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Occasional Review

Neurosurgical management and pathology of lumbosacral lipomas with tethered cord

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Lumbosacral lipomas are the most common form of occult spinal dysraphism. The development of lumbosacral lipomas is from the premature disjunction of the neural tube from the surrounding ectoderm, leaving the neural plate open posteriorly and allowing for the infiltration of mesodermal tissue, including fatty tissue. Since lumbosacral lipomas are a common cause of spinal cord tethering that can lead to progressive neurological deficits, prophylactic neurosurgery for lumbosacral lipomas, including untethering of the spinal cord, is recommended. We briefly review the embryology, classification, clinical presentation, imaging evaluation, surgical indication, neurosurgical management and pathological examination that are involved in recognizing these complicated malformative pathologies.

Key words: lumbosacral lipomas, occult spinal dysraphism, premature disjunction, tethered cord, untethering.

INTRODUCTION

Spinal lipomas, in particular lipomas of the conus medullaris and terminal filum ("lumbosacral lipomas"), are the most common form of occult spinal dysraphism (spina bifida occulta), which are a broad collection of spinal malformations loosely grouped together by their common characteristic of being skin covered.[1,2] The suffix "oma" implies a neoplastic event in the pathophysiology of the lesion; however, the lumbosacral lipomas arise from a disorder of embryogenesis.[1,2] Clinically, lumbosacral lipomas are a common cause of "spinal cord tethering" that can lead to progressive neurological deficits such as bladder and bowel disturbance and sensorimotor dysfunction of the legs.[1,2] Prophylactic surgery for lumbosacral lipomas, including untethering of the spinal cord, is currently recommended, although the natural history of the lumbosacral lipoma is not fully understood.[1–5] The purpose of this article is to briefly review the embryology, classification, clinical presentation, imaging evaluation, surgical indication, neurosurgical management, and pathological examination that are involved in recognizing these complicated malformative pathologies.

EMBRYOLOGY

In normal development, there is complete disjunction of the cutaneous ectoderm from the neural ectoderm and closure of the neural tube (Fig. 1A–E). However, in the development of spina bifida aperta (or cystica), as typified by myelomeningocele with myeloschisis, the absence of this normal disjunction ("non-disjunction") between the cutaneous and neural ectoderms means that the neural plate remains open (open neural placode, myeloschisis) and is spread out on the surface of a fluid-filled meningeal sac throughout the spina bifida (Fig. 1F).[6]

A current theory that may account for the development of lumbosacral lipomas is that there is "premature disjunction" of the neural tube from the surrounding ectoderm, thus leaving the neural plate open posteriorly and allowing mesenchymal cells to enter this cleft, where they are induced by the primitive ependyma to form fatty tissue (Fig. 1G,H).[1,2,5,7–10]

A congenital dermal sinus, which is also one of the occult spinal dysraphisms, consists of a tract lined by stratified squamous epithelium found on or near the midline. It is thought to result from a focal premature disjunction between the ectoderm that is destined to form the neural tube and that which will form the overlying skin (Fig. 1G,I).

CLASSIFICATION

Numerous classifications have been proposed for the differing anatomical variations of lumbosacral lipomas.[1–3,11]

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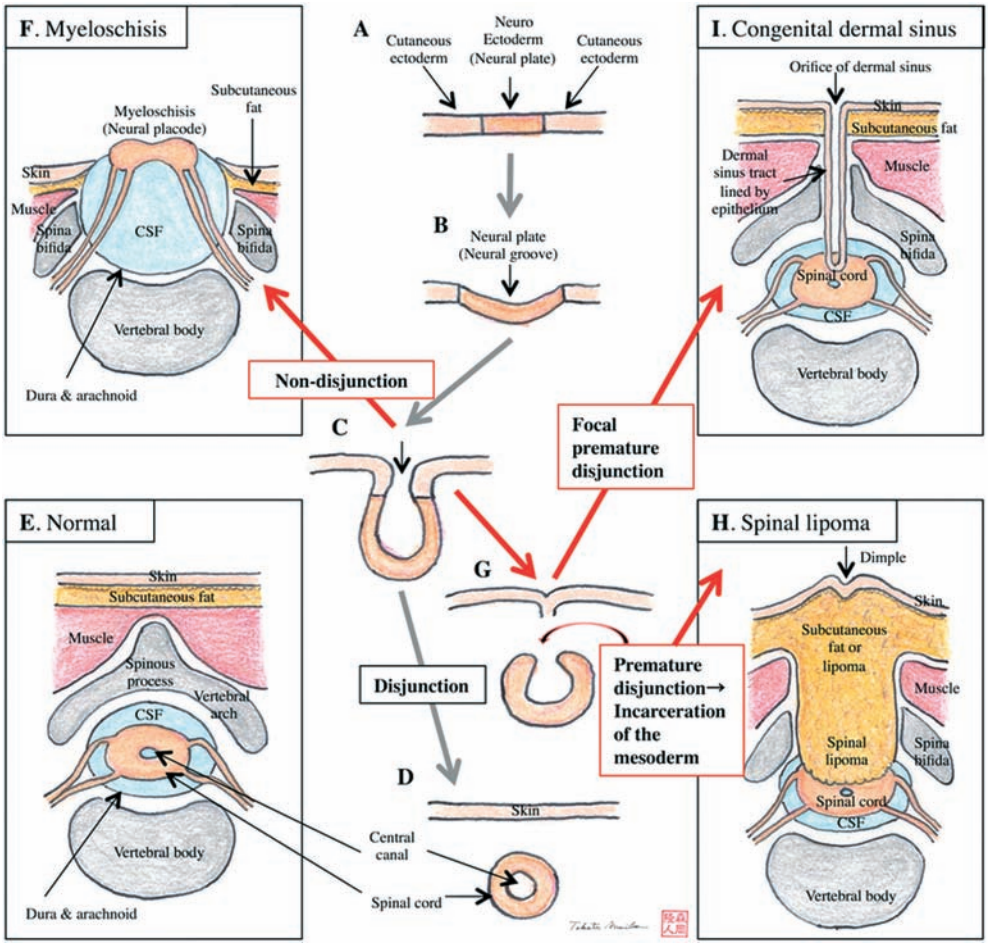

