Tethered Cord Syndrome; Surgical Indication, Technique and Outcome

Objective: The authors tried to reveal some unique features of lipomyelomeningocele (LMMC), including clinical presentation, factors precipitating onset of symptoms, pathologic entities of LMMC associated with tethered cord syndrome, and surgical outcome in LMMC patients.

Methods: Seventy-five patients with LMMC were enrolled in this study. Neuro-imaging and intraoperative findings allowed classification of LMMC into three Types. The patients were divided into two groups by age: A (61 patients), from birth to 3 years, and B (24 patients), from 3 to 24 years. For prevention of re-tethering of the cord, a mega-dural sac rebuilding procedure was performed in 15 patients.

Results: During a mean postoperative follow-up period of 4 years, the surgical outcome was satisfactory in terms of improved pain and motor weakness, but disappointing with reference to the resolution of bowel and bladder dysfunction. Among these 75 patients with LMMC, preoperative deficits were improved after surgery in 29 (39%), remained stable in 28 (37%), changed slightly in 13 (17%), and worsened in 5 (7%). Patients in group A achieved better outcomes than those in group B. Depending on the type of lesion, patients with types I and II LMMC have better outcomes than those with type III LMMC. Finally, re-tethering of the cord with neurological deterioration occurred in 4 (5.3%) of the 75 patients, but no re-tethering was found in the 15 patients who were recently treated with a mega-dural sac rebuilding procedure.

Conclusion: Our data continue to support the opinion that early diagnosis and optimal surgery are still essential for the treatment of patients with LMMC, since there is a high likelihood of residual neurological functions that can be preserved. Based on our surgical experience of Untethering and decompression of lipomas, a mega-dural sac repair is useful to prevent re-tethering of the cord.

KEY WORDS: Tethered cord syndrome - Lipomyelomeningocele - Untethering - Urodynamic function.

INTRODUCTION

Tethered cord syndrome (TCS) caused by a tight terminal filum has been known for more than 20 years and is recognized as a clinically important problem in children and adults. Tethered cord syndrome (TCS) is a complex clinicopathological entity that remains poorly understood. Nevertheless, it is well appreciated that optimal surgical outcome can be achieved only through aggressive and complete microsurgical detethering. Such an approach must be tempered with the understanding there may be a significant risk of neurological complications. To minimize the complications, it is imperative for the surgeon to appreciate the highly variable anatomy of the conus medullaris and cauda equina, particularly when these structures are distorted by secondary tethering lesion. TCS is typically associated with a low-lying conus medullaris (its tip is located below the L2 vertebra). The thickened terminal filum and subsequent tethering of the spinal cord are thought to be resulted from abnormal development of the filum during the retrogressive differentiation phase of secondary neurolution. Other features of spinal dysraphism such as myelomenigocele, lipomyelomeningocele (LMMC) and osseous spina bifida are commonly identified in these patients. TCS can include voiding dysfunction, lower extremities weakness, sensory abnormalities, pain, and gait disturbance. It is associated both with major spinal dysraphism such as LMMC and with more subtle structural abnormalities of the terminal filum, including thickening, fat infiltration, and high content of dense fibrous tissue.

The pathogenesis of the clinical syndrome in TCS is believed to arise from traction on the lower end of the spinal cord by a thickened terminal filum. Traction on the conus medullaris then leads to decreased blood flow and decreased oxidative metabolism, which may eventually cause the clinical symptoms and signs of the TCS. Its clinical features include neurological,
musculoskeletal, and urological abnormalities that may be improved after untethering of the filum terminale.

There are several corollaries to ischemic hypothesis in TCS. The first is that the mechanical properties of the cauda equine are important, so that a thicker terminal filum or one with fat infiltration of the filum causes higher tension\(^\text{26}\). Second such mechanical properties prevent normal spinal cord migration, which exacerbates this traction. Finally, asymmetrical growth of the vertebrae vis a vis the spinal cord is particularly evident during period of rapid growth, which explains the onset of neurological abnormalities at that time\(^\text{29}\).

