

Sacral myolipoma with involuntary contraction causing tethered cord syndrome: illustrative case

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BACKGROUND Spinal lipomas sometimes involve various ectopic tissues originating from the ectoderm, mesoderm, and endoderm in the process of morphological development.

OBSERVATIONS A 29-year-old male patient with myolipoma of the conus medullaris at the S2 and S3 levels was described. The unusual finding, involuntary muscle contraction, was presented in an operative video and a literature review. In the present case, sacral myolipoma with involuntary contraction caused tethered cord syndrome in adulthood, and untethering surgery resolved continuous buttock and leg pain.

LESSONS This rare finding is considered a surgical indication for adult patients with myolipoma.

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KEYWORDS spina bifida; neural tube defect; spinal dysraphism; lipoma; lipomeningocele; lipomyelomeningocele

Spinal lipomas are congenital malformations characterized by tethered cord syndrome.¹⁻³ In childhood, symptoms such as gait abnormalities with foot deformity, continuous urinary dribbling, and scoliosis are aggravated by growth spurts. Pain is not common.⁴ In adulthood, pain in the back, leg, or perineal region is common and aggravated by trauma, stretching, spinal stenosis, or progressive deposition of fat during rapid weight gain.^{2,3}

Spinal lipomas develop at the conus medullaris or filum terminale via different morphological processes.¹ Lipomas of the conus medullaris are associated with primary neurulation, in which the neural tube is formed by a change in the shape of the neural plate. Lipomas of the filum terminale are associated with secondary neurulation, in which the most caudal part of the neural tube is formed by the growth of the tail bud. Primary and/or secondary neurulation disorders lead to different types of congenital neural tube defects, including spinal lipomas.⁵ Spinal lipomas sometimes involve various ectopic tissues originating from the ectoderm, mesoderm, and endoderm in the process of morphological development.^{1,6} We herein describe an adult patient with sacral myolipoma of the conus medullaris with involuntary contraction. Unusual and previously unreported findings are presented using an operative video along with a literature review.

Illustrative Case

A 29-year-old man presented with a 2-year history of gait difficulty and urinary incontinence during walking. He had moved from a local area to Tokyo 2 years earlier and had to walk long distances because of his work. He had a previous history of untethering of the sacral lipoma at another hospital at the age of 14 years, which was attributed to distal weakness of the left leg with a foot deformity. At that time, he did not have gait difficulty or urinary incontinence and had the ability to run but underwent surgery because spontaneous neurological deterioration frequently occurred during periods of growth.

On admission to our hospital, he had distal weakness of the left leg, muscle atrophy of the left thigh and calf, and hypotrophic club foot deformity of the left foot. He walked without a stick but became fatigued when walking. He also had buttock and left leg pain with a numeric rating scale score of 3 not only during walking but also when resting on a bed. He had no previous history of trauma, stretching, or rapid weight gain.

Magnetic resonance imaging showed a low-lying conus medullaris due to a mass dorsal to the conus at the S2 and S3 levels (Fig. 1). The mass was diagnosed as a lipoma of the conus medullaris. It did not compress the spinal cord but retethered it to the

ABBREVIATIONS MRI = magnetic resonance imaging.