Fig. 1 Formation of the spinal cord and its surrounding structures with normal (gray bold arrows) and abnormal (red bold arrows) primary neurulation. (A) The neural plate is composed of neural ectoderm, which is continuous with cutaneous ectoderm. (B,C) The central portion of the neural plate is invaginated, forming the neural groove, and closure of the neural tube begins. (D,E) The cutaneous ectoderm fuses in the midline, forming the overlying integument. There is complete disjunction of the cutaneous ectoderm from the neural ectoderm and closure of the neural tube occurs. (F) Because of the absence of disjunction ("non-disjunction") between the cutaneous and neural ectoderms, the open neural plate (open neural placode, myeloschisis) lies spread out on the surface of a fluid-filled meningeal sac throughout the spina bifida. (G,H) When the cutaneous ectoderm prematurely separates from the neural ectoderm prior to closure of the neural tube, the surrounding mesoderm, such as subcutaneous fat, gains access to the ependymal surface of the developing neural tube. This mesoderm evolves into a spinal lipoma. (G, I) A congenital dermal sinus consists of a tract lined by stratified squamous epithelium found on or near the midline, and is thought to result from a focal premature disjunction between the ectoderm that is destined to form the neural tube and that which will form the overlying skin.

The simplest is the division of lumbosacral lipomas into two broad categories, as proposed by Pierre-Kahan *et al.*:[12] lipomas of the terminal filum (filar lipomas) and lipomas of the conus medullaris (conus lipomas). Chapman (1982) further classified conus lipomas into three types: caudal, dorsal and transitional types, based on the site of attachment of the lipoma to the cord and the position of the posterior nerve roots in relationship to the lipoma;[13] however, there is considerable confusion in the literature regarding their terminology and proper classification.[3] In contrast, the classification of lumbosacral lipomas into five types (filar type lipoma, caudal type lipoma, dorsal type lipomas, combined (or transitional) type lipoma, and lipomyelomeningocele) proposed by Arai *et al.*[14] is thorough and widely accepted, especially in Japan (Fig. 2). In this manuscript, terminology is based on Arai's classification.

CLINICAL PRESENTATION

The initial clinical presentation of a child with lumbosacral lipoma is variable and depends largely on their age.[1,2] Older children often present with neurological symptoms attributed to the tethering of the spinal cord, whereas younger children without neurological deficits are more likely to present with cutaneous stigmata, such as a subcutaneous mass (Fig. 3A), dimple (Fig. 3A), focal hypertrichosis, tail-like skin appendage (Fig. 4A) or capillary hemangioma.[1,2] These cutaneous stigmata are a