Pang and Wilberger\(^\text{26}\) and others\(^\text{10}\) have described optimistic surgical outcomes. Others, however, have reported significantly less favorable outcomes after detethering surgery\(^\text{28}\). LMMC is one of the main causes of the tethered cord syndrome (TCS)\(^\text{5,11,16,20,22,27}\). Symptoms of LMMC are mainly related to traction on the lower spinal cord and include motor deficits, spasticity, sensory disturbance, urodynamic and urorectal dysfunctions, and secondary orthopedic deformities. The natural course of the disease is usually poor: lower spinal cord function gradually deteriorates without appropriate treatment of tethered cord\(^\text{9}\). Release of a tethered cord with anatomical reconstruction of the various tissue layers during infancy or early childhood, on the other hand, has been shown to prevent this symptomatic progression\(^\text{6,16}\). However, even after an initially successful untethering procedure, many patients exhibit radiological evidence of adhesion of the residual lipoma to the dural sac\(^\text{16,29}\). Also, it is well known that not all patients experience functional improvement following surgery for LMMC with TCS. These unfavorable results may be due to damage to the spinal cord during the operation or to re tethering of the spinal cord by the surrounding scar tissue. Various methods have been developed by many pediatric neurosurgeons to prevent postoperative retethering. Authors have used a mega dural sac repair procedure after untethering and have obtained favorable results with this. In the present study, authors tried to find unique features of LMMC in terms of clinical presentation, precipitating factors for onset of symptoms, various pathologic entities, and surgical outcome from a long-term follow-up review of 75 LMMC patients. Also, various factors involved in postoperative retethering are evaluated.

**MATERIALS AND METHODS**

From 1980 to 2001, 115 patients with TCS were treated at the Kangnam St. Mary's Hospital. Study included 75 patients who were operated on for a condition diagnosed as LMMC, and these were enrolled in this study. Clinical characteristics, radiological features, pathological classifications and surgical outcome were evaluated by retrospective analysis. In cases of lumbosacral LMMC we classified the patients into three types according to the radiological features and operative findings: type I, “caudal type”; type II, “dorsal type”; and type III, “transitional or mixed type” (Fig. 1).

All of these patients were treated initially in our department, and the operative procedures were mostly consistent in these series as follows. An effort was made to delineate the lipoma extradurally and trace it back until the point of insertion into the spinal cord was exposed. The lipoma was debulked microsurgically to permit the conus to move more freely within the spinal canal, and any tethering bands

**Fig. 1.** Classification of lipomeningomyelocele. Type I caudal type, with lipoma terminating the caudal portion of the cord through the dural defect; type II dorsal type, with lipoma attached to the placode and neural elements suspended from the undersurface of the placode; type III transitional or mixed type, where lipoma is mixed with neural elements and placode through the dural defect.

**Fig. 2.** Schematic drawings of the operation. A: After debulking of lipomas, reconstruction of dura with Gore-Tex (G) is performed. B: A mega-dural sac is constructed to maintain the CSF circulation (arrows) and prevents retethering of the cord.
were identified and divided. The pia was approximated by suturing with 8-0 prolene over the residual lipoma on the site of placode. Finally, a mega-dural sac was constructed with lyophilized dura or Gore-Tex, with the purpose of maintaining the CSF circulation and preventing re-tethering of the cord (this procedure was performed in 15 of the patients referred most recently) (Fig. 2). As a result, this procedure allowed the spinal canal to expand in the posterior direction, creating a circumferentially wide space capable of accommodating the neural element within the dural sac (Fig. 3).

Authors also divided the patients into two groups according to age: group A, less than 3 years old; group B, 3-24 years. For the assessment of surgical outcomes, patients were graded as clinically improved, stable, mildly changed (only mild voiding difficulty postoperatively), and worsened. Pre- and post-operative urodynamic studies were conducted in 17 patients to evaluate the functional outcome (Fig. 4).

RESULTS

This patient population was made up of 40 male and 35 female patients. There were 51 patients under 3 years old (group A) and 24 over 3 years old (group B). The median follow-up period was 4 years (range 6 months to 18 years). Age distribution and types of the lesions are summarized in Table 1.

Initial clinical symptoms and signs were as follows: diffuse and nondermatomal leg pain referred to the anorectal region was the most common presenting symptom, and sensori-motor deficits in the lower extremities and bladder and bowel dysfunction were also common findings (Table 2). Patients with type III LMMC had more neurological deficits than the patients with type I LMMC.

