Embryopathogenetic surgicoanatomical classification of dysraphism and surgical outcome of spinal lipoma: a nationwide multicenter cooperative study in Japan

Clinical article

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Object. The natural history of asymptomatic spinal lipoma in infancy remains unclear, and the indication for the prophylactic untethering operation is still debatable. To address this question, a multicenter cooperative study for the treatment of spinal lipoma was performed by the 7 most active institutions in neurosurgical care for spina bifida in Japan between 2001 and 2005.

Methods. Patients were classified using the embryopathogenetic surgicoanatomical classification. Their neurosurgical postoperative course was analyzed using the Spina Bifida Neurological Scale. Among 261 patients, 159 were asymptomatic and 102 were symptomatic.

Results. Of the 136 patients for whom prophylactic surgeries were performed, 135 remained asymptomatic and only 1 (0.4%) of the 261 patients presented with mild sensory disturbance. Mild foot deformity was identified in 1 (4.3%) of 23 conservatively observed patients. Of 100 symptomatic patients, deterioration after surgery was seen in 6%, and improvement in 44%. Complete resolution of symptoms was seen in only 14.2%. Filar types for patients > 3 years old improved in Spina Bifida Neurological Scale scores from 12.3 to 14.0. The mean age of symptomatic patients with lipomyelomeningocele was the youngest of all (1.3 years), which indicates lipomyelomeningocele may deteriorate in early infancy. Improvements from surgery were seen for all types of lipoma except the caudal type, presenting at an older mean age (15 years).

Conclusions. A low rate of postsurgical worsening indicates that surgeries for asymptomatic and symptomatic lipomas are safe. Surgeries done after the onset of symptoms seldom cure the patients. These two results support early untethering for any kind of lipoma; however, further study of the natural history is required.

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KEY WORDS • spinal lipoma • prophylactic surgery • multicenter cooperative study • embryopathogenetic surgicoanatomical classification • Spina Bifida Neurological Scale

T he indications for prophylactic untethering surgery and the natural history of infantile asymptomatic spinal lipoma remain controversial. Discussion at the international level on the efficacy of early surgical intervention with untethering of the low-set conus medularis as a prophylactic operation has been inconclusive.3 Some aspects of the natural history of this condition have been clarified,1,2 but the information revealed has been insufficient to satisfy the supporters of either radical early prophylactic operation or conservative observation. In addition to an unclear natural history, no analyses have been performed based on consistent classifications of the vari-

Abbreviations used in this paper: COE-SB = Centers of Excellence in Spina Bifida; EPSAC = embryopathogenetic surgicoanatomical classification; EPSAC-dyr = EPSAC of dysraphism; LMMC = lipomyelomeningocele; LMMCC = lipomyelomeningocystocele; SBNS = Spina Bifida Neurological Scale.
Surgical outcome for spinal lipoma: multicenter cooperative study

The SBNS Score

The SBNS score provides a rating of 3 (worst) to 15 (normal) by grading motor (M: 6 points), reflex or sensory (R or S: 4 points), and bladder and bowel (B/B: 5 points) functions (Table 1). Grading is essentially based on the functional segmentation of the spinal cord. The maximum score is 11, rather than 15, in patients < 3 years old, because the bladder and bowel grade is confined to 1 regardless of the level of function. Whereas adults with 15 points are defined as asymptomatic, the definition of asymptomatic in children < 3 years old is 11 points and no obvious urological deficit. The SBNS was designed so that doctors or parents can score it without radiological or urological examination. In the present study, to determine that an infant was asymptomatic, urinary retention was ruled out by repeated ultrasonography examinations. A urodynamic study was not performed for all patients.

The SBNS grade classifications are defined as follows: Grade I, normal ambulation and bladder and bowel control; Grade II, normal ambulation and disturbed bladder and bowel control; Grade III, ambulatory without a brace and/or aid; Grade IV, ambulatory with a brace and/or aid; and Grade V, not ambulatory.

