Surgical treatment of the retethered spinal cord after repair of lipomyelomeningocele

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In a series of 75 patients with surgically treated lipomyelomeningoceles, the neurological condition of six patients deteriorated 6 months to 14 years after the operation due to repeat tethering of the spinal cord. The tethering resulted from postoperative dense adhesion between the cord and the overlying dura mater. Two of the six patients underwent conventional repeat untethering procedures, and the remaining four were successfully treated with a new surgical technique developed by the authors to prevent such dural adhesion. For this procedure, after complete untethering of the spinal cord, the lumbosacral cord is retained in the center of the dural sac by fine stay sutures between the pia mater of the conus medullaris and the ventral dura mater. In addition, the dura mater is tacked to the posterior arch which is reconstructed with bone grafts at one or two bifid vertebral levels. During a postoperative follow-up period of 1 to 3 years, no further deterioration has been observed and magnetic resonance studies have demonstrated a space filled with cerebrospinal fluid (CSF) around the lumbosacral cord. The authors conclude that long-term observation, both neurological and radiological, is essential even after successful repair of a lipomyelomeningocele. This new surgical procedure can maintain a CSF bath around the lumbosacral cord, thus preventing dural adhesion. Application of this technique will hopefully be beneficial in lipomyelomeningocele patients with a high risk of cord retethering after initial repair.

KEY WORDS • lipomyelomeningocele • tethered cord syndrome • spinal dysraphism • spina bifida

The aim of surgical treatment for a lipomyelomeningocele is to release the tethered spinal cord. Surgical procedures such as debulking of the lipomatous tissue, untethering of the spinal cord, and watertight closure of the dura mater with a graft are regarded as appropriate techniques to achieve this goal[1,3,8,10,13,17]. Reconstruction of the conus medullaris is also recommended[13,16]. Postoperative retethering, however, occurs not infrequently, even when these repair procedures have been carried out. Up to 10% of patients manifest late neurological deterioration due to a postoperative tethered spinal cord[2,8,10,13,15]. Furthermore, most surgically treated patients display a potential for cord retethering in magnetic resonance (MR) studies[6,12].

Between 1975 and 1988, we treated 75 patients with lipomyelomeningoceles using standard recommended surgical techniques[2]. Six of these patients showed neurological deterioration in the follow-up period and two of them were operated on in the usual manner for repeat untethering of the spinal cord. After the repeat untethering operation, however, MR studies showed a potential for further tethering of the spinal cord. We then developed a new surgical technique to prevent postoperative retethering and successfully applied this to the last four patients. The improved operative technique for lipomyelomeningocele repair is detailed in this report.

Summary of Cases

The new surgical technique was applied in four patients with postoperative retethering of the spinal cord. The age of the patients at the repeat operation ranged from 4 to 14 years. Table 1 summarizes their clinical course. Their lipomyelomeningoceles were classified on the basis of the operative findings at initial surgery (Table 2).[1]

Neuroradiographic Studies

The patients were evaluated by computerized tomography (CT) and MR imaging. The MR imager em-
ployed was a 0.5-tesla superconducting system. We used T₁- and T₂-weighted spin-echo pulse sequences. The repetition time (TR) and echo time (TE) were TR 600 to 1000 msec and TE 40 msec for the T₁-weighted spin-echo images, and TR 2000 msec and TE 120 msec for the T₂-weighted images.

The sagittal MR images at the time of deterioration showed a low-lying spinal cord running straight along the dorsal portion of the dural sac, with no space for cerebrospinal fluid (CSF) between the dorsal surface of the conus medullaris and the dura mater (Figs. 1A and 2A). Preoperative MR and CT studies suggested that the spinal cord was retethered by adhesion between the dorsal surface of the lumbosacral cord and the dura mater.

Operative Technique

Repeat untethering was performed in the standard manner (Fig. 3a and b). At two or three vertebral levels proximal to the bifid lamina, an osteotomy was performed with a chisel or a sagittal saw. Keeping the interconnected ligamentous structures, the laminar flap was turned upward and retracted. The rostral portion of the dura mater was opened. As the dural incision was extended caudally, the spinal cord was dissected from the dense scar tissue. A Malis bipolar coagulator and cutter system† and a contact Nd:YAG (neodymium: yttrium-aluminum-garnet) laser,‡ with a 50-μ sapphire tip and 1 to 3 W in output power, were used for dissection of the neural tissue. Electrical stimulation was utilized for intraoperative monitoring of the contractions of the segmental muscle group and of the anal sphincter.† † ‡ This monitoring helped to identify the nerve roots involved in the lipomatous and scar tissue. Further removal of the residual lipomatous tissue involving the conus medullaris reduced the volume of the conus, while the pial layer was preserved for reconstruction of the conus medullaris. Then, the pial flaps were approximated for reconstruction of the conus medullaris. Thus, the spinal cord was completely released from the transdural adhesion.

