>
<td>6</td>
</tr>
<tr>
<td>hypertrichosis</td>
<td>5</td>
</tr>
<tr>
<td>hemangioma</td>
<td>1</td>
</tr>
<tr>
<td>no cutaneous lesion</td>
<td>1</td>
</tr>
<tr>
<td>spina bifida</td>
<td>17</td>
</tr>
<tr>
<td>intramedullary tumor</td>
<td>20</td>
</tr>
</tbody>
</table>

* Hyperpigmentation included port-wine stain, café-au-lait areas, and pigmented nevi.

low-up period of 22 months was obtained in these patients (range 6 months to 12 years). All patients who were neurologically intact on presentation have remained so (Table 3).

Patients with gradually progressive deficits fared better with early surgical intervention. Three (75%) of four such patients 12 years of age or less improved postoperatively, while six (67%) of nine such patients older than 18 years of age improved. Diminished sensation was the symptom most frequently encountered at presentation, with motor weakness and sphincter abnormalities occurring less commonly (Table 5). Urinary
incontinence was most amenable to improvement. The findings most resistant to treatment were orthopedic foot deformities and abnormalities of rectal sphincter tone.

**Discussion**

**Embryology and Pathophysiology**

Closure of the medullary tube and ectodermal differentiation into neural and epithelial tissue occur between the 3rd and 5th week of intrauterine development. Errors in this differentiation can lead to central nervous system and cutaneous abnormalities categorized as spinal dysraphism. Leptomelyelolipomas are one group of dysraphic states. The posterior neuropore is the last region of the tube to fuse, and therefore the caudal end of the spine is the most common area in which to encounter dysraphism. In addition, it is known that the more severe the caudal dysraphism, the greater the affiliation with cranial dysraphic states and other congenital anomalies. Although Chiari malformations have been reported with leptomelyelolipoma in as many as 7% of some series, they are generally considered to be rare in these patients.

At the 15-mm stage of fetal development, the cord is a structure extending to the lower end of the sacrum. The process of tube closure and formation of the vertebral column by mesodermal investment of the spinal cord must be altered early in order to account for the partial dorsal myeloschisis and the presence of spina bifida (identified in 85% of our patients). It is the developmental process to this point which must give rise to leptomelyelolipoma. Alvord, et al., proposed that these abnormalities arise from a clone of lipomatous cells that has escaped normal controls. The presence of ependyma, neuroglia, smooth and striated muscle, and other elements in leptomelyelolipomas supports this contention.

In the case of leptomelyelolipomas, the conus is tethered and compressed by the lipoma and is unable to undergo its normal ascent. With progressive lengthening of the vertebral axis, greater traction is placed upon the cord, producing the low-lying conus medullaris. Leptomelyelolipomas produce neurological impairment by traction, compression, and direct transmission of force. Patients frequently become symptomatic during growth spurts. Childbirth, sexual intercourse, and bending during exercise have all been implicated in the precipitation of symptoms.

Although adipose tissue in leptomelyelolipomas does not undergo mitotic activity, the tumor does change in size, depending on the nutritional status of the individual. Patients may experience neurological deterioration with weight gain. Coexisting conditions which narrow the spinal canal will also increase the compressive effect of the lipomatous mass and exacerbate symptoms.

Transient paralysis due to a blow on the back is caused by the direct transmission of force to the lipoma by a water-hammer effect. Hilal and Keim and others demonstrated that tension on the conus caused impaired blood flow with resultant ischemia and spinal cord damage.

**Evaluation of Cases**

**Clinical Studies.** Leptomelyelolipoma can produce both upper and lower motor neuron deficits, mimicking a myriad of urological and orthopedic problems. Urinary tract infections may be the first symptom of the disease, along with the onset of an orthopedic foot deformity. Apparently neurologically intact patients may have abnormal electromyographic (EMG) studies, and EMG examination of the perineum and lower extremities is advisable in all patients.

Somatosensory evoked potentials (SSEP) monitoring may help to measure baseline neurological function.

**Neuroradiographic Studies.** Plain radiographs may offer details of bone in cases of spina bifida (seen in 100% of patients in many series) and other spinal abnormalities (Fig. 1). Although in the past the neuroradiographic mainstay has been metrizamide myelography followed by computerized tomography (CT)
Leptomyelolipoma

FIG. 2. Axial computerized tomography scans in two patients. Upper: Scan obtained through the midlumbar spine showing a lipoma, which is located posteriorly within the spinal canal (arrow). Lower: Scan obtained through the lower lumbar spine showing fatty tissue (arrow) which surrounds a cord-like structure (arrowhead) extending through a neural arch defect.

