Surgical treatment for lipomyelomeningocele in children

Sheng-Li Huang, Wei Shi, Li-Gen Zhang

Xi'an, China

Background: **Lipomyelomeningocele (LMM) is a common and severe closed neural tube defect in children. Because of the complex anatomy of LMM and the difficulty in assessing the value of surgery, the management of patients with LMM is controversial. This study was undertaken to evaluate effective techniques and procedures in surgical treatment of LMM and to assess the value of early neurosurgical intervention.**

Methods: **Twenty-five children with LMM aged from 2 months to 6 years underwent surgery between January 2004 and December 2006. Magnetic resonance imaging (MRI) of the spine and electromyography (EMG) of the lower limbs was conducted in all patients preoperatively. Urodynamic studies were appropriate even in asymptomatic patients. Hoffman's functional grading was used to assess the pre and postoperative status of the patients. The operation was composed of subtotal excision of lipoma, suturing of the spinal pia mater, and section of the filum terminale. Suturing of the spinal pia mater was performed in a wide process of reconstruction of meningeal layers including the dura. Follow-up lasted 1-4 years (mean 2.1 years), in which all children underwent neurological examination, EMG and MRI.**

Results: **After surgery a temporary neurological deterioration was found in two patients including slight weakness of a leg in one patient and urinary retention in the other, but it recovered completely a few days later. No postoperative complications were encountered. During the follow-up, 20 asymptomatic patients remained symptom-free. Symptoms disappeared totally in 2 of the 5 patients with neurological deficits, improved in one patient and stabilized in the remaining 2. MRI showed no tethered cord in all patients who underwent surgical procedures.**

Conclusions: **Early operation for LMM patients, even asymptomatic ones, should be performed to prevent the development of neurological deficits. Subtotal excision of lipoma, suturing of the spinal pia mater, and section of the filum terminale are recommended in the surgical treatment of LMM. The longitudinal cut of the filum terminale, a technique we have established in our surgical practice, is a simple and practical way to identify the filum terminale by visual inspection. And suturing the spinal pia mater is of extreme importance in preventing postoperative tethering.**

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Author Affiliations: Department of Neurosurgery, the Second Affiliated Hospital, School of Medicine, Xi'an Jiaotong University, Xi'an 710004, China (Huang SL, Shi W); Department of Surgery, Xi'an Children's Hospital, Xi'an 710003, China (Zhang LG)

Corresponding Author: Wei Shi, Department of Neurosurgery, the Second Affiliated Hospital, School of Medicine, Xi'an Jiaotong University, Xi'an 710004, China (Tel: 86-29-87679366; Fax: 86-29-87678634; Email: sweins@163.com)

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Introduction

Lipomyelomeningocele (LMM) is a closed neural tube defect commonly seen in children, which is invariably in the lumbosacral region. Though Johnson[1,2] first described this entity in 1857, the literature about these lesions has been confusing. The term "lipomyelomeningocele" is used in a broad sense to include all forms of congenital spinal lipomatous malformations.[2-6] Few authors have provided a specific definition of LMM. Such a liberal usage of this term leads to confusion in analyzing the result of its treatment. The term "lipomyelomeningocele" used in this study refers to the following conditions: a subcutaneous lipoma, which is typically present in the midline of the lumbosacral region, continues through a midline defect in the lumbodorsal fascia, vertebral neural arch and dura, attaches to an elongated and tethered spine cord and conus, but not to the roots of the cauda equina. The condition may prevent upward mobility of the conus medullaris of the spinal cord during normal childhood development and consequently may lead to progressive neurological and urinary deterioration.