Preoperative urodynamic studies (UDS) were performed in 17 patients (8 in group A, 9 in group B), 11 of whom (3 in group A, 8 in group B) had urodynamic dysfunction and 6 (5 in group A, 1 in group B) had normal urodynamic function.

Among these 75 patients with LMMC, preoperative neurological deficits improved after surgery in 29 patients (39%), remained stable in 28 (37%), changed slightly in 13 (17%), and worsened in 5 (7%) (Table 3). Patients in group A obtained better outcomes than those in group B (Table 4). According to the type of lesions, patients with types I and II LMMC obtained better outcomes than those with type III LMMC. Improvement or stable disease was observed in 24 out of 25 patients with type I and 23 out of 32 patients with type II LMMC, but only 10 out of 18 patients with type III showed improved or stable disease, and the symptoms worsened in 4 patients.

Postoperative UDS were performed in 13 patients, and
Table 1. Age distribution and types of lesion in 75 lipomeningomyelocele (LMMC) patients

<table>
<thead>
<tr>
<th>Age distribution</th>
<th>Type of lesion</th>
<th>( I (N=25) )</th>
<th>( II (N=32) )</th>
<th>( III (N=18) )</th>
<th>Total (N=75)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth to 1 month</td>
<td>1</td>
<td>6</td>
<td>1</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>2–5 months</td>
<td>7</td>
<td>9</td>
<td>4</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>7–12 months</td>
<td>3</td>
<td>6</td>
<td>2</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>13 months to 3 years</td>
<td>6</td>
<td>4</td>
<td>2</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>4–6 years</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>7–10 years</td>
<td>5</td>
<td>1</td>
<td>1</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>11–16 years</td>
<td>2</td>
<td>2</td>
<td>4</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>17–25 years</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>7</td>
<td></td>
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</tbody>
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Table 2. Clinical symptoms and signs in 75 patients with LMMC

<table>
<thead>
<tr>
<th>Symptoms and signs</th>
<th>Type of lesion</th>
<th>( I )</th>
<th>( II )</th>
<th>( III )</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensory deficits</td>
<td>5</td>
<td>12</td>
<td>13</td>
<td>30</td>
<td></td>
</tr>
<tr>
<td>Motor weakness</td>
<td>9</td>
<td>11</td>
<td>14</td>
<td>34</td>
<td></td>
</tr>
<tr>
<td>Pain in back, legs</td>
<td>9</td>
<td>18</td>
<td>17</td>
<td>44</td>
<td></td>
</tr>
<tr>
<td>Incontinence</td>
<td>8</td>
<td>13</td>
<td>17</td>
<td>38</td>
<td></td>
</tr>
<tr>
<td>Increased reflex</td>
<td>5</td>
<td>7</td>
<td>8</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>Diminished reflex</td>
<td>7</td>
<td>14</td>
<td>16</td>
<td>39</td>
<td></td>
</tr>
<tr>
<td>Deformity of foot</td>
<td>7</td>
<td>7</td>
<td>18</td>
<td>14</td>
<td>26</td>
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</table>

Table 3. Surgical outcomes in 75 patients with LMMC by clinical grade

<table>
<thead>
<tr>
<th>Type</th>
<th>Improved</th>
<th>Stable</th>
<th>Mild change</th>
<th>Worse</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>( I (N=25) )</td>
<td>17</td>
<td>7</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>( II (N=32) )</td>
<td>11</td>
<td>12</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>( III (N=18) )</td>
<td>1</td>
<td>9</td>
<td>4</td>
<td>4</td>
<td>29 (39%)</td>
</tr>
<tr>
<td>Total</td>
<td>29 (39%)</td>
<td>26 (37%)</td>
<td>13 (17%)</td>
<td>5 (7%)</td>
<td></td>
</tr>
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</table>