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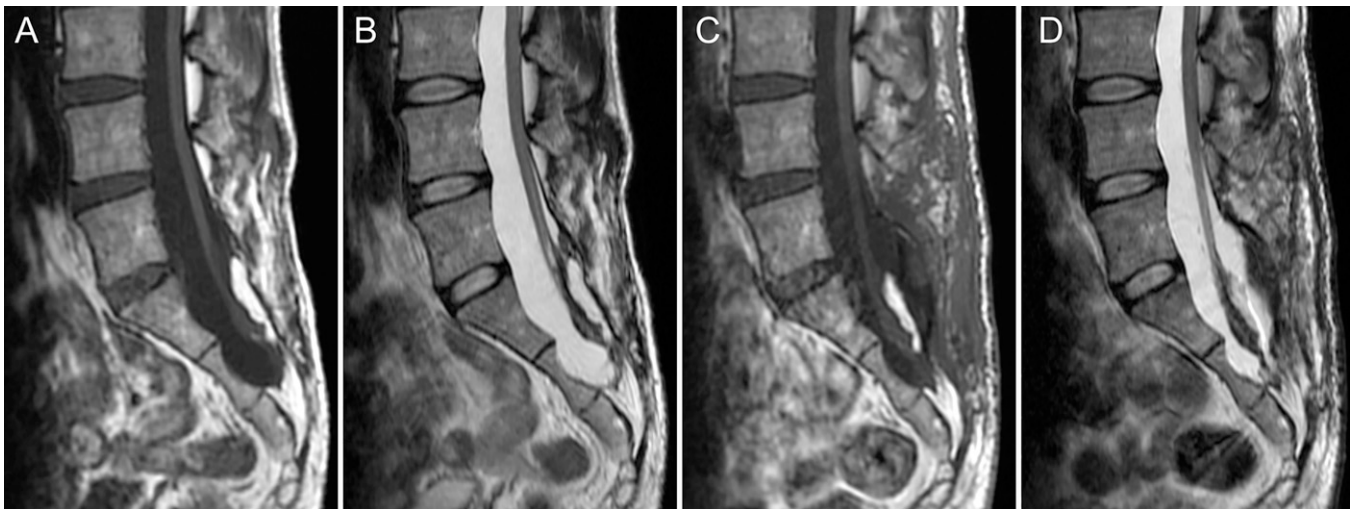


FIG. 1. Preoperative sagittal T1- (A) and T2-weighted (B) magnetic resonance imaging (MRI) showing a low-lying conus medullaris due to a high-intensity mass dorsal to the conus at the S2 and S3 levels. Postoperative sagittal T1- (C) and T2-weighted (D) MRI showing untethering of the spinal cord as well as partial removal of the myolipoma.

sacral spine. The patient underwent untethering surgery because his symptoms were potentially due to additional tugging of the spinal cord during long-distance walking.

After sacral laminotomy, the lipoma was dissected circumferentially from surrounding tissues under nerve stimulation. Intraoperatively, we noted beating of the lipoma along with the conus medullaris, similar to a heartbeat (Video 1). Muscle-like pinkish tissue was observed between the caudal aspect of the lipoma and the dura mater. Stimulation of the exiting nerve roots did not affect movement of the lipoma. The tissue contracted in an involuntary and irregular pattern. Removal of the tissue attenuated the contraction. The lipoma was partially removed, and the dura was closed with autologous tissue. A histological examination revealed mature striated skeletal muscle and adipose tissue (Fig. 2). Focal perivascular inflammation, which is a secondary change, was also detected. Postoperatively, the conus medullaris was successfully released. Although no neurological complications occurred, wound dehiscence required additional repair surgery. Buttock and left leg pain disappeared; however, there were no improvements in leg weakness, atrophy, or deformity or in urinary incontinence in the 6-month follow-up.

VIDEO 1. Clip showing involuntary muscle contraction causing additional tugging of the tight conus medullaris. [Click here to view.](#)

Discussion

Observations

The present case was unique because involuntary muscle contraction caused additional tugging of the tight conus medullaris. The patient in the present case had foot deformities diagnosed in childhood, but he remained well until the onset of new neurological deficits in adulthood. To the best of our knowledge, only five cases of lumbosacral myolipoma with muscle contraction, including the present case, have been reported in the literature (Table 1).⁷⁻¹⁰ Four patients (80%) were ≥ 18 years old at the onset of new neurological deficits. Four patients (80%) had myolipoma of the conus medullaris, whereas the other had