Because of the complexity of the defect, great risk exists in surgery for LMM. Besides, as LMM usually appears to be asymptomatic in its early stage, some clinicians are inclined to adopt conservative treatment for this condition.[7-9] Without effective intervention,

however, the situation may worsen with neurological deterioration developing with age. In 1950, Bassett[10] emphasized the progressive deterioration of nerve functions in patients with LMM, and stressed the value of early diagnosis and prophylactic surgery. Since then, more attention has been paid to the early surgical treatment of this disease. But it is difficult to perform such an operation as well as to assess the value of early surgery for LMM in spite of the advances in medical sciences. Thus the management of LMM patients is still controversial and the value of early surgery remains uncertain.[11,12] The present paper introduces our experience in early surgical treatment of children with LMM, the surgical techniques, and the value of early neurosurgical intervention.

Methods

Patients

From January 2004 to December 2006, 25 children with LMM, 13 boys and 12 girls, aged from 2 months to 6 years, were admitted to our hospital. These patients, including asymptomatic ones, underwent surgery once the diagnosis of LMM was made. Two patients were under the age of 3 months when they received operation, 9 were 3-6 months, 7 were 7-12 months, 5 were 1-3 years, and 2 were more than 3 years (Fig.).

Neurological examination was performed preoperatively to assess sensation, strength, muscle tone, deep tendon reflexes, volitional control of muscles of the lower extremity and perineal area, and gait abnormalities when appropriate. A subcutaneous fatty mass was observed at the midline or just off the midline in the lumbosacral region in all patients. Some of them had other skin lesions associated with the fatty masses including skin dimples (7 patients), hemangioma (6), hair nevus (2), tail-like appendages (1), and scar-like skin patches (1). Normal functions of the lower extremities were observed in 20 patients, weakness in 3, and deformed foot in 2. One of the 3 patients with weakness had significant gait problems. None of the patients suffered from loss of sensation in the legs and/or sacral region. However, detailed sensation examination was not possible in very young children. Many of the older children complained of pain when they were lying supine or whenever there was pressure on the lipomatous swelling. Urodynamic studies were conducted in some patients when appropriate. Usually normal continence and voiding pattern suggest normal bladder and bowel control, while incontinence with pathologic urodynamics and impaired bladder voiding indicates a neuropathic bladder. Seven children were subjected to urodynamic studies. All the 25 patients demonstrated adequate bladder and bowel control. Hoffman's functional grading[2] was used to assess their status pre and postoperatively (Table 1). All the patients underwent electromyography (EMG) of the lower limbs and magnetic resonance imaging (MRI) of the spine preoperatively. The diagnosis of LMM was made preoperatively by MRI and was confirmed pathologically after operation. Two patients had associated anomalies: one had diastematomyelia and the other, anal stenosis. None of the patients had an associated Chiari malformation or hydrocephalus.

Surgical procedures

Under general endotracheal anesthesia, a patient was placed in a prone position with all pressure points padded. An oval skin incision was made around the base of the mass which was dissected circumferentially toward the fascia where the mass narrowed into a stalk and merged with the dura. The adjacent laminae were resected. The dura was opened under a microscope at the midline superiorly and then reflected laterally. The dura was opened from the normal area towards the lesioned

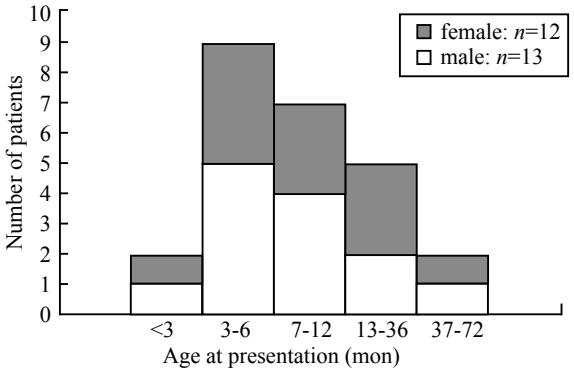

Fig. The number of patients with lipomyelomeningocele and their ages at surgery.

