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Case Reports & Case Series

Spinal intramedullary dermoid cyst associated with filar lipoma: A case report and literature review

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ABSTRACT

Background: Several authors have reported the various patterns of coexistence of spinal dermoid cysts and lipoma; however, the association of intramedullary dermoid cysts with lipoma is extremely rare. In addition to the embryological and pathological aspects of this rare condition, we discuss the feasibility of combined microscopic and endoscopic procedures for the management of intramedullary dermoid cysts.

Case description: An 18-year-old woman presented with right buttock pain. Magnetic resonance (MR) imaging revealed a large, well-defined mass extending from L2 to L4. The conus medullaris terminated in the mass and was tethered by a fatty filum. According to the signal intensities on MR images, the mass could be divided into two components. The upper component existed intramedullary and was iso- to hypo-intense relative to the spinal cord on T1-weighted images. The lower component exhibited homogeneous hyper-intensity signals on both T1- and T2-weighted images. Partial removal of the cyst wall and evacuation of the cyst contents followed by untethering of the spinal cord were performed by the combined microscopic and endoscopic procedures. The patient's symptoms were relieved postoperatively and pathological studies confirmed the diagnosis of dermoid cysts associated with lipoma.

Conclusions: We present a rare case of an embryological "collision" of an intramedullary dermoid cyst associated with filar lipoma underlining the spectrum of intradural pathologies in spinal dysraphism. Surgical management is creative in these circumstances and multimodal. Our surgical management shows that the use of endoscopes can be effective in the surgical removal of long sectional spinal dermoid cysts.

1. Introduction

Spinal dermoid cysts are gradually progressive benign tumors, which are covered by keratinized squamous epithelium. They are distinguished from epidermoid cysts by the presence of dermis and dermal glands [1]. Spinal dermoid cysts are estimated to account for 0.8–1.1% of all primary spinal tumors [2–4]. The majority of these tumors develop extramedullary or juxtamedullary in the lumbosacral region [3,4], and those that occur intramedullary are uncommon. Furthermore, cases of spinal intramedullary dermoid cysts with concurrent spinal lipoma are extremely rare [2,3,5]. In this report, we describe a rare case of intramedullary spinal dermoid cyst associated with filar lipoma, which was treated with combined microscopic and endoscopic procedures.

2. Case report

2.1. Preoperative course and neurological and radiological examinations

An 18-year-old woman presented with a 1-month history of pain in the right buttock. She complained of intermittent pain in the right S2 dermatome, which was aggravated by standing and relieved by lying down. She had no history of trauma, infection, lumbosacral surgery, congenital anomaly, or developmental disorders. No abnormal findings, with the exception of a small dimple above the anus, were detected on the skin of the lower back. On neurological examination, the muscle strength and deep tendon reflexes were normal. Urinary and bowel functions were intact. Magnetic resonance (MR) imaging of the lumbar spine demonstrated a large, well-defined mass extending from L2 to L4

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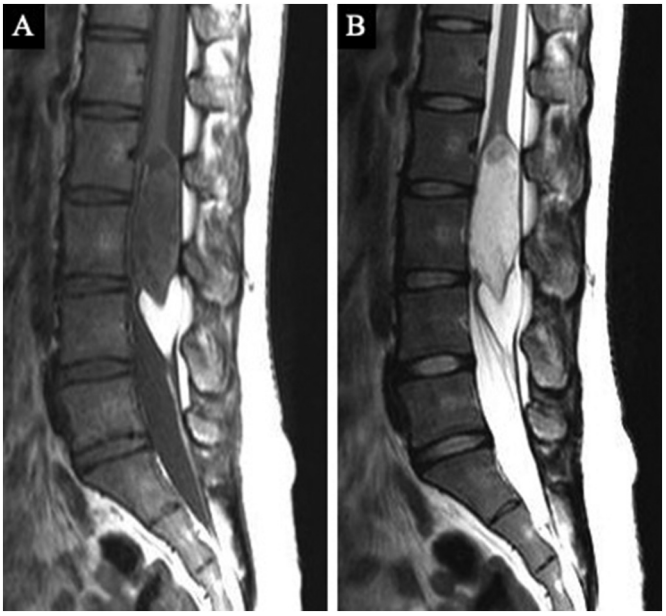

Fig. 1. Preoperative imaging findings.
Preoperative sagittal T1-weighted (A) and T2-weighted (B) magnetic resonance images revealed the intradural mass extending from L2 to L4 associated with the spinal cord, tethered by a fatty filum.