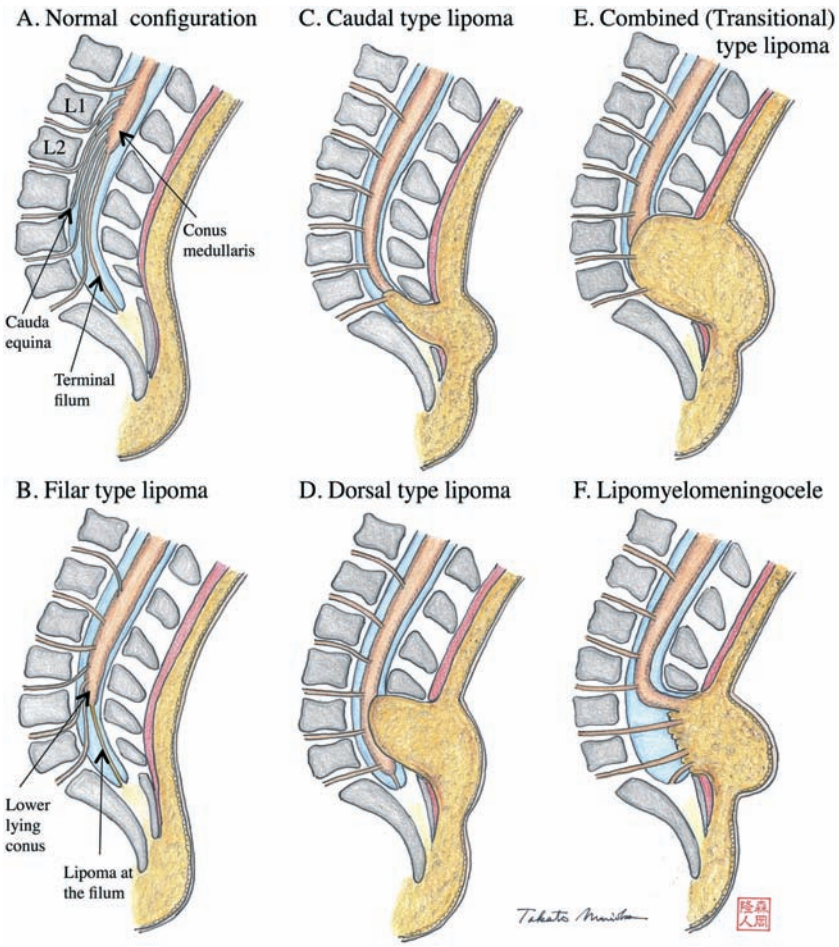

Fig. 2 Classification of lumbosacral lipomas proposed by Arai *et al.* (2001). (A) The normal anatomical relationship between the conus medullaris, cauda equina and terminal filum. The caudal end of the conus is located at the vertebral level of L1–2. The cauda equina runs in a rostro–caudal direction. (B) Filar type lipoma. A thin lipoma attaches to the caudal portion of the conus and caudally extends to the terminal filum. Note the lower-lying conus. The cauda equina runs in a caudo–rostral direction. (C) Caudal type lipoma. A relatively small lipoma, in most cases fusiform in shape, extends rostrally into the subarachnoid space by penetrating the dehiscence of the caudal-most end of the dural sac, and intermingles with the most caudal portion of the spinal cord. (D) Dorsal type lipoma. An extradural lipoma extends into the intradural space by penetrating the dehiscence in the dorsal aspect of the dural sac, and intermingles with the dorsal portion of the distal spinal cord. (E) Combined (or transitional) type lipoma. An extradural lipoma penetrates the dorsolateral and caudal dehiscence of the dural sac and intermingles with the conus. The axis of the spinal cord sometimes rotates with a shift to the side of the dural dehiscence. (F) Lipomyelomeningocele. A lower-lying tethered spinal cord enters an extraspinal and subcutaneous cystic meningeal sac and terminates in the lipomatous tissue on the wall of the meningeal sac.

tell-tale sign of occult spinal dysraphism and usually provide a clue for the initial diagnosis. Approximately 90% of children with lumbosacral lipomas have at least one cutaneous stigmata.[2]

IMAGING EVALUATION

The evaluation of lumbosacral lipomas has been greatly enhanced by MRI.[1,2] The T1- and T2-weighted images (T1WI and T2WI, respectively) are extremely useful in planning surgery because they show the neurosurgical anatomy of the lesion and its relationship to the surrounding structures (Fig. 3B, Fig. 4B). MRI also aids in detecting associated malformations, such as split cord malformations and syringomyelia. The conus medullaris is low-lying in the majority of cases and is an indication for untethering surgery in patients with filar type lipomas (Fig. 2A,B). However, the 2-5 mm-thick sections provided by conventional MRI are insufficient for detailed imaging, especially in infants. We previously reported the usefulness of three-dimensional T1WI (3D-T1) and heavily T2WI (3D-hT2), such as 3D Fourier transformation constructive interference in steady state (CISS), to demonstrate the fine anatomical structures of lumbosacral lipomas.[15–18] Another advantage of using 3D-T1 and 3D-hT2 is the flexibility that they provide for creating curvilinear or curved parallel reconstructions.[15–18] Because some patients have deformities of the vertebral column such as lordosis and scoliosis, this method is useful in demonstrating on a single plane the complicated anatomical relationship between the spinal cord and the lipoma. Curved parallel coronal reconstructions can provide an identical view to the operative findings, which cannot be performed with conventional T1WI and T2WI (Fig. 3C,D, Fig. 4C,D). Postoperatively, untethering and retethering of the cord are also evaluated with MRI, especially 3D-T1 (Fig. 3E, Fig. 4E).