Methods

The EPSAC-Dysr System

As shown in Fig. 1 and Table 2, the fundamental concept of the EPSAC-dysr is based on the embryological stage of development of the individual type of dysraphism and the surgical reversibility of the neurological deficit after the anatomical reconstructive procedure. The basic background for this classification is based on identical morphogenetic findings of myeloschisis in caudal dysraphism (SB) and encephaloschisis in cephalic dysraphism (cranium bifidum), as demonstrated in previous experimental and clinicopathological studies. The clinical form of myeloschisis and encephaloschisis (or anencephaly/exencephaly) is a critical dysraphic state with arrest of the neuronal maturation process at approximately the 4th week of gestation. Unlike this dysraphic entity, meningocele and myelomeningocele/encephalomeningocele are surgically reversible structures, both in anatomical deformity and neurofunctional deficit, as consequent secondary changes following normally completed neuronal maturation.

The primary definitive cause of secondary deformity of the brain or spinal cord is a structural defect (hole) in the dura mater (meningoschisis) together with a herniated arachnoid sac (meningocele) containing CSF and the brain or spinal cord. Pulsatile CSF force aggravates this secondary structural deformity. The resulting neurological deficits are progressive but possibly reversible by reconstructive surgery. Defects in the lamina (laminoschisis) should always be associated with the aforementioned dysraphic states of myeloschisis/encephaloschisis and meningocele. Spinal lipoma associated with SB is always classified with meningocele (Figs. 2 and 3). A subcutaneous lipomatous fatty mass is a continuous structure penetrating the dura mater through the defect of meningocele. This may or may not be associated with meningocele, but the spinal cord will be severely deformed if the meningocele contains a spinal cord structure herniated through the meningocele defect, as in LMMC or LMMCC (Fig. 3 right).

In the analysis performed in this study, LMMC was included with the 3 historically important subtypes of lipoma (dorsal, caudal, and transitional) proposed as the Chapman classification. The filar type, as a significant subtype of spinal lipoma without meningocele, was also included in this widely used classification, as reported by Arai et al. One asymptomatic patient with a thickened filum terminale, who was 8 years old at registration and 9 years old at surgery, was also included in the filar-type lipoma group.

Clinical Cases: COE-SB Top 7, Japan

Between 2001 and 2005, patients with spinal lipoma treated in 7 neurosurgical institutions in Japan (The Jikei
University Hospital, Aichi Prefectural Colony–Welfare Center for Persons with Developmental Disabilities, Jun-tendo University Hospital, Miyagi Children’s Hospital, Kansai Medical University, Saitama Children’s Medical Center, and Chiba Children’s Hospital) were analyzed retrospectively. According to a nationwide survey on SB care in Japan in 2003 reported by the Spina Bifida Study Group of the Ministry of Health, Labor, and Welfare of Japan, more than half (87 [51.8%] of 168) of patients with spinal lipomas in Japan were treated in these 7 institutions.

Inclusion criteria were the presence of a spinal lipoma with attachment to the conus or filum, as demonstrated on MR imaging. Cases were classified for analysis according to the EPSAC-SB. Spinal lipomas with meningocele such as LMMC were included, in addition to spinal lipomas without meningocele. Surgeons in the 7 institutions recommend surgery to all patients fulfilling the inclusion criteria. After appropriate information on the uncertain natural history, the controversial surgical indication, and surgical risks was provided, surgeries were performed when the parents or patient agreed. Care was taken not to provide biased information based on predictions of deterioration or.