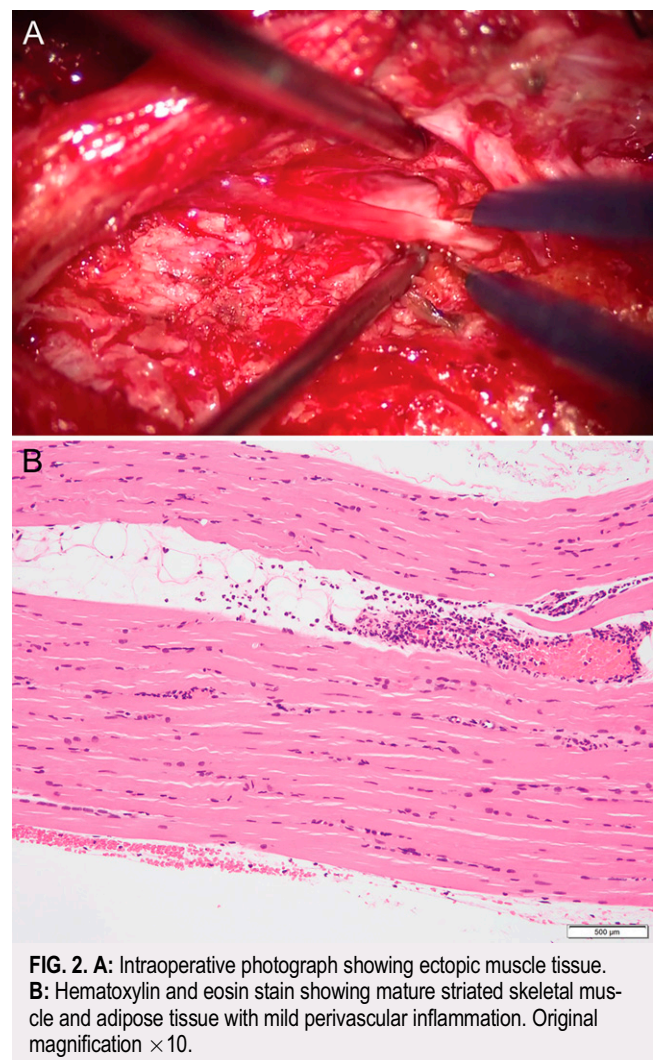


FIG. 2. A: Intraoperative photograph showing ectopic muscle tissue. **B:** Hematoxylin and eosin stain showing mature striated skeletal muscle and adipose tissue with mild perivascular inflammation. Original magnification $\times 10$.

TABLE 1. Clinical characteristics of cases of lumbosacral myolipoma with muscle contraction

Authors & Year	Age	Sex	Symptoms	Level	Lipoma Type	Striated Muscle	Contraction	Outcome
Knierim et al., 1987 ⁷	1 mo	M	Lumbosacral mass	L4–S1	Conus & filum	Intratumoral	Stimulation-induced	No change
Brown & Shaver, 2000 ⁸	18 yrs	M	Urological	S2	Filum	Intratumoral	Stimulation-induced	Amelioration of urological symptoms
Shimizu et al., 2014 ⁹	18 yrs	M	Urological, pain	S1–3	Conus	Intratumoral	Stimulation-induced	Attenuation of pain
Iliescu et al., 2015 ¹⁰	26 yrs	M	Urological, motor, sensory	S3–5	Conus	Extratumoral	Stimulation-induced	Amelioration of all symptoms
Present case	29 yrs	M	Urological, motor, pain	S2–3	Conus	Extratumoral	Involuntary	Attenuation of pain

myolipoma of the filum. Striated muscle fibers were observed intratumorally in three cases and extratumorally in two. Myolipoma contracted with nerve stimulation in four cases and was associated with contraction of the external anal sphincter⁹ or gastrocnemius muscle⁷ in one case each. Four cases (80%) showed the attenuation of preoperative symptoms.

Histologically, spinal lipomas are composed of mature adipocytes. They sometimes involve various ectopic tissues originating from the ectoderm (neuroglia and ependyma), mesoderm (muscle fibers, cartilage, and vessels), and endoderm (respiratory epithelium) in the process of primary and secondary neurulation.^{1,4,6,11} They most frequently involve striated muscle fibers originating from the mesoderm but are commonly located intratumorally, are surrounded by dense fibrosis, and do not contract.^{1,4,6}

Lessons

In the present case, sacral myolipoma with involuntary contraction caused tethered cord syndrome, and untethering surgery resolved continuous pain. This rare finding is considered a surgical indication for adult patients with myolipoma.

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Disclosures

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

Author Contributions

Conception and design: Takai, Fujimoto. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Takai, Fujimoto. Critically revising the article: Takai, Agari. Reviewed submitted version of manuscript: Takai. Approved the final version of the manuscript on behalf of all authors: Takai. Study supervision: Takai.

Supplemental Information

Video

Video 1. <https://vimeo.com/689702510>.

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