(Fig. 1). The conus medullaris terminated in the mass and was tethered by a fatty filum. According to the signal intensities on the MR images, the mass could be divided into two components. The upper component of the mass, which accounted for approximately two-thirds of the lesion, existed intramedullary and was iso- to hypo-intense on T1-weighted images and hyper-intense on T2-weighted images, relative to the spinal cord. The signals were heterogeneous on both T1- and T2-weighted images. In contrast, the lower component exhibited homogeneous hyper-intensity on both T1- and T2-weighted images and was connected to the tight filum terminale. The mass was not enhanced after administration of gadolinium. Spinal dysraphism was observed from S1 to S5, but neither a dermal sinus nor subcutaneous lipoma was found.

2.2. Operative procedures and postoperative course

The patient was intubated and placed in the prone position under intravenous general anesthesia with propofol and remifentanil. Electrodes were placed for free-run and direct stimulation of motor evoked potentials (MEPs) in the lower limbs (bilateral quadriceps femoris, biceps femoris, anterior tibialis, and gastrocnemius) and anal sphincter. The bulbocavernous reflex was also recorded for intraoperative neurophysiological monitoring. An incision was made in the skin between the levels of the spinous processes of L1 and L5, and laminoplastic laminotomies were performed at L2, 3, and 4. The dura mater was intact and incised at the midline. The spinal cord was bulging at the L2-L3 level and the intradural lipoma was located adjacent to the conus medullaris (Fig. 2A). Debulking of the lipoma from the caudal side to the rostral side was performed under an operative microscope (OPMI PENTERO 800, ZEISS) (Fig. 2B). The rostral margin of the lipoma was adjacent to the shiny white intramedullary mass via a thin capsule. Using the microscope, evacuation of the intramedullary mass was performed (Fig. 2C). To explore the rostral margin of the intramedullary mass, another small window (5 mm) was made at approximately 3 cm rostral to the conus medullaris along the posterior median sulcus (Fig. 2D, E). A 2.7 mm rigid endoscope (OLYMPUS Optical Co.) was then introduced into the operative field and supported by EndoArm (OLYMPUS Optical Co.) (Fig. 3A, B). Using the endoscopic view, evacuation of the intramedullary mass was performed using a ring curette and suction through the small myelotomy (Fig. 4A). A yellowish, lipid-rich material that contained tiny hairs and a small amount of calcification was confirmed through endoscopic view. Partial removal of the cyst wall and evacuation of the cyst content were conducted by the combined use of endoscope and microscope. Subsequently, untethering of the cord with careful electrophysiological monitoring was performed under the microscope. The pial edges were sutured with 5-0 nylon sutures, and the dura mater was closed using VCS clips (AnastoClip VCS, LeMaitre VASCULAR). MEP monitoring was intact throughout the surgery, and the patient exhibited no postoperative neurological deterioration. Postoperative MR images indicated subtotal removal of the intramedullary mass and untethering of the spinal cord (Fig. 4B).

2.3. Histopathological examination

The specimens were fixed in formalin before being processed into paraffin sections. Paraffin-embedded samples were processed for hematoxylin-eosin staining. The intradural lipoma mainly consisted of mature adipocytes and contained Pacinian corpuscles (Fig. 5A). The intramedullary mass contained abundant yellowish lipid-rich debris in which hairs and a small amount of calcification were present. The wall of the mass was lined with stratified squamous epithelium and contained sebaceous glands and sweat glands, which confirmed the diagnosis of a dermoid cyst (Fig. 5B).

3. Discussion

Cases of dermoid cysts associated with spinal lipoma are rare [6–9]. Table 1 indicates the various patterns of coexistence of the two pathologically different lesions that have been described previously [6–8,10,11]. Among them, the association of intramedullary dermoid cysts with spinal lipoma is extremely rare, and only two cases have been reported to date [7,10]. The human spinal cord is formed following primary and secondary neurulation. During primary neurulation, disjunction between the cutaneous and neuro ectoderms occurs after fusion of the neural tube. At the end of primary neurulation, the caudal neuropore closes and the caudal cell mass is formed distal to the neural tube. During secondary neurulation, the caudal cell mass undergoes cavitation and connects to the central canal present in the neural tube at the junctional region, which comprises the secondary neural tube [9]. De novo spinal dermoid cysts are reported to originate from the inclusion of epithelial cell rests in the neural tube by the absence of normal disjunction between the cutaneous and neural ectoderms, and are sometimes accompanied by spinal dermal sinuses or other congenital abnormalities [4]. However, incomplete degeneration or apoptosis error of the caudal cell mass in the late phase of secondary neurulation have been reported to result in the formation of the filum terminale [12].