Table 1. Hoffman's functional grading[2]

Grade	Description
0	No significant neurological, orthopedic, or urological problem. The patient may have reflex changes and/or sensory deficits. Minor neurological deficits and sphincter problems may not be recognized in infants under 6 months of age, and therefore some patients who are in grade 0 may belong to a higher grade.
1	Minimal weakness and/or muscle wasting and/or foot deformity affecting only one leg without significant gait disturbance. Normal bladder and sphincter function.
2	Neurogenic bladder alone or combined with weakness of one leg; or intact bladder function with minimal weakness affecting both legs.
3	Moderate to severe weakness of one leg producing gait disturbance with or without neurogenic bladder or minimal weakness of both legs combined with neurogenic bladder.
4	Severe paraparesis requiring aids for walking with or without neurogenic bladder.
5	Inability to ambulate.

Table 2. Functional status of the 25 patients before surgery and one year after surgery

Age groups	*n*	Preoperative grade						Postoperative grade					
		0	1	2	3	4	5	0	1	2	3	4	5
<3 mon	2	2	0	0	0	0	0	2	0	0	0	0	0
3-6 mon	9	9	0	0	0	0	0	9	0	0	0	0	0
7-12 mon	7	5	2	0	0	0	0	7	0	0	0	0	0
1-3 y	5	4	0	1	0	0	0	4	1	0	0	0	0
>3 y	2	0	1	0	1	0	0	0	1	0	1	0	0
Total	25	20	3	1	1	0	0	22	2	0	1	0	0

site and then around the stalk, and intradural exploration was thus performed. The operation for LMM was mainly composed of subtotal excision of lipoma or fatty mass, suture of the spinal pia mater, and resection of the filum terminale. The lipoma was debulked with standard microsurgical techniques without use of an ultrasonic aspirator. Once the cord was completely free, the dura was closed. The lumbodorsal fascia might be released laterally and mobilized towards the midline in some of the patients. Finally, redundant skin was excised and the wound was closed. Intraoperative monitoring was not done in all patients. Fortunately no additional plastic surgeon collaboration was required.

Results

Postoperative course

After operation, the 25 patients recovered and were released from the hospital. There was a temporary neurological deterioration in 2 patients, one of whom had temporary worsening bladder control (urinary retention) which required catheterization, and recovered four days after catheterization; the other with slight weakness of one leg recovered one week later. No postoperative complications, cerebrospinal fluid leakage, wound infection or flap necrosis were encountered.

Follow-up

The postoperative follow-up of the 25 patients ranged from 1 to 4 years, on average 2.1 years. All these patients were subjected to neurological examination. EMG was performed when the patient presented with detectable neurological deterioration, and postoperative EMG was compared with preoperative EMG. Only detectable neurological worsening was considered as postoperative deterioration. Deterioration was observed in none of the patients during the follow-up and 20 asymptomatic patients maintained symptom-free. In five patients with neurological deficits, symptoms disappeared totally in 2, improved in 1 and stabilized in 2. The functional status of the 25 patients before surgery and one year after surgery is shown in Table 2. According to Hoffman's functional grading, 3 patients showed improvement of symptoms (one Hoffman's grade), two patients were stable and no patients deteriorated at the end of the first year follow-up. Weakness and pain were improved more markedly than other deficits. As expected, long-standing motor deficits and fixed orthopedic deformities were not improved. None of the patients had scoliosis. Postoperative MRI showed that there was no tethering of the spinal cord to the dura mater. No lipoma recurrence was observed at the end of the follow-up.