TABLE 2: The “classical” and the “clinical” classification of spina bifida in relation to embryopathogenic and surgicoanatomical classification of spina bifida (EPSAC-SB)

<table>
<thead>
<tr>
<th>Embryopathogenic Concept of Neural Tube Defects</th>
<th>Spina Bifida Aperta</th>
<th>Spina Bifida Occulta</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. myeloschisis [+ II + III]</td>
<td>1) myeloschisis</td>
<td>1) myelodysplasia</td>
</tr>
<tr>
<td>II. meningoschisis [+ III]</td>
<td>1) myeloschisis (cystic form)</td>
<td>1) lipo-meningocele</td>
</tr>
<tr>
<td></td>
<td>1) meningocele</td>
<td>2) lipo-myelo-meningocele</td>
</tr>
<tr>
<td></td>
<td>2) myelo-meningocele</td>
<td>3) lipo-myelo-meningo-cystocele</td>
</tr>
<tr>
<td></td>
<td>4) others</td>
<td>4) others</td>
</tr>
<tr>
<td>III. laminoschisis</td>
<td>5) spinal lipoma without meningocele</td>
<td>6) congenital meningocele</td>
</tr>
<tr>
<td></td>
<td>1) congenital meningocele</td>
<td>1) congenital meningocele</td>
</tr>
<tr>
<td></td>
<td>2) others (laminodysplasia and others)</td>
<td>2) others (laminodysplasia and others)</td>
</tr>
</tbody>
</table>

* Inside the shaded area: spina bifida cystica. (Table modified for typographical and style considerations. Reprinted with permission from Oi et al. Nervous System in Children 27:213–222, 2002.)
Surgical outcome for spinal lipoma: multicenter cooperative study

Surgical difficulty assumed from the neuroimaging findings. The distributions of asymptomatic and symptomatic patients who underwent surgery or observation are shown in Table 3.

Surgical procedures, including untethering and partial removal without insult to the spinal cord or roots, were commonly performed by expert pediatric neurosurgeons, and more detailed surgical options were determined at each institution. Preoperative and postoperative neurological findings were recorded according to the SBNS (Table 1).\(^\text{16,19}\) The SBNS was also scored at the end of 2005 as a final follow-up score, regardless of whether the patient chose surgery or conservative management. The median follow-up period was 125 weeks (range 1 month–5 years), and immediate postoperative outcomes were analyzed within 4 weeks of the operative procedure. In addition to the overall results from the 7 institutions, detailed data according to patient age, subtype classification of the lipoma as proposed by Chapman,\(^\text{4}\) and degree of symptoms are available from the Jikei University Hospital Women’s and Children’s Medical Center, Tokyo.

**Results**

**Summary of the Multicenter Cooperative Study**

As shown in Table 3, a total of 261 patients were enrolled from the 7 institutions. The mean number of patients at a single institution was 37.3 (range 20–53 patients). Among the 159 asymptomatic patients, 136 underwent surgery and 23 received conservative observation without surgery. Of the 102 symptomatic patients, 100 underwent surgery. Despite being symptomatic, 2 patients were observed without surgery because their symptoms improved to an acceptable degree (SBNS scores improved from 11 to 14 in one, and from 12 to 15 in the other) immediately after admission.

Surgical results for asymptomatic patients are shown in Table 3. Of the 136 asymptomatic patients who underwent surgery, urinary retention was seen in 1 patient (0.7%) postoperatively. However, the deficit resolved after a few weeks and the patient was not suffering from any symptoms as of the final follow-up. Only 1 patient (0.7%) displayed deterioration in the follow-up period. A second surgery was not performed because symptoms were limited to mild sensory disturbance. Of the 100 symptomatic patients who underwent surgery, 20 (20%) improved immediately after surgery, and the improvement rate increased to 44% at the final follow-up. Worsening of symptoms was also seen in 9 (9%) of 100 patients immediately postoperatively, but decreased to 6% at the final follow-up. Nonsurgical observation was chosen for 25 patients. Of 23 asymptomatic patients, only 1 patient (4.3%) presented with varus in the follow-up period. The patient remained under observation because the symptoms were mild and did not affect daily activities. The other 22 asymptomatic and 2 symptomatic patients (95.7 and 100%) had stable neurological states.