In the literature, Muthukumar et al. hypothesized that the co-existence of intramedullary dermoid cysts and spinal lipoma originated during secondary neurulation when the residual totipotent cells of the caudal cell mass were differentiated into two different cell types; one became the intramedullary dermoid cyst and the other became the filar lipoma [7]. Our intraoperative findings may support their theory, because the dermoid cyst existed adjacent to the lipoma and anatomical continuity was observed between the two histologically different lesions. Although a dimple on the skin of the lower back and spinal dysraphism were found from S1 to S5 in our patient, the dura was intact and neither dermal sinus nor subcutaneous lipoma was observed. This suggests that developmental errors occurred in the process of secondary neurulation rather than primary neurulation.

In general, dermoid cysts are surrounded by a capsule, which consists of epithelial lining, and adhere tightly to the adjacent neural tissue [13,14]. Although complete removal of the cyst wall may reduce the

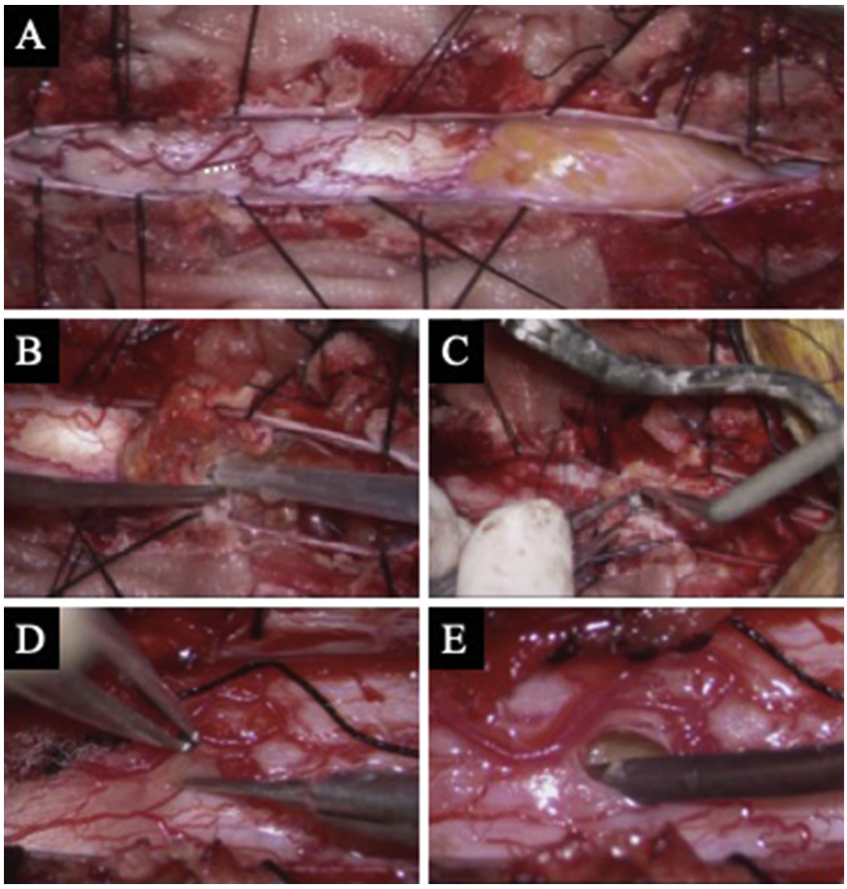

Fig. 2. Intraoperative photographs.
A: Intraoperative photograph showing swelling of the conus medullaris adjacent to the intradural lipoma. The white dotted line showing the point of myelotomy.
B: Debulking of the lipoma was performed using microscope.
C: The rostral margin of the lipoma was adjacent to the shiny white intramedullary mass via a thin capsule. Evacuation of the intramedullary mass was performed using microscope.
D, E: Small myelotomy was made at approximately 3 cm rostral to the conus medullaris along the posterior median sulcus.

recurrence rate, it also involves a risk of spinal cord damage that may cause severe postoperative neurological deterioration. It has been reported that the risk of cyst recurrence after incomplete removal does not justify the risk involved in attempting complete cyst wall resection [2,4,15].