Table 4. Comparison of surgical outcomes in groups A and B

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Group A (%)</th>
<th>Group B (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Improved</td>
<td>25 (49%)</td>
<td>4 (16%)</td>
</tr>
<tr>
<td>Stable</td>
<td>21 (41%)</td>
<td>7 (29%)</td>
</tr>
<tr>
<td>Mildly changed</td>
<td>4 (7.8%)</td>
<td>9 (37.5%)</td>
</tr>
<tr>
<td>Worse</td>
<td>1 (1.9%)</td>
<td>4 (16%)</td>
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</table>

the mean follow-up period was 2.5 years. Postoperatively, 3 patients showed an improvement in bladder functions (2 in group A, 1 in group B), 9 patients showed no change in bladder dysfunction (4 in group A, 5 in group B), and 1 patient in group B had aggravated bladder dysfunction after surgery. Complications related to operations were postoperative CSF leakage (5 cases, 6.7%) and newly developed neurological deficits (6 cases, 8%). Four patients (5.3%) required reoperation because of subsequent symptomatic retethering. Neither retethering nor new neurological deficits occurred in the 15 patients who underwent mega-dural sac grafting procedures. The surgical outcome was satisfactory in terms of improvements in pain and motor weakness, but disappointing with reference to resolution of bowel and bladder dysfunction.

DISCUSSION

Hoffman et al.\(^6\) have stressed the importance of early treatment of LMMC patients, because the likelihood of avoiding or reversing a neurological deficit by surgical treatment is greatest early on and decreases with age, presumably indicating that with delayed treatment, chronic traction on the spinal cord ultimately leads to irreversible deficits. McLone et al.\(^7\), Lamacra et al.\(^8\), and Herman et al.\(^9\) have also recommended early surgical intervention for LMMC. In agreement with these authors, we advocated repair of these lesions within 1-6 months of birth or as soon as they are detected in patients referred after this age.

Chapman\(^5\) classified intraspinal lipomas into three types: caudal, dorsal and transitional, which were related to the attachment pattern of the lipoma on the caudal portion of the cord. Many authors use LMMC as a general term indicating all varieties of intraspinal or lumbosacral lipomas\(^10-11,12,13,14\). Arai et al.\(^11\) defined a classification scheme with five types of lumbosacral lipomas: dorsal type, caudal type, combined type, filar type, and lipomeningomyelocele. These authors defined LMMC as one of the subtypes of lumbosacral lipoma, and classify it as follows: type I (caudal), lipoma terminating the caudal portion of the cord through the dural defect; type II (dorsal), lipoma attached to the placode and neural elements suspended below the placode; and type III (transitional or mixed), lipoma mixed with neural elements and placode through the dural defect.

The aims of surgery are to remove the fibroadipose mass, to relive the tethering effect on the spinal cord, to preserve neural tissues, and to prevent retethering of the spinal cord. Basic surgical procedures are the same for each type of lipoma, but the degree of difficulty of the surgery is different for each type. Arai et al.\(^11\) reviewed their large series of patients with lumbosacral lipoma and reported that caudal and filar lipomas had simple attachments and tethered the caudal end of the cord, so that untethering of the spinal cord could easily be achieved. However, complete untethering was often not achieved because of injury to the functional neural elements, especially when an LMMC of the combined type was concerned, in which case surgery is accompanied by some risks. According to Pierre-Kahn et al.\(^20\), lipomas of the conus are compatible with dorsal and combined types of lipomas: lipoma of the filum, in which surgery is harmless and beneficial, and lipoma of the conus, for which surgery involves considerable risk. We have found that surgery is relatively simple and its results rather beneficial in types I (dorsal) and II (caudal) LMMC, while it involves considerable risk in type III (transitional or mixed) LMMC, especially
when its aim is extensive separation of the lipoma from the attached spinal cord.

Without therapy or with inappropriate therapy, the neurological function of patients deteriorates with increasing age and the patients finally become incapacitated. Fixation and/or tethering of the spinal cord and roots are undoubtedly the major causes of worsening. These factors probably explain the late or slowly progressive deterioration noted in these patients. Koyanagi et al. reviewed a lumbosacral lipoma series, finding that in at least 38% of the patients with urinary symptoms the onset of these had been quite late, after the establishment of bladder control, and that 89% of the patients with motor deficits had first been seen to have motor problems a relatively long time after birth. Previous clinical studies also demonstrated progressive neurological deficits with age in this disorder. Bulsara et al. reviewed their series of lipomeningomyelocele, intraspinal lipoma and filum terminale lipoma patients; they found that all groups showed the most significant improvement in motor function after surgery. Greater improvements in pain, bladder function, and sensation were observed following surgery for lipomas of the filum terminale. The least improvement in these groups was seen in the LMMC group.