**Results for Subtypes of Spinal Lipoma**

The largest number of patients was recorded at the...
Jikei University Hospital Women’s and Children’s Medical Center, and more detailed results are provided for these patients. Distributions of lipoma types according to EPSAC and subtypes according to the Chapman classification are shown in Table 4. Lipomeningocele was not seen during the research period, and LMMCC was excluded from the analysis because the number of cases was too small. The mean age at first examination was high for caudal-type lipomas (15 years; range 0–64 years), and low for transitional-type lesions (1.3 years; range 0–7 years) and LMMC (1.5 years; range 0–6 years). The ratio of symptomatic to asymptomatic patients varied depending on the lipoma type (Table 4). The incidence of symptomatic disease was high for the caudal type and LMMC.

Surgical results for asymptomatic and symptomatic patients were similar to those seen in the overall data, with no deterioration seen in asymptomatic patients (Table 5), and improvement and deterioration rates for symptomatic patients of 39 and 4%, respectively (Table 6). The single patient who deteriorated after surgery was a 1-month-old child with LMMC whose preoperative SBNS of 4 represented one of the most severely affected patients. This patient's neurological state worsened and then stabilized by the time of the surgery. Normalization of neurological symptoms after surgery was seen in 4 (14.2%) of 28 symptomatic patients. The highest incidence of improvement was seen for the filar type (50%) and, unexpectedly, for the transitional type (66.7%). These improvements were also revealed in the SBNS scores (Tables 7 and 8). Changes in the mean SBNS scores were −0.1 (filar type) to 1.3 (LMMC) in patients < 3 years old (maximum SBNS score 11); and 0 (transitional type, full scale [perfect score: 15 of 15 points] preoperative) to 1.7 (filar type and LMMC) in patients ≥ 3 years of age (maximum SBNS score 15). The initial and final SBNS scores were characteristic. The difference in scores between the best and worst type of lipomas at first examination was 2.1 for patients < 3 years of age, compared with 7.3 at the final follow-up examination in patients ≥ 3 years of age. The scores for filar-type lipomas were significantly better than those for LMMC at the final follow-up (p < 0.01). Permanent aggravation of the motor deficit was seen postoperatively in 1 patient with LMMC.

Postoperative Complications

Complications other than neurological deterioration were seen in 19 of the 236 surgical patients. No deaths were recorded during the study period. Wound infection and subcutaneous CSF collection that prolonged the admission period were seen in 6 patients (2.5%) and 9 patients (3.8%) in the surgical group, respectively, but meningitis did not develop. Complications unrelated to surgical procedures such as upper respiratory tract infection were also seen in 4 patients (1.6%). These events did not influence the neurological outcomes.

Discussion

Symptomatology in Spinal Lipoma

Although controversy remains, many neurosurgeons

### TABLE 4: Mean age in years at first examination for patients with individual tumor subtypes

<table>
<thead>
<tr>
<th>Lipoma Type</th>
<th>Asymptomatic (no. of cases)</th>
<th>Symptomatic (no. of cases)</th>
<th>Total (no. of cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td>dorsal</td>
<td>1.9 ± 2.9 (6)</td>
<td>8.5 ± 7.1 (3)</td>
<td>4.1 ± 5.4 (9)</td>
</tr>
<tr>
<td>caudal</td>
<td>7.6 ± 0.1 (1)</td>
<td>16.0 ± 23.0 (7)</td>
<td>15.0 ± 21.5 (8)</td>
</tr>
<tr>
<td>transitional</td>
<td>1.2 ± 2.8 (6)</td>
<td>1.7 ± 1.6 (7)</td>
<td>1.3 ± 2.2 (13)</td>
</tr>
<tr>
<td>filar</td>
<td>1.5 ± 3.3 (7)</td>
<td>2.4 ± 2.6 (8)</td>
<td>2.0 ± 2.9 (15)</td>
</tr>
<tr>
<td>LMMC</td>
<td>0.4 ± 0.4 (3)</td>
<td>1.8 ± 2.2 (5)</td>
<td>1.3 ± 2.2 (8)</td>
</tr>
<tr>
<td>total</td>
<td>1.7 ± 2.3 (23)</td>
<td>5.9 ± 12.4 (30)</td>
<td>4.1 ± 9.8 (53)</td>
</tr>
</tbody>
</table>