A new method was applied at this point. First, transposition of the spinal cord was carried out (Figs. 3c and 4A); the conus medullaris, dorsally and/or laterally shifted in the dural sac even after untethering of the cord, was kept near the center of the dural sac so that it was not in contact with the dura mater. The dentate

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† Bipolar coagulator and cutter system manufactured by Codman & Shurtleff, Randolph, Massachusetts.
‡ Contact ND:YAG laser manufactured by SLT Japan, Tokyo, Japan.
Retethered spinal cord

ligaments, if present, were divided close to the dura mater two or three vertebral levels rostral to the conus medullaris. The end of each spinal cut was sutured to the dura mater. A couple of stay sutures were also applied between the thick pial layer of the ventrolateral surface of the conus and the ventral dura mater. When, after untethering, the lumbosacral spinal cord was shifted to the right in the dural sac, stay sutures for transposition were applied mainly in the left ventrolateral surface of the conus medullaris. When the cord was shifted to the left, stay sutures were applied mainly in the right ventrolateral surface of the conus. For these stay sutures, 8-0 or 9-0 monofilament nylon thread was used. Thus, the conus that had been adherent to the surrounding dura mater was maintained around the center of the dural sac, and kept away from the surrounding dura mater.

After transposition of the spinal cord, the dural sac was reconstructed with a large patch graft of fascia lata and the dura mater was closed in watertight fashion. The laminar flap was brought back to its anatomical position.

A new technique was also employed in the extradural space of the bifid vertebral levels. Remnants of the bifid laminae on both sides were exposed at one to three vertebral levels, usually at levels L-4, L-5, and S-1. For laminoplasty, bone grafts of the rib or iliac bone were modified to resemble a lamina (Fig. 3d), and were then approximated to the exposed remnants of the laminae. The underlying dura mater, after repair with a graft, was sutured to the newly constructed posterior arches (Figs. 3e and 4b). These tenting sutures permitted sufficient CSF space to be preserved behind the conus medullaris which might otherwise readhere to the surrounding dura mater.

Operative Findings

All four patients had dense transdural adhesion between the dorsal and lateral surfaces of the lumbosacral cord and the overlying dura mater, even though two (Cases 3 and 4) had undergone reconstruction of the conus medullaris at the initial repair. The adhesion anchored the spinal cord in an abnormally low position, shifting it posteriorly and/or laterally in the dural sac and rotating it. The spinal nerve roots often ran through the dense scar tissue around the conus and/or through the residual lipomatous tissue involving the conus medullaris. The safely dissectable plane of scar tissue between the spinal cord and the dura mater was so narrow that the neural tissue was dissected only with great difficulty. None of the patients exhibited a defect of the dura mater or a thick filum terminale. The spinal cord was successfully untethered in all four patients. However, even after the cord was released from the transdural adhesion, the conus medullaris remained shifted dorsally and/or laterally in the dural sac by the nerve roots. These roots emerging from the conus were still tight and they seemed to anchor the spinal cord.

Clinical Course

Table 1 summarizes the clinical results. One patient (Case 1) manifested no further deterioration, two (Cases 2 and 3) showed transient bladder dysfunction and motor weakness of the legs, and the fourth (Case 4) showed transient motor weakness in the legs with improvement of sensory disturbance.
Neuroradiographic Findings

In the late postoperative period, MR studies disclosed a CSF space between the dorsal surface of the conus medullaris and the overlying dura mater; the course of the spinal cord through the dural sac was also seen to be relaxed and less straight (Figs. 1B and 2B). These findings indicated no significant adhesion between the spinal cord and the surrounding dura mater and that the cord remained untethered even in the late postoperative period.

Discussion

Retethering of the Spinal Cord

Retethering of the spinal cord is far from a rare postoperative complication of lipomyelomeningoceles after appropriate initial repair. The original procedure usually consists of debulking of the lipomatous tissue, untethering of the cord, reconstruction of the conus medullaris, and duraplasty. In the present series, postoperative tethered cord syndrome developed in six of 75 lipomyelomeningocele patients after initial surgical repair. The first two patients underwent surgery for repeat untethering by the standard procedure and the later four patients described here underwent a new technique to prevent further cord tethering. The rate of postoperative tethering was similar to that reported in other recent studies. The incidence of this complication, neurosurgeons should realize that retethering of the spinal cord even after satisfactory repair is not at all uncommon. The radiological and operative findings in such patients indicate that late deterioration due to tethering results from dural adhesion. Thus, long-term neurological and radiological observation is essential.

Operative Procedure

Standard surgical procedures can provide release of the spinal cord from the dural adhesion only during surgery; untethering does not always last long after surgery. The spinal cord tends to attach to the dorsal dura mater because the conus medullaris is still shifted dorsally in the dural sac even after the spinal cord is released from the adhesion. Therefore, we developed