One possible solution we devised to address this issue was to shorten the myelotomy length to the fullest extent possible, and attempt cyst removal using an endoscope. The contents of the cysts are typically soft and waxy substances, which can be easily removed by suctioning or curetting via the small surgical window. In addition, the use of an angled endoscope allowed us to change the optical axis to eliminate the blind spot and suction or curette the tumor from multiple directions.

In our case, we performed the operation to relieve the intermittent pain in the right S2 dermatome and confirm the histological diagnosis of the mass. The tethering of the spinal cord might have been the cause of the symptoms; however, we could not exclude the possibility that the intramedullary mass was inducing pain by compressing or irritating the conus medullaris. Therefore, we opted to perform untethering of the

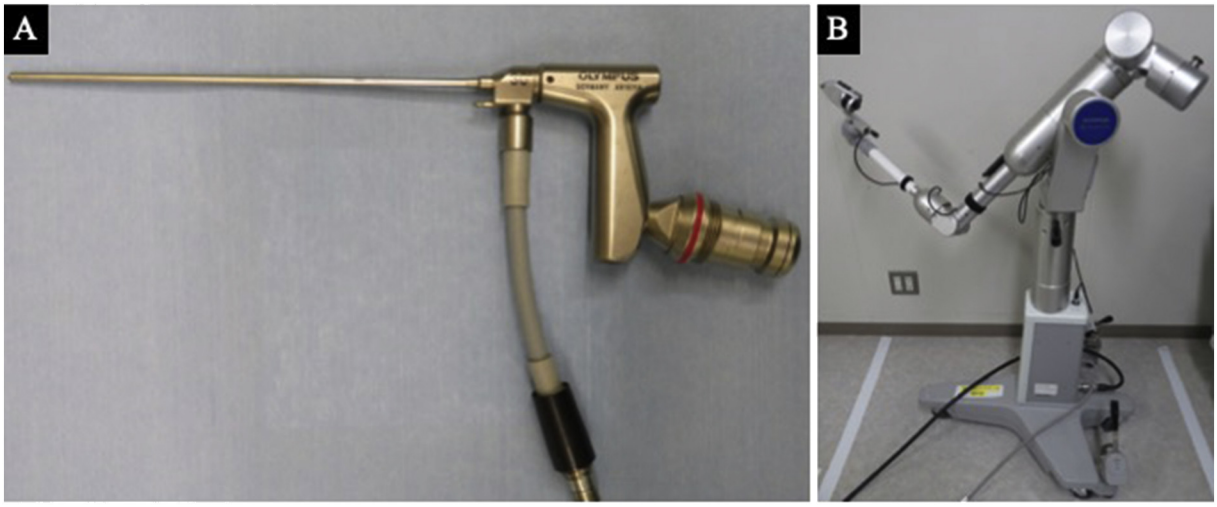

Fig. 3. Photographs of surgical instruments.
A: 2.7 mm rigid endoscope (OLYMPUS Optical Co.)
B: EndoArm (OLYMPUS Optical Co.)

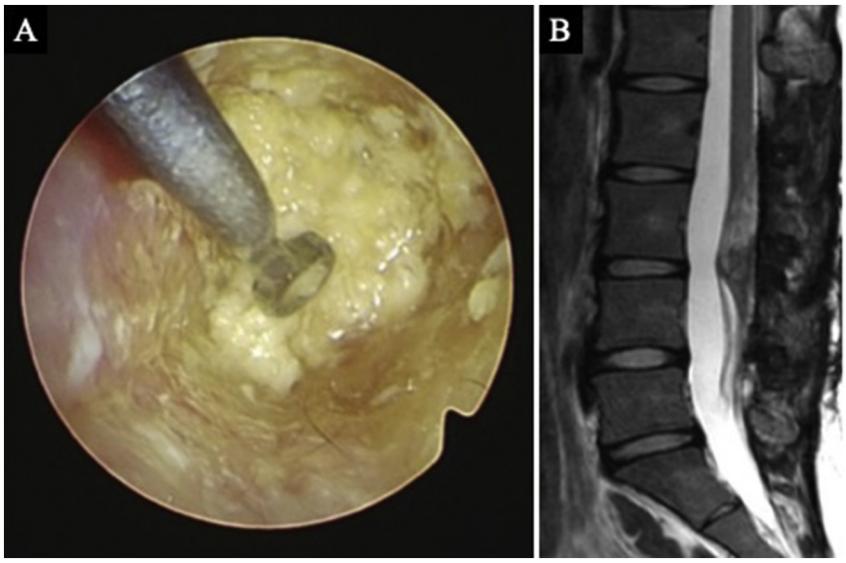

Fig. 4. Intraoperative photographs and postoperative magnetic resonance images.
A: Neuroendoscopic view of the inside of the cyst cavity showing a yellowish, lipid-rich material that contained tiny hairs and a small amount of calcification.
B: Six months postoperative sagittal T2-weighted image showing subtotal removal of the intramedullary mass and untethering of the spinal cord.