It is important to emphasize that the goal of the operations in this study was to untether the cord and resect as much of the lipoma as possible without injury to the cord and nerve roots. In addition, it is important to restore the dural sac to prevent postoperative retethering and to maintain a normal CSF circulation space. Bulsara et al. report that early operative intervention leads to a good outcome and that younger patients tend to improve more than older patients. In our series, group A patients showed less pronounced neurological deterioration after surgery than did group B patients.

The exact mechanisms of progressive neurological deterioration in patients with lumbosacral lipoma have not been fully clarified. Tethering of the spinal cord and compression by the lipoma are thought to be responsible for neurological deterioration. It is well documented that the spinal cord and canal are similar in length in early fetal life but the conus medullaris ascends rapidly in the later stages of intrauterine development. Further ascent to a level adjacent to L-1 and L-2 is noted by the 2nd postnatal month in the normal child. An arrest of the normal ascent could produce tension with local ischemia of the lower spinal cord and nerve roots. Lipoma prevents the normal ascent of the spinal cord by tethering and causes neuronal dysfunction. Acute deterioration may occur following extreme flexion of the spinal column, as reported by Pang and Wilberger.

Our results indicate that the type of lesion of the lipoma is related to the preoperative neurological status. Chapman et al. reported that 36% (5/14) of patients were improved, 43% (6/14) unchanged, and 21% (3/14) worse after surgery for intraspinal lipoma patients. In the series of Pang et al., 29% (6/21) were improved, 67% (14/21) were unchanged and 4% (1/21) were worse. However, in Harrison et al.'s series of lumbosacral lipoma patients the authors reported a good outcome: 65% (13/20) improved and 35% (7/20) were unchanged, while Koyanagi et al. obtained improvement in 34.6% (9/26) of symptomatic patients. In our own series of 75 patients with LMMC, 39% (29/75) were improved, in 37% (28/75) disease remained stable, there was mild change in 17% (13/75), and 7% (5/75) were worse after surgery.

The prevalence of urological dysfunction in TCS is extremely high. Vernet et al. investigated the urodynamic studies in the management of children with tethered spinal cord. They evaluated the results of pre- and postoperative urodynamic studies in their 25 patients. Ten of 15 patients who had preoperative urological deterioration had experienced improvement or stabilization. With respect to urodynamic studies, there were significant increases in total and pressure-specific bladder capacities following untethering. In our series, among 13 patients in whom postoperative urodynamic studies were performed, 2 patients in group A (2/6) and 1 patient in group B (1/7) obtained improvement of bladder dysfunction. In 1 patient with worsened bladder function this is thought to have resulted from mechanical and vascular insults to the cord during the operation.

Retethering usually results from postoperative dural adhesion, which is undoubtedly common after LMMC repair. Authors devoted some thought to wondering what we could do during the primary repair to prevent recurrence of tethering. Closure of the meningeal components, pia mater, pia arachnoid and dura mater is essential for prevention of retethering. Attempted mega-dural sac grafting on the dural defect area to maintain the CSF flow suspended the neural elements. Posterior expansion of the spinal canal can create a wide space to accommodate the neural elements within the subarachnoid space. In our series, 4 patients (5.3%) who underwent conventional dural repair experienced retethering of the cord. However, there was no retethering in 15 patients in whom mega-dural sac repair was performed.

**CONCLUSION**

Our data continue to support the opinion that early diagnosis and adequate release of the tethered cord are of key importance to successful management of patients with
The improvements in pain and motor weakness after surgery were disappointing, but the resolution of bladder dysfunction was disappointing in older patients. With reference to the type of lesion, a better outcome was noted in patients with types I and II LMMC than in those with type III LMMC. For the prevention of retethering of the cord and promotion of functional recovery from neurological deterioration, authors would like to emphasize the importance of a well-planned mega-dural graft procedure in the dural defect area in every primary surgical intervention.

References