Discussion

LMM is a closed neural tube defect or a subcutaneous mass, which occurs approximately once in every 4000 births.[12] The mass occurs when a fatty mass protrudes posteriorly beyond the bony cavity of the intervertebral space with meningeal covering. A subcutaneous lipoma is typically present in the lumbosacral region, usually at the midline, but occasionally placed eccentrically. The lesion is not exposed to air and there is no drainage of cerebral spinal fluid. Children with LMM may develop progressive neurological deterioration characterized by varying degrees of lower extremity paralysis, decreased sensation and neurogenic bowel and bladder.[10,13,14] Surgery is not an effective treatment for installed neurological deficits but can prevent the progression of the deficits. The surgery is intended to reduce the bulk of the intradural part of lipoma, to decompress the terminal spinal cord, to untether the cord, to reconstruct the dura, and to prevent the progression of neurological deficit. Though children with LMM present no neurological symptoms at its early stage, operation is still a necessary intervention to restrict the progressive deterioration.[10,15] The operation is risky because of complexity of the condition, but the neurological symptoms of the condition may worsen if the operation is not properly performed. Thus it is critical to explore appropriate surgical techniques for the treatment of LMM.

LMM typically consists of normal mature adipocytes separated into clusters by numerous collagen bands. It is a hamartomatous fat with unclear relation to the neural tissue and its vascularity.[16] Exploration of the intradural part of lipoma is important and should be proceeded

carefully. At surgery, an effective approach involves the exposure of the normal dura and the upper part of the lipoma for at least two vertebral levels. Laminectomy of the adjacent normal lamina may be necessary to expose the lesion. Subcutaneous lipoma is usually resected completely. As to the intradural part of lipoma, it is still controversial over how much of the lipoma should be removed and opinions vary from minimal, partial, to complete or near complete removal.[7,11] Roots are intermingled within a lipoma and are difficult to separate. But there are no neural elements within the superficial lipomatous tissue. No attempt has been made to establish a plane of cleavage between the lipoma and the conus. As recommended by many researchers, a thin sheet of lipoma is left adherent to the conus in all cases.[17,18] Total removal of a lipoma carries an intrinsic risk of injury to the posterior columns and provokes a durable postoperative pain in the lower extremities and lower urethral canal. For these reasons, we advocate subtotal rather than total removal. After most of the lipoma is removed, the lipoma-placode interface is readily followed because it is made of connective tissue with strong resistance and whitish color and subjected to hemorrhage from blood vessels.

Untethering of the spinal cord is the goal of the treatment for LMM, which is achieved by sectioning the filum terminale. Section of the filum terminale is necessary in both symptomatic and asymptomatic patients to avoid progressive neurological deterioration. However, there is a risk of injury to neighboring motor and sensory nerve roots involved in bowel and bladder control. A complete transection of a single nerve root could cause neurologic deterioration immediately after operation. Therefore it is important for surgeons to recognize the filum terminale and differentiate it from functional nerve roots at surgery. Intraoperative neurophysiological monitoring, including somatosensory evoked potentials,[19-22] electromyography,[19-21,23] and compound muscle action potentials,[20,22,23] have been used to distinguish the filum terminale from functional nerve roots. Unfortunately, in some cases, intraoperative neurophysiological monitoring can not be established prior to the sectioning, and it would be ideal to visually distinguish between the filum terminale and the nervous tissue to allow sectioning of the former while sparing the latter. In view of this, we have established a technique to identify the filum terminale by visual inspection, which is based on the intrinsic anatomical differences between the filum terminale and the cauda eguina. The filum terminale is known as a fibrovascular band that is composed mainly of 5- to 20-μm thick longitudinal bundles of type 1 collagen separated by 3- to 10-μm intervals.[24] In our practice, it is initially recognized by its whitish color in superficial tissues, and subsequently ascertained by a 0.5-cm long longitudinal cut profile which appears notably to be fibrous tissues but not nerve fibers. In three of our patients we mistook the cauda eguina for the filum terminale initially, but identified the tight filum terminale after careful observation of the longitudinal cut profiles. Thus this technique helps clinicians reassure the filum terminale and avoid mistakes. Besides, such a short longitudinal cut on nerve bundles will not influence the function of the nerves, particularly the cauda eguina. If the cauda eguina is mistakenly cut open at surgery, the lesion will not result in signs and symptoms of neurologic involvement of the lower extremities, bowel and bladder postoperatively. All our patients underwent this procedure, and no postoperative deterioration was observed. In short, our technique allows anatomical identification of nerve roots and fibrous structures, such as the cauda eguina and the filum terminale during the operation.