* Classified according to the EPSAC system (see refs. 15 and 16) and Chapman and colleagues’ classification (see refs. 4 and 5).
Surgical outcome for spinal lipoma: multicenter cooperative study

TABLE 6: Number of improved, unchanged, and worsened patients after surgery for symptomatic lipoma

<table>
<thead>
<tr>
<th>Lipoma Type</th>
<th>Improved (%)</th>
<th>Unchanged (%)</th>
<th>Worsened (%)</th>
<th>Total Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>dorsal</td>
<td>1 (33)</td>
<td>2 (67)</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>caudal</td>
<td>1 (17)</td>
<td>5 (83)</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>transitional</td>
<td>4 (67)</td>
<td>2 (33)</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>filar</td>
<td>4 (50)</td>
<td>4 (50)</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>LMMC</td>
<td>1 (20)</td>
<td>3 (60)</td>
<td>1 (20)</td>
<td>5</td>
</tr>
<tr>
<td>total</td>
<td>11 (39)*</td>
<td>16 (57)</td>
<td>1 (4)</td>
<td>28</td>
</tr>
</tbody>
</table>

* Four of the 11 improved patients displayed resolution of symptoms, among 28 symptomatic patients (4 of 28: 14.2%).

believe in the effectiveness of prophylactic surgery for asymptomatic lipoma. To justify prophylactic surgery as the first-choice treatment, objective evidence is required showing that: 1) immediate- or late-onset neurological deterioration and surgical complications are acceptably low; 2) neurological symptoms that appear while patients are conservatively observed are seldom cured even when radical surgery is applied after the onset; and 3) the ratio of symptomatic to asymptomatic patients without surgery increases with age more than does the ratio for patients who receive surgery.

In our study, symptoms were partially improved at as high an incidence as 44%. However, complete remission was relatively uncommon at 14.2%. Radical surgery is widely considered useless for longstanding stable neurological deficits. Although the majority of neonates diagnosed with spinal lipoma present with stigmata and other skin manifestations, children in whom lipoma is diagnosed at an older age present with progressive neurological symp-

toms. Patients with caudal-type lipomas in this study had the highest mean age and the lowest improvement rate of symptoms of the 5 types. Correcting neurological or urological symptoms to a normal status is thus expected to be difficult after untethering surgery. The present study confirmed that the rate of surgical complications is acceptably low and that neurological improvements for symptomatic patients were not adequate after surgery. However, the ratio of symptomatic to asymptomatic patients without surgery was not significantly higher than for surgical patients in the follow-up period. These results indicate that prophylactic surgery is possibly recommended, although further investigation of natural history is needed. The SBNS scoring system, designed and clinically analyzed by Oi and Matsumoto in 1992, is valuable for the evaluation of chronological changes in neurological function. Unlike other available grading scales for spina bifida, the SBNS rating includes the level of spinal function.

Because bladder and bowel functions in infants are not reflected in the SBNS, scores for patients < 3 years old were analyzed separately. Completely assessing urological deficit in infants is difficult, which is another reason to recommend prophylactic surgery. Patients whose occult symptoms may be present in the infantile period and progress later, particularly in late childhood, may miss the chance to have deterioration prevented.

Classification of Spina Bifida/Spinal Lipoma and Surgical Outcomes

Kulkarni et al., supported conservative management for any type of asymptomatic lipoma. In the present study, results for lipomas with and without meningocele based on the EPSAC-SB classification were analyzed. Among these, no cases of lipomeningocele were treated during the study period, but the senior author (S.O.) has encountered a case of infantile spinal lipoma in which the form changed from a spinal lipoma without meningocele to a type with progressive meningocele during the developmental period in late infancy. A significantly better functional prognosis has been reported for filar type lipoma than for lipomas of the conus. Surgical indications for filar lipoma were objectively acceptable, with a high improvement rate and no surgical morbidity for the prophylactic operation. The natural history of LMMC is worse than any other type of lipoma. This patient with LMMC who postoperatively deteriorated had the most severe form in neurology. It is difficult to help a very young infant with a severely damaged neural element. Only asymptomatic or mildly symptomatic patients treated appropriately at early ages (between 1 and 8 months) remained neurologically unchanged or improved (Tables 6 and 7). However, when surgical treatment is delayed or not appropriately performed, these patients may be left with significant neurological sequelae, or their symptoms may even be aggravated, with relatively rapid