SURGICAL INDICATION

When considering surgery in a patient with a newly diagnosed lumbosacral lipoma, it is important to consider the type of lipoma and whether the patient is symptomatic.[1–5,13,15] The decision to operate is straightforward in all symptomatic patients and in patients with asymptomatic filar lipomas with lower-lying coni. However, the decision to operate in asymptomatic patients with conus lipomas is controversial. Some physicians advocate prophylactic surgery for all patients, regardless of symptoms, while others propose that

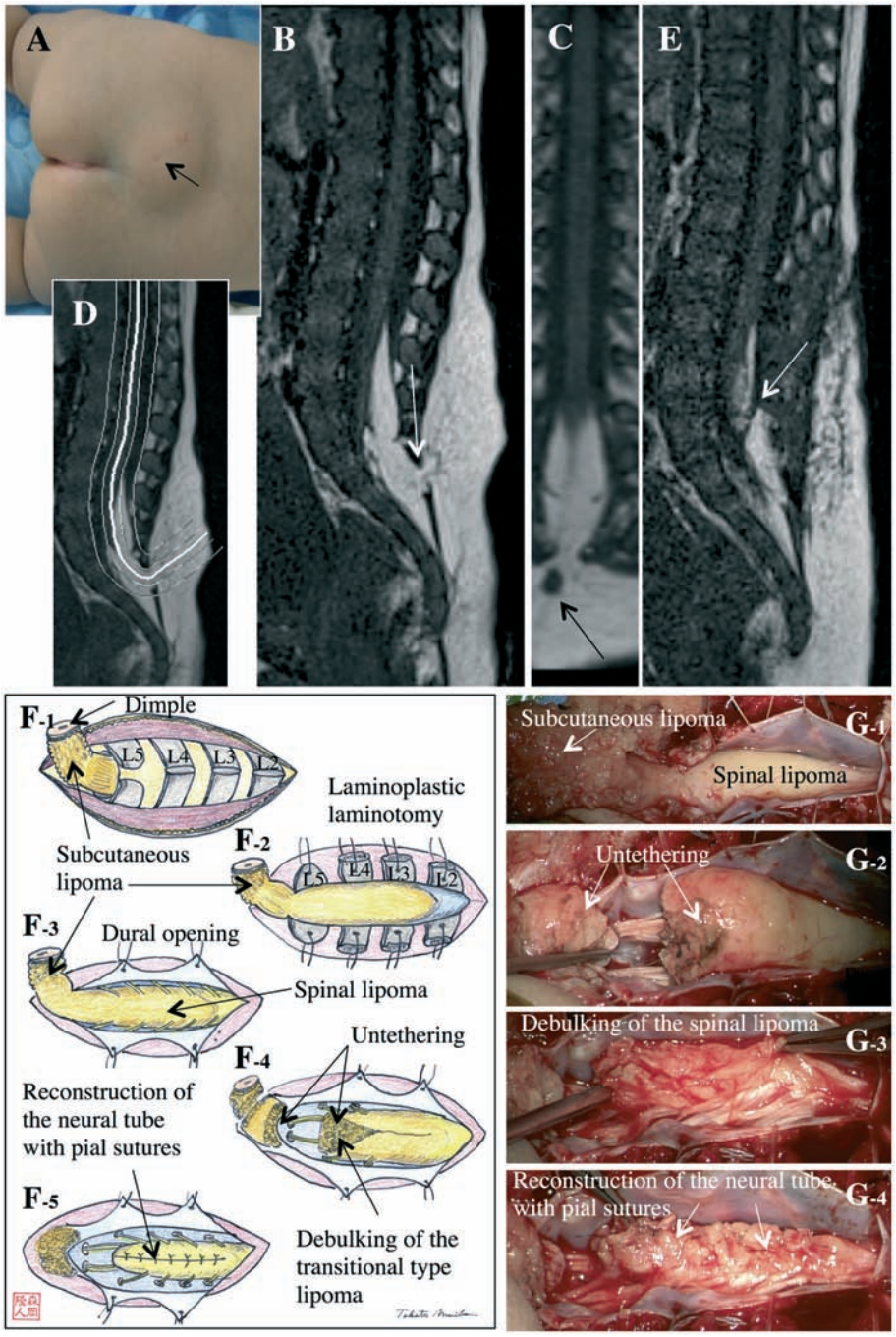

Fig. 3 Case 1. A 4-month-old boy with transitional type lipoma. (A) Photograph showing the subcutaneous mass with a dimple (black arrow). (B) Sagittal view of a three-dimensional T1-weighted image (3D-T1) shows a lower-lying conus that is tethered by the spinal and subcutaneous lipomas (white arrow). (C) Curved parallel coronal reconstructions of 3D-T1 along with the spinal cord, spinal lipoma and subcutaneous lipoma (D) provide an identical view of the operative findings. Cartilage is visible as a low-intensity mass (black arrow) in the subcutaneous lipoma. (E) Post-operative 3D-T1 shows untethering of the cord (white arrow) and rostral movement of the conus. (F) Schematic drawing and (G) microscopic view of operative findings. The subcutaneous lipoma is continuous with the spinal lipoma, and is of a transitional type (F-1,2,3, G-1). Untethering of the cord is performed at the caudal end of the dural sac (F-4, G-2). After debulking the lipoma (G-3), reconstruction of the neural tube is performed using pial sutures (F-5, G-4).

the surgical option be withheld until symptoms develop, because conus lipomas, especially transitional-type lipoma and lipomyelomeningocele, have relatively high surgical morbidity.[1–4,14] A resolution to this debate cannot be reached based on current data; however, Kanev and Bierbrauer have shown that neurological deterioration demonstrates a logarithmic relationship with advancing age.[19] Further evidence from a large number of studies suggests that any loss of neurological function is unlikely to be regained in the majority of cases.[1,2] Additionally, the rate of surgery-related morbidity has decreased substantially due to acquired experience and newer technologies, making the risk–benefit ratio of prophylactic surgery an evolving and dynamic concept.[1,2,4,5]

SURGICAL TREATMENT

Because lumbosacral lipomas are not neoplasms, the surgical goal is not total removal of the lipoma, but rather the protection of neurological function and the prevention of delayed neurological decline attributed to a tethered cord. The surgical principles employed to meet these goals