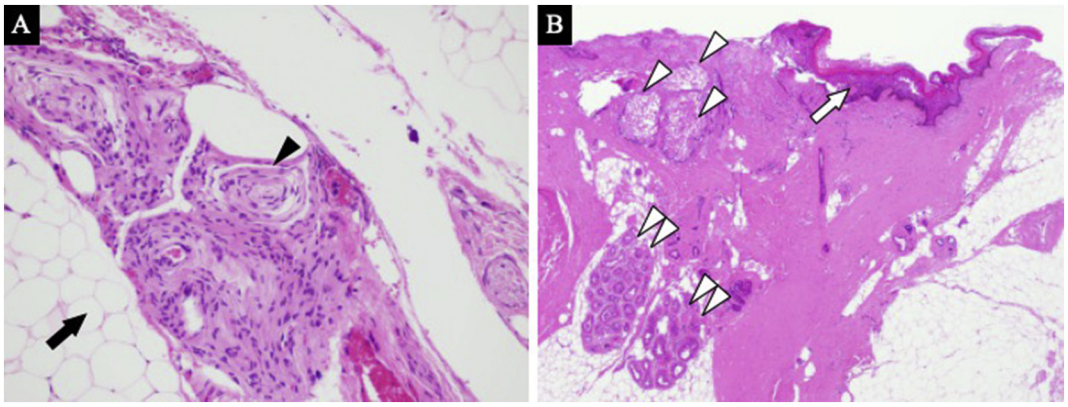

Fig. 5. Histological analysis of the specimens using hematoxylin-eosin staining.
A: The intradural lipoma mainly consisted of mature adipocytes (black arrow) and contained Pacinian corpuscles (black arrowhead) (original magnification ×200).
B: The intramedullary mass had a lining of stratified squamous epithelium (white arrow), sebaceous glands (white arrowheads), and sweat glands (white double arrowheads) (original magnification ×40).

Table 1
Reported cases of the association of spinal dermoid cysts with lipoma.

Author, year	Age/ gender	Clinical presentation	Dermal or vertebral abnormalities	Locations of dermoid cyst	Types of lipoma
Present case	18/F	Buttock pain	Dimple Spinal dysraphism	Intramedullary at conus medullaris	Filar lipoma
Garat et al. 1985 [11]	N/A	Bladder dysfunction Motor paralysis in lower extremities	Hypertrichosis	N/A	Conus lipoma
Hillman et al. 1992 [10]	6/F	Neurogenic foot deformity motor and sensory paralysis in lower extremities	Spinal dysraphism Diastematomyelia	Intramedullary at conus medullaris	Conus lipoma
	1/M	Neurogenic foot deformity	Hypertrichosis Spinal dysraphism Diastematomyelia	Extradural	Conus lipoma
Misago et al. 1996 [8]	2/M	Asymptomatic	Telangiectasia Hypertrichosis Spinal dysraphism	Subcutaneous	Extradural (subcutaneous)
Miyagi et al. 2006 [6]	6/F	Neurogenic foot deformity Bladder dysfunction	Imperforate anus Spinal dysraphism	Intradural (adjunctive to cauda equina)	Filar lipoma
Muthukumar et al. 2007 [7]	19/M	Gait disturbance Motor and sensory paralysis in lower extremities bladder dysfunction	Spinal dysraphism	Intramedullary at conus medullaris	Filar lipoma

spinal cord and mass reduction simultaneously. With endoscopic procedures, we were able to remove the cyst content via an extremely small myelotomy. Long-term follow-up with a larger number of patients is necessary to determine the recurrence rate and efficacy of the combined use of endoscopes and traditional microsurgical techniques with use of conventional microscopes in the removal of long sectional spinal dermoid cyst.

Abbreviations

MEP	motor evoked potential
MR	magnetic resonance

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Declaration of competing interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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