FIG. 3. Sagittal T₁-weighted magnetic resonance image showing a lipoma with high signal intensity (arrow) within the spinal canal, extending posteriorly into the subcutaneous soft tissues through a neural arch defect.

Scanning, this has been replaced by magnetic resonance (MR) imaging. Malis and Carem, et al., do not suggest myelography for fear of traumatizing the low-lying conus or an undiagnosed Chiari malformation. The use of CT scanning with contrast enhancement allows delineation of nerve root emergence and the lipoma-cord relationship (Fig. 2 upper) in a complementary fashion to the studies obtained by MR imaging. Other abnormalities, such as diplomyelia, thickened filum, diastematomyelia, dermoid cyst, and terminal hydromyelia may also be diagnosed by CT (Fig. 2 lower).

The usefulness of MR studies in visualizing cord lesions is well documented. On T₁-weighted spin-echo images, the lipoma shows a high signal intensity, the cord an intermediate intensity, and the cerebrospinal fluid (CSF) a low intensity (Fig. 3). On T₂-weighted images, the lipoma and cord show intermediate intensity, the lipoma being of equal or slightly lower intensity than the cord. In general, CSF shows high intensity on T₂-weighted images.

Axial images demonstrate a low-intensity band between the lipoma and the spinal cord. On sagittal images, the band is cephalad and anterior to the lipoma, separating it from the cord (Fig. 4 left). The low-intensity band has been likened to a halo, but is more accurately described as a chemical shift artifact. It has a characteristic appearance and occurs perpendicular to the direction of the frequency-encoding gradient and the interface of tissues with different chemical-shift properties. The greater the magnetic field, the greater is the intensity of this artifact. With leptomyelolipomas, this phenomenon is particularly fortuitous in providing exact demarcation at the lipoma-cord interface. This line assists in demonstrating the relationship of the nerve roots to this interface (Fig. 4 right).

Magnetic resonance imaging is particularly adept at revealing the presence of terminal hydromyelia, and allows for evaluation of the paraspinal structures; MR imaging is now the diagnostic method of choice for leptomyelolipoma.

Surgical Indications

All patients with leptomyelolipoma should undergo surgical correction. If the neoplasm is asymptomatic,
the goal is to prevent the onset of neurological symptoms. If it is symptomatic, the goal is to halt progression and alleviate deficits where possible. Bassett,\textsuperscript{10} recognizing the progressive nature of the disease, advocated early prophylactic surgery. Others have recommended surgical repair only after the onset of neurological deficit\textsuperscript{65} or for the repair of cosmetic deformity alone.\textsuperscript{50} Malis\textsuperscript{48} and others\textsuperscript{18,43} have observed that superficial removal of the mass leads to intense arachnoidal scarring with escalated neurological deterioration and the need for a more dangerous second exploration often resulting in a poor outcome. It has been advocated that surgery be delayed until 1 year of age in asymptomatic infants to allow technically easier removal. A sudden neurological deterioration has been described; four of these patients were under 6 months of age.\textsuperscript{50} It is our belief that all patients with leptomyelolipoma should undergo surgical correction as early as possible.

Only rarely do neonates demonstrate severe abnormalities. This situation has been encountered by other investigators,\textsuperscript{18,56} and is probably due to congenitally absent or malformed nerve roots. Many reports describe a neurological deficit increasing with age. Although neurologically intact patients aged in their 50's have been reported, this is rare.\textsuperscript{5,11,46} Our oldest patient without a deficit was 25 years old.