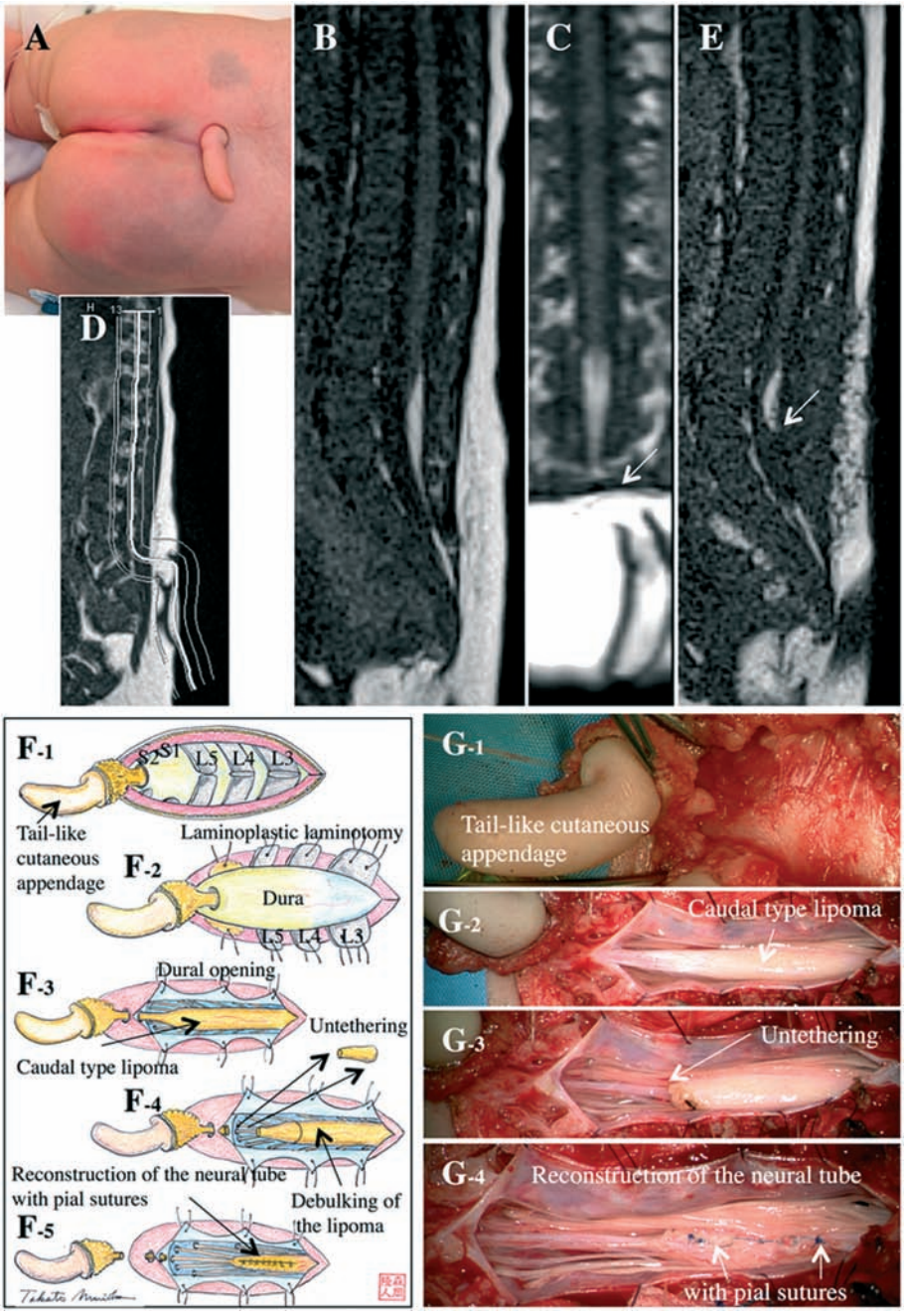

Fig. 4 Case 2. A 3-month-old girl with caudal type lipoma. (A) Photograph showing a tail-like skin appendage. (B) Sagittal view of a three-dimensional T1-weighted image (3D-T1) shows a lower-lying conus that is tethered by the spinal lipoma. (C) Curved parallel coronal reconstructions of 3D-T1 along with the spinal cord, spinal lipoma and skin appendage (D) fail to reveal the communication between the spinal lipoma and the skin appendage (white arrow). (E) Post-operative 3D-T1 shows untethering of the cord (white arrows) and rostral movement of the conus. (F) Schematic drawing and (G) microscopic view of operative findings. The skin appendage is continuous with the spinal lipoma of a caudal type (F-1,2,3, G-1,2). Untethering of the cord is performed at the caudal end of the dural sac (F-4, G-3), while the preoperative imaging failed to reveal the communication. After debulking the lipoma, reconstruction of the neural tube is performed using pial sutures (F-5, G-4).

include: (i) untethering of the spinal cord (Fig. 5A-1,2, 5B-1,2); (ii) debulking of the lipoma (Fig. 5A-3, B-3); and (iii) re-establishing normal anatomical structures (such as reconstructing the neural tube with pial sutures) (Fig. 5A-4, B-4).[1,2] The specifics of the surgical procedure vary somewhat depending on the surgeon's preference, and several excellent detailed descriptions have been published.[1,2,5,12,13] Here we offer a brief summary of our technique.

Through a midline skin incision, the lumbodorsal fascia is exposed in a rostral–caudal direction until the defect through which the lipoma traverses is found (Fig. 5C-1,2). Next, the fascia is opened and the paraspinous muscles are stripped in preparation for the laminotomy (Fig. 5C-3). After the laminotomy, the dural surface and the site where the subcutaneous lipoma enters intradurally through the dural defect are exposed (Fig. 5C-4). Under an operative microscope, the dura is opened in a rostro–caudal direction to expose a normal subarachnoid space rostral to the spinal lipoma. As the dural defect is approached, care is taken to avoid injury to neural elements, especially the spinal roots (Fig. 5C-5). The monitoring of evoked electromyography (EMG) from the external anus and leg muscles can aid in safe dissection.[2] Once the caudal end of the dural sac is