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Present Age (yrs), Sex</th>
<th>Age at Onset</th>
<th>Initial Symptom</th>
<th>Age at Initial Op</th>
<th>Type of Lipoma*</th>
<th>Interval Between Ops</th>
<th>Second Presentation</th>
<th>Outcome</th>
<th>Follow-Up Period (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11, F</td>
<td>at birth</td>
<td>urinary incontinence</td>
<td>4 mos</td>
<td>Ib</td>
<td>8 yrs</td>
<td>urinary incontinence</td>
<td>unchanged</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>16, F</td>
<td>at birth</td>
<td>subcutaneous lipoma</td>
<td>6 mos</td>
<td>IIa</td>
<td>14 yrs</td>
<td>foot deformity</td>
<td>unchanged</td>
<td>2.5</td>
</tr>
<tr>
<td>3</td>
<td>12, F</td>
<td>1 yr</td>
<td>weakness of legs</td>
<td>4 yrs</td>
<td>IIIa</td>
<td>7 yrs</td>
<td>foot deformity</td>
<td>unchanged</td>
<td>1.5</td>
</tr>
<tr>
<td>4</td>
<td>5, F</td>
<td>at birth</td>
<td>weakness of legs</td>
<td>1 mo</td>
<td>Iib</td>
<td>4 yrs</td>
<td>foot deformity, urinary incontinence</td>
<td>improved</td>
<td>1</td>
</tr>
</tbody>
</table>

* Classification according to Hakuba, et al.; see Table 2.
Retethered spinal cord

TABLE 2

<table>
<thead>
<tr>
<th>Type</th>
<th>Classification of lipomyelomeningoceles*</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>anomalous type: this type is accompanied by other severe spinal malformations</td>
</tr>
<tr>
<td>II</td>
<td>extraspinal type: the spinal cord protrudes out of the spinal canal via a spinal bursa</td>
</tr>
<tr>
<td>IIa</td>
<td>protruded spinal cord curves back into the spinal canal</td>
</tr>
<tr>
<td>IIb</td>
<td>conus medullaris protrudes through a dorsocaudal defect and fuses with the extraspinal lipoma</td>
</tr>
<tr>
<td>IIc</td>
<td>conus medullaris protrudes outside the spinal canal, accompanied by the dural tube, and fuses with the extraspinal lipoma</td>
</tr>
<tr>
<td>III</td>
<td>intraspinal type: the spinal cord stays within the spinal canal</td>
</tr>
<tr>
<td>IIIa</td>
<td>conus medullaris fuses with the intraspinal lipoma through a defect of the caudal dural sac</td>
</tr>
<tr>
<td>IIIb</td>
<td>lipoma inserts broadly into the spinal cord dorsal to the dural root entry zone</td>
</tr>
<tr>
<td>IIIc</td>
<td>a fibrolipomatous stalk attaches to the spinal cord</td>
</tr>
<tr>
<td>IV</td>
<td>epidural type: a fibrous band originating from the epidural lipoma is connected to the spinal cord</td>
</tr>
</tbody>
</table>

*Classification according to Hakuba, et al.* Untethering of the spinal cord is difficult in Type I, IIa, IIb, and IIia cases.

Timing of Surgery

Most neurosurgeons believe that patients with a lipomyelomeningocele must undergo initial repair as early as possible, even if they have no apparent neurological deficits. As to the timing of repeat surgery for untethering, the patients in whom retethering of the cord was suspected from the MR images were operated on as soon as neurological deterioration was recognized.

Case Selection

There were two reasons for limiting the repeat operation to the patients with possible retethered spinal cord. One was that dissection of the neural structures from the dense scar tissue presented a risk of injuring the vulnerable spinal cord. In fact, three of the four patients operated on displayed transient postoperative deterioration, most likely due to minor injury of the neural structure caused by manipulation during surgery. The other reason was that neither MR nor CT studies were able to indicate clearly which candidates should undergo the repeat untethering procedure. They did not differentiate tethered spinal cords from untethered low-positioned spinal cords. Tethered cords require untethering, whereas low-positioned cords, which are associated with the development of dysgenetic spinal cords, do not. At present, only the neurological symp-toms and signs can be used to differentiate between these two pathological conditions. Thus, we conclude that patients with a possible tethered cord as shown on an MR image should undergo repeat surgery as soon as further neurological deterioration is found.

Lumbosacral lipoma with occult spinal dysraphism is classified on the basis of anatomical investigation or surgical considerations. Based on the operative findings at initial repair, untethering of the spinal cord is quite difficult in patients with lipomas of Types I, IIa, IIb, and IIa (Table 2). The patients in the present series had lipomas of either Type IIb or Type IIIa (Table 1), which suggests that such patients run a larger risk of a postoperative tethered cord. We will use the new technique at initial surgery for patients with a lipoma of Types I, IIa, IIb, or IIa, because releasing a tethered cord is much more difficult at a second intervention than at the first operation.

Tethered cord syndrome is seen in patients suffering dysraphism. In patients with myelomeningocele, the shallow spinal canal, the large mass of neural plaque, and the paucity of the dura mater are responsible for postoperative tethering of the cord. The new technique introduced in this study is applicable to cases with various spinal diseases which present a large risk of a tethered cord.

Conclusions

Retethering of the spinal cord after appropriate repair is not at all uncommon. This complication usually results from postoperative dural adhesion. Long-term
observation, both neurological and radiological, is mandatory after repair of a lipomyelomeningocele. The new surgical procedure reported here can maintain a CSF bath around the lumbosacral cord, preventing dural adhesion. Application of this technique will be beneficial in lipomyelomeningocele patients with a high risk of cord re tethering after initial repair.

References

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