Anatomical Considerations

Leptomyelolipomas have been classified into four types, as follows.\textsuperscript{18,42,62}

\textit{Type 1 (Dorsal Type).} In Type 1, the lipomatous mass inserts itself directly into the dorsum of the cord. If the mass is not situated in the midline, it will deviate and rotate the cord toward the lipoma (Fig. 5). There is a small dural defect through which the lipoma passes. The dura and pia arachnoid fuse to the interface of the

![Fig. 4. Sagittal T₁-weighted magnetic resonance images. Left: A lipoma with high signal intensity (large arrow) is seen in the posterior spinal canal and a syringohydromyelia with low signal intensity (small arrow) is shown in the distal spinal cord. Right: In another patient, a lipoma (arrow) is seen lying anterior to a meningocele (arrowhead), which extends through a neural arch defect.](image)

![Fig. 5. Coronal T₁-weighted magnetic resonance image showing a lipoma on the left side of the lumbar spine (large arrow). The distal spinal cord is pulled to that side (small arrow).](image)
Leptomyelolipoma

lipoma and cord, but the sensory nerve roots emerge anterior to the interface and none of the nerve roots run through the lipoma. The nerves that emerge at this level are shorter than normal and demonstrate abnormal angulation.

Type 2 (Caudal Type). Type 2 lipoma traverses a defect in the most distal part of the dural sac. There is no fusion of the leptomeninges to the lipoma-cord interface. The lipoma extends cephalad through the subarachnoid space to engulf the tip of the conus. The rootlets run within the fibrous fatty mass, usually in the anterolateral regions. The filum terminale may not be identifiable.

Type 3 (Transitional Type). In Type 3, the lipoma-cord interface resembles the dorsal lipoma superiorly, then extends around the conus inferiorly to resemble the caudal type. The leptomeninges fuse with the cord at the lipoma-cord interface and attach to the exiting nerve roots.

Type 4 (Filar Type). Type 4 is relatively rare and is not discussed by many authors. It consists of a short thick filum terminale widened by infiltrating fat and fibrous tissue. The fat may extend into the conus medullaris. Although the conus is tethered, the nerve roots have a normal configuration.

Surgical Approach

In the management of our patients we followed the protocol established by Malis. Monitoring of SSEP's was employed throughout positioning and the surgical procedure. The procedure was performed with the patient in the lateral oblique position and the side of greatest lipomatous involvement upward, allowing free movement of the thorax and abdomen without compression. The SSEP and sphincteric EMG findings were used to avoid damage to functional neural elements.

An omega-shaped flap was created so that the base lay inferiorly over the natal cleft, as previously depicted by Rosenblum, et al. The vascularity from the lumbar and gluteal arteries was incorporated into the flap. The flap design prevented urinary and fecal contamination of the wound, particularly in younger patients. A margin of adipose tissue was left adherent to the skin to prevent vascular compromise. Amputation of the lipoma at the point where it penetrated the lumbosacral fascia defect allowed reflection of the flap inferiorly. A laminectomy above the lipomatous insertion was needed to visualize the cephalic extent of the dural dehiscence and the transitional area between the lipoma and normal cord.

When the laminae were exposed, a vascular fibrous band might be seen bridging the gap between the laminae of the most superior bifid arches. If present, this band produced kinking of the herniating meningocele and cord in a configuration Malis has likened to a hairpin turn. This band should be tested for neural function prior to its division.

The remainder of the procedure entailed untethering the conus from lipomatous, leptomeningeal, and dural attachments, and debulking the lipomatous mass with decompression of the conus. The filum terminale was severed as it was shortened and acted as a further tethering agent, with occasional lipomatous infiltration. Reconstruction of the dural canal with a watertight closure provided ample space for CSF to flow around the cord, preventing adhesions with retethering. Reapproximation of muscle mass and lumbosacral fascia was performed over the canal to avoid future trauma to the repaired region.

If the dura was deficient, we preferred to use tensor fascia lata for reconstruction of the dural tube. If the canal was too tight, increases in the size of the residual adipose tissue might produce compression and neurological symptoms during periods of weight gain. A watertight closure will prevent accumulation of CSF in the subcutaneous space.

Conclusions

Leptomyelolipomas are common dysraphic lesions produced by aberrant neural tube closure during early embryological development. They may be associated with anomalies in other organ systems. The abnormal spinal development produces four discrete anatomical configurations of the lipoma-spinal cord interface. In our experience the natural history of the disease is that of progressive neurological deterioration with deficits arising during periods of rapid growth and weight gain. Deficits are produced by traction, compression, and direct transmission of force to the cord. Although improvement is common with surgical correction, only a minority of patients are cured and few will regain full function after deficits appear. In view of the recent advances in neuroimaging, intraoperative monitoring, and microsurgical techniques, early surgery is recommended for all patients.

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Leptomyelolipoma


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