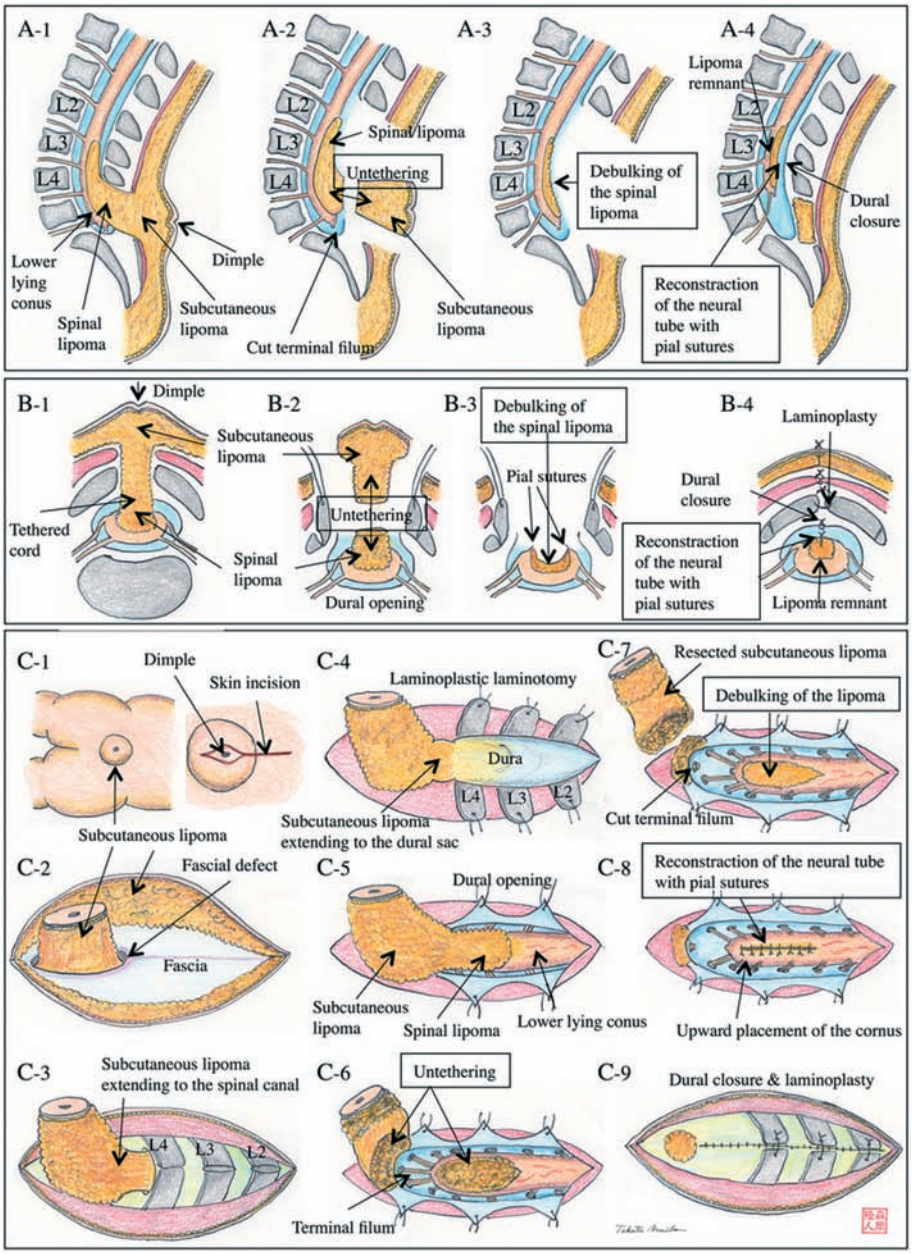

Fig. 5 Schematic drawing of the surgical procedures for spinal lipoma of a dorsal type with tethered cord. (A, B) Sagittal and axial views show the spinal lipoma with lower-lying conus (A-1, A-2). The surgical principles include untethering the spinal cord (A-2, B-2), debulking the lipoma (A-3, B-3) and reconstructing the neural tube with pial sutures (A-4, B-4). (C) Operative views of this case. A detailed explanation is given in the text.

identified and the lipoma is freed from the surrounding tissue, untethering of the cord can be performed (Fig. 5C-6).

The terminal filum is cut when no evoked EMG is obtained (Fig. 5C-7). By using an ultrasound aspirator, we debulk enough of the lipoma so that the neural placode can be reconstituted with pial sutures (Fig. 5C-7). Although there is significant disagreement on the amount of lipoma that should be removed,[5,12,20–22] we do not pursue complete resection. We reconstruct the neural tube using pial sutures, to decrease the risk of retethering by reducing the amount of raw fatty tissue exposed (Fig. 5C-8).[23] Although it is unclear if the incidence of retethering is truly reduced by this technique, it is conceivable that closing by non-absorbable threads makes second operations easier by reconstituting a more normal anatomy.[2] Finally, we reconstruct the dura and perform the laminoplasty (Fig. 5C-9).

Figure 3 (F,G) and Figure 4 (F,G) show schematic drawings and photos of the individual operative view in a 4-month-old boy with transitional-type lipoma and a 3-month-old girl with caudal-type lipoma, respectively.

PATHOLOGICAL EXAMINATION

Histologically, lumbosacral lipoma consists of mature adipose tissue arranged in lobules with loose connective tissue (Fig. 6A), as is seen in the subcutaneous adipose tissue (Fig. 6D), or often with a striking amount of fibrous tissue interspersed between the adipose lobules.[24]