The incidence of retethering after the initial surgery is between 3.4% and 20.2%.[1,11,12,17,25,26] Retethering results from postoperative dural adhesion. The use of artificial dural graft may also prevent from cord retethering. In fact, the pia mater surrounded by cerebrospinal fluid appears to be the best protector against adhesions and retethering. In patients with LMM, the intradural part of lipoma shares a similar structure on the surface—a membrane surrounding the fatty mass. This membrane is continuous with the pia mater covering the adjacent spinal cord and is essentially pia mater. Most surgeons ignor this membrane at surgery. To avoid postoperative retethering, we preserve this part of the pia mater covering the intradural part of a lipoma, and if possible, the cord is re-convoluted with a pia to pia closure to maintain the surface of the spinal cord smooth and complete. Postoperative 1-year follow-up MRI showed that the spinal cord was untethered to the dura mater in all of our patients and no patient developed symptoms of re-tethering. Our results suggest that suturing of the spinal pia mater is an effective way to prevent re-tethering.

Surgical treatment has been widely used for symptomatic patients to reduce the bulk of lipoma and to untether the cord. But the management of asymptomatic patients is still controversial.[7,11] Conservative treatment has been used by some researchers[7-9] because of operative risk and its unclear preventive effect. Late worsening after operation was found in almost 50% of cases and postoperative deterioration in 4%,[7] and the incidence and patterns of neurological deterioration were very similar regardless of whether early surgery was performed or not.[8] But in patients with LMM, neurological function is frequently intact at birth but insidious deterioration increases with age.[15] The likelihood of reversing neurological deficit after surgical treatment decreases with age. Hence it is preferable

to perform early operation on children with LMM before they have developed any neurological deficit. And the likelihood of increasing a neurological deficit after surgical therapy is minimal.[2] In children, all forms of spinal dysraphia are indicated for surgery. Our asymptomatic patients remained neurologically intact after early operation, and the patients under 3 years old obtained a functional improvement of more than one Hoffman's grade, which demonstrate the significant help of early prophylactic surgery for asymptomatic LMM in improving cosmesis and preventing development of neurological deterioration. We favor surgery for LMM patients within 3-6 months of birth or at their earliest detection after this age.

In conclusion, early operation for LMM patients, even asymptomatic ones, should be performed to prevent the development of neurological deficits. Subtotal excision of lipoma, suturing of the spinal pia mater, and sectioning of the filum terminale are recommended in the surgical treatment of LMM. The longitudinal cut of the filum terminale as we have used is a simple and practical way to identify the filum terminale by visual inspection. And suturing of the spinal pia mater is of vital importance in preventing postoperative tethering.