TABLE 7: The mean SBNS scores in patients < 3 years of age before and after surgery and at follow-up*

<table>
<thead>
<tr>
<th>Lipoma Type</th>
<th>Preop</th>
<th>Postop</th>
<th>Final FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>dorsal</td>
<td>10.8 (4/5)</td>
<td>10.8 (4/5)</td>
<td>11.0 (4/4)</td>
</tr>
<tr>
<td>caudal</td>
<td>10.0 (0/2)</td>
<td>10.5 (0/2)</td>
<td>10.0 (0/1)</td>
</tr>
<tr>
<td>transitional</td>
<td>9.8 (4/10)</td>
<td>10.4 (6/10)</td>
<td>10.6 (4/7)</td>
</tr>
<tr>
<td>filar</td>
<td>10.4 (4/10)</td>
<td>10.5 (4/10)</td>
<td>10.3 (1/6)</td>
</tr>
<tr>
<td>LMMC</td>
<td>8.7 (3/7)</td>
<td>9.3 (3/7)</td>
<td>10.0 (3/5)</td>
</tr>
<tr>
<td>total</td>
<td>9.9 (15/34)</td>
<td>10.3 (17/34)</td>
<td>10.4 (12/23)</td>
</tr>
</tbody>
</table>

* The maximum SBNS score in patients < 3 years is 11. Numbers in parentheses denote the number of asymptomatic/total patients.

TABLE 8: The mean SBNS scores in patients ≥ 3 years of age before and after surgery and at follow-up*

<table>
<thead>
<tr>
<th>Lipoma Type</th>
<th>Preop</th>
<th>Postop</th>
<th>Final FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>dorsal</td>
<td>12.0 (0/2)</td>
<td>12.0 (0/2)</td>
<td>13.3 (1/3)</td>
</tr>
<tr>
<td>caudal</td>
<td>8.8 (1/5)</td>
<td>8.8 (1/5)</td>
<td>10.0 (1/6)</td>
</tr>
<tr>
<td>transitional</td>
<td>15.0 (1/1)</td>
<td>15.0 (1/1)</td>
<td>15.0 (4/4)</td>
</tr>
<tr>
<td>filar</td>
<td>12.3 (1/4)</td>
<td>13.3 (2/4)</td>
<td>14.0 (3/8)†</td>
</tr>
<tr>
<td>LMMC</td>
<td>6.0 (0/1)</td>
<td>6.0 (0/1)</td>
<td>7.7 (1/3)†</td>
</tr>
<tr>
<td>total</td>
<td>10.6 (3/13)</td>
<td>10.9 (4/13)</td>
<td>12.3 (10/24)</td>
</tr>
</tbody>
</table>

* Numbers in parentheses denote the number of asymptomatic/total patients.
† p < 0.01.
progressive neurological deterioration. The untethering surgery must be performed as early as anesthesia and surgical procedures can be done safely, because the mean age for symptomatic LMMC was nearly the youngest, second only to transitional-type lipoma (Table 4).

There are similarities between LMMC and transitional-type lipoma in age at onset and surgical difficulty in detaching the conus from the lipoma without injuring the nerve roots. On the other hand, the SBNS grade of transitional-type lipoma is I or II, which is milder than that of LMMC (III or higher) (Table 8). Prophylactic surgery for transitional-type lipoma has not been recommended by some authors.2 These authors have supported a strategy in which transitional lipomas should be surgically treated only when the patient is symptomatic. Milder symptoms compared with LMMC may suggest that surgery for transitional-type lipomas can be delayed until symptoms occur.