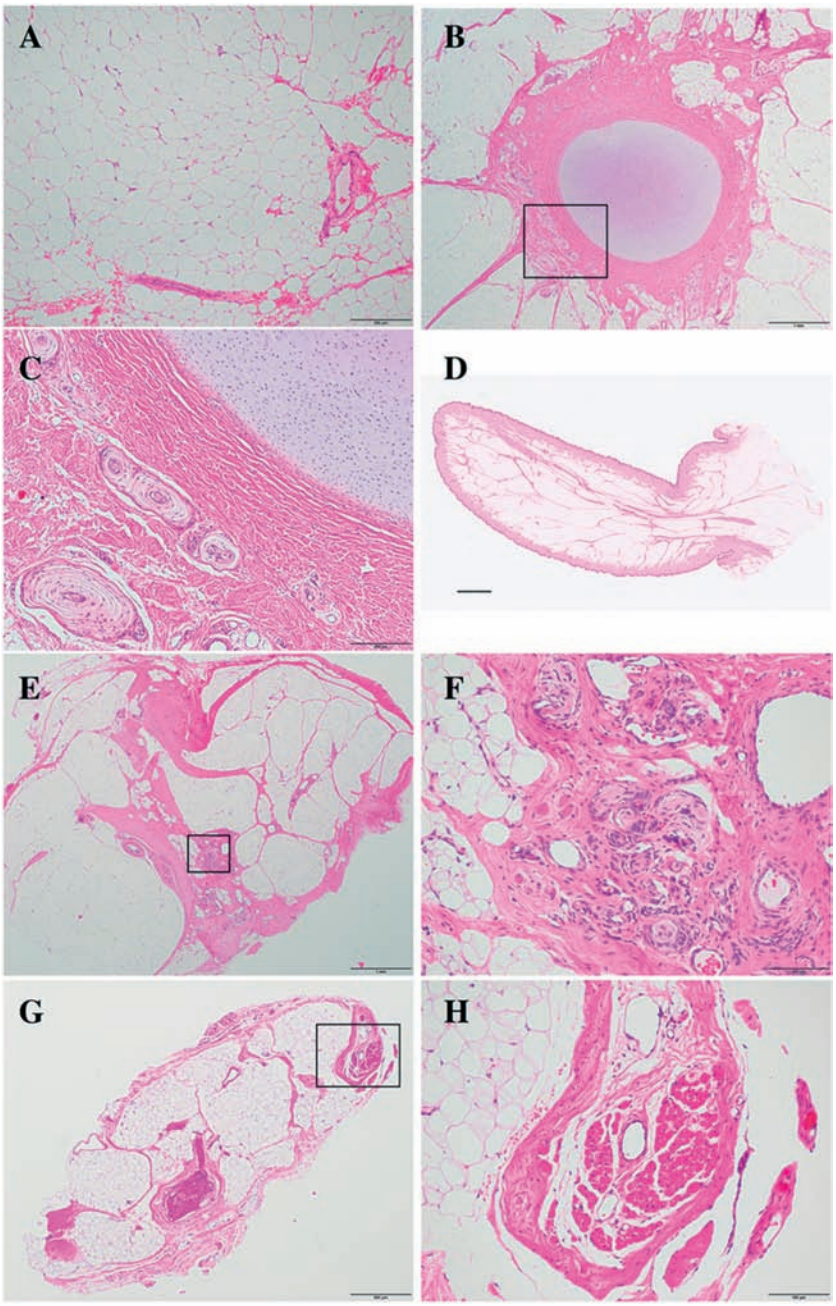

Fig. 6 (A-C) Histological findings in Case 1. (A) The lipoma consists of mature adipose tissue with loose connective tissue. (B) There is a small, relatively circular hyaline cartilage tissue embedded in the subcutaneous adipose tissue. (C) Higher magnification (from the square in B) reveals numerous Vater-Pacini corpuscles surrounding the cartilage. (D-H) Histological findings in Case 1. (D) The skin appendage consists of mature adipose tissue covered with skin tissue. The continuing extradural (E,F) and intradural (G,H) components of the lipoma, respectively, show skeletal muscle fibers in the fibrous connective tissue. Black bar indicated 200 μm in A and C, 1 mm in B and E, 3 mm in D, 100 μm in F and H, and 500 μm in G, respectively.

As previously described in the embryology of the lumbosacral lipoma, premature disjunction of the neural tube from the surrounding ectoderm occurs, leaving the neural plate open posteriorly and allowing mesenchymal cells to enter this cleft.[1,2,5,8–10] However, lipomas do not consist entirely of mesenchymal tissues, with a variety of ectodermal and endodermal tissues also present.[24] In a large series of 234 cases,[24] the most frequently encountered tissues were striated skeletal muscle (37%) (Fig. 6E-H), sensory nerve corpuscles such as Vater-Pacini corpuscles (21%) (Fig. 6B,C), neuroglial tissue (18%), meningeal tissue (7%) and cartilage (6%) (Fig. 6B,C). Although some researchers have postulated that lumbosacral lipomas may be a form of teratoma,[11,24,25] lumbosacral lipomas are congenital, complex lesions in which adipose tissue is associated with a variety of tissue patterns.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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