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References

1 Sutton LN. Lipomyelomeningocele. Neurosurg Clin N Am 1995;6:325-338.
2 Hoffman HJ, Taecholarn C, Hendrick EB, Humphreys RP. Management of lipomyelomeningoceles. Experience at the Hospital for Sick Children, Toronto. J Neurosurg 1985;62:1-8.
3 Kang JK, Lee KS, Jeun SS, Lee IW, Kim MC. Role of surgery for maintaining urological function and prevention of retethering in the treatment of lipomeningomyelocele: experience recorded in 75 lipomeningomyelocele patients. Childs Nerv Syst 2003;19:23-29.
4 Tortori-Donati P, Rossi A, Cama A. Spinal dysraphism: a review of neuroradiological features with embryological correlations and proposal for a new classification. Neuroradiology 2000;42:471-491.
5 Sandberg DI, Lavyne MH. Symptomatic spinal epidural lipomatosis after local epidural corticosteroid injections: case report. Neurosurgery 1999;45:162-165.
6 Schut L, Bruce DA, Sutton LN. The management of the child with a lipomyelomeningocele. Clin Neurosurg 1983;30:464-476.
7 Pierre-Kahn A, Zerah M, Renier D, Cinalli G, Sainte-Rose C, Lellouch-Tubiana A, et al. Congenital lumbosacral lipomas. Childs Nerv Syst 1997;13:298-334.
8 Kulkarni AV, Pierre-Kahn A, Zerah M. Conservative management of asymptomatic spinal lipomas of the conus. Neurosurgery 2004;54:868-875.
9 Van Calenbergh F, Vanvolsem S, Verpoorten C, Lagae L, Casaer P, Plets C. Results after surgery for lumbosacral lipoma: the significance of early and late worsening. Childs Nerv Syst 1999;15:439-442.
10 Bassett RC. The neurologic deficit associated with lipomas of the cauda equina. Ann Surg 1950;131:109-116.
11 Xenos C, Sgouros S, Walsh R, Hockley A. Spinal lipomas in children. Pediatr Neurosurg 2000;32:295-307.
12 La Marca F, Grant JA, Tomita T, McLone DG. Spinal lipomas in children: outcome of 270 procedures. Pediatr Neurosurg 1997;26:8-16.
13 Chapman PH. Congenital intraspinal lipomas: anatomic considerations and surgical treatment. Childs Brain 1982;9:37-47.
14 Bruce DA, Schut L. Spinal lipomas in infancy and childhood. Childs Brain 1979;5:192-203.
15 Kanev PM, Lemire RJ, Loeser JD, Berger MS. Management and long-term follow-up review of children with lipomyelomeningocele, 1952-1987. J Neurosurg 1990;73:48-52.
16 Lellouch-Tubiana A, Zerah M, Catala M, Brousse N, Kahn AP. Congenital intraspinal lipomas: histological analysis of 234 cases and review of the literature. Pediatr Dev Pathol 1999;2:346-352.
17 Muthukumar N. Congenital spinal lipomatous malformations: part II—Clinical presentation, operative findings, and outcome. Acta Neurochir (Wien) 2009;151:189-197.
18 McLone DG, Naidich TP. Laser resection of fifty spinal lipomas. Neurosurgery 1986;18:611-615.
19 Albright AL, Pollack IF, Adelson PD, Solot JJ. Outcome data and analysis in pediatric neurosurgery. Neurosurgery 1999;45:101-106.
20 Kothbauer K, Schmid UD, Seiler RW, Eisner W. Intraoperative motor and sensory monitoring of the cauda equina. Neurosurgery 1994;34:702-707.
21 Legatt AD, Schroeder CE, Gill B, Goodrich JT. Electrical stimulation and multichannel EMG recording for identification of functional neural tissue during cauda equina surgery. Childs Nerv Syst 1992;8:185-189.
22 Shinomiya K, Fuchioka M, Matsuoka T, Okamoto A, Yoshida H, Mutoh N, et al. Intraoperative monitoring for tethered spinal cord syndrome. Spine (Phila Pa 1976) 1991;16:1290-1294.
23 Phillips LH 2nd, Jane JA. Electrophysiologic monitoring during tethered spinal cord release. Clin Neurosurg 1996;43:163-174.
24 Fontes RB, Saad F, Soares MS, de Oliveira F, Pinto FC, Liberti EA. Ultrastructural study of the filum terminale and its elastic fibers. Neurosurgery 2006;58:978-984.
25 Colak A, Pollack IF, Albright AL. Recurrent tethering: a common long-term problem after lipomyelomeningocele repair. Pediatr Neurosurg 1998;29:184-190.
26 Ross M, Brewer K, Wright FV, Agur A. Closed neural tube defects: neurologic, orthopedic, and gait outcomes. Pediatr Phys Ther 2007;19:288-295.

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