Release of tethered neural elements should normally be conducted, and volume reduction is also needed, for dorsal- and transitional-type lipomas. These lipomas may display mass effects and displace the conus and nerve roots, producing spinal canal stenosis. Motor deficits are noted in patients with lipomas extending cranially beyond the L-5 level. In addition, the fat mass index peaks in both boys and girls at 6 months of age.28 Early decompression surgery is recommended for patients with large dorsal-type lipomas.9

Although the mechanisms causing symptoms are related not only to stretching of the spinal cord, but also to factors such as natural history that are not controllable by surgery,23,26 untethering and decompression are to date the only available prophylactic interventions. A previous theory that dissociated growth rates between the vertebral column and the fixed spinal cord after birth causes tethered cord syndrome23 has not recently been supported. The conus has already reached the adult level by the time of birth.2,29 Dynamic factors caused by spinal movement, not static factors, are crucial in exerting mechanical force on the spinal cord. Intermittent stretching force on the caudal spinal cord causes hypoxemia and ischemia, and then inhibits oxidative metabolism and electrical activity in the spinal cord.31 The length of the vertebra is increased by flexion of the spinal column.24 Longitudinal stretching force on the spinal cord as a result of body motion ultimately causes neurological deficits. Because this traction of the spinal cord also occurs even in thick filum terminale without lipomatous masses,27 complete untethering is required. Several methods have been attempted to prevent retethering. Grafting of a dural substitute to posteriorly enlarge the caudal dural sac is one appropriate procedure. A dural substitute is recommended as long as CSF leakage, the most common postoperative problem, is prevented.

Further Prospects for Study Design of COE-SB Top 7
Japan

To advance the present ongoing discussions on this unresolved and controversial issue, the natural history of and indications for asymptomatic spinal lipomas will be objectively analyzed as a nationwide multicenter cooperative prospective study by the COE-SB Top 7, Japan study group. As materials and methods for any prospective study, the data obtained regarding immediate postoperative outcomes in this retrospective analysis will offer highly useful reference material to provide a common standard of informed consent, particularly for patients or their parents to decide on treatment, either a prophylactic operation or conservative observation. Rates of surgical morbidity or complications aggravating the neurological state were extremely low in patients with nonsymptomatic lipoma in the present retrospective study (0.7%) compared with other reported retrospective analyses from any single institution. The complication rate was as low as 6% in cases of symptomatic lipoma at this group of institutions. It may be concluded that patients or their families may not get a feeling of “risky surgery” when making decisions if these institutions are chosen for initial treatment. However, patients with nonsurgical follow-up also showed low risk of symptomatic change (0.7%) during the same relatively short period of follow-up (< 5 years). Definitive support for either treatment modality is thus impossible, even though surgical outcomes for symptomatic patients were somewhat hopeful, with a 44% improvement rate, although the “normalization” of neurological deficits was disappointingly low at 14.2%. One positive aspect may be that these surgical outcomes and the efficacy of the untethering procedure depended on the type of spinal lipoma. Definitive consensus on classification and common terminology should be established to resolve these controversial issues.

Future study designs for the COE-SB Top 7, Japan will focus on the following subjects: 1) a prospective cohort study with highly skilled pediatric neurosurgery techniques, as a well-designed controlled study without randomization (Level 2a of the Oxford Center for Evidence-Based Medicine’s Levels of Evidence); 2) a prospective cohort study over the long term (that is, in the course of 20 years), in which objective quantitative evaluation is used for neurological and chronological changes; and 3) final conclusions based on a scientifically supported classification, with various subtypes among dissimilar patient populations.

Conclusions

The safety of untethering surgery for asymptomatic and symptomatic lipomas was confirmed in the 7 most active institutions in neurosurgical care for spina bifida in Japan. Among the lipomas classified according to the EPSAC, LMMC presented severe symptoms in early infancy, and untethering surgery within the period of infancy is recommended. Further investigation of the natural history would indicate the surgical criteria for each type of asymptomatic lipoma. The SBNS is useful for chronological assessment and follow-up.

Disclosure

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References


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