Case report

Duplicated filum terminale in non-split cord malformations: An underrecognized cause for treatment failure in tethered cord syndrome

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Context: Anatomical variations of the filum terminale (FT) have been described in association with split cord malformations (SCM) but they appear to be a rare finding in its absence. We report the first case in literature of a duplicated FT in a patient presenting with tethered cord syndrome (TCS) without any radiological evidence of SCM.

Findings: A 47-year-old man presented with invalidating back pain radiating to both legs. Magnetic resonance imaging revealed an intradural dorsal lipoma in a low-lying conus. Intraoperatively two distinct fibrous bands were anatomically and electrophysiologically identified as the FT and both were sectioned. The diagnosis of FT was confirmed for both specimens by histology.

Conclusion: In absence of SCM, a duplicated FT has not been previously described as a cause of TCS. It may be a cause of treatment failure for TCS if unrecognized on preoperative imaging and during surgery if one filum remains intact. We highlight the importance of a meticulous cauda equina dissection supported by intraoperative nerve stimulation to identify this rare anomaly. We hypothesize that this entity may represent a variant of SCM involving the caudal neural tube but which requires further validation at an embryological level.

Keywords: Filum terminale, Spinal cord, Split cord malformation, Tethered cord syndrome, Conus medullaris

Introduction

The term “tethered spinal cord” as defined by Hoffman¹ refers to a low lying conus medullaris (CM) with a thickened (>2 mm diameter) filum terminale (FT). The term tethered cord syndrome (TCS) is a clinical diagnosis in which there is progressive neurological worsening in the functions of the lower spinal cord due to traction on the CM.² The FT in these cases is often abnormal with adhesions to the adjacent dura. This prevents ascent of the spinal cord during embryonic life and growth spurts after birth results in traction causing anatomic and metabolic changes in the conus leading to neurological problems.³

Frequent presenting symptoms include back pain radiating to the legs, sensory deficit of the lower extremities, and loss of bladder/bowel control.⁴⁻⁶

TCS in adults is uncommon, and the diagnosis is often challenging with back pain being the only

presenting symptom.^{7,8} Most of these patients obtain significant symptomatic relief after undergoing FT sectioning.

Anatomical variations of the CM and FT are well known and are often associated with varied etiologies including spinal dysraphism, vertebral and orthopedic abnormalities such as scoliosis and limb deformities, caudal regression and anorectal malformations.⁹ A duplicated FT is well described in association with two distinct hemicords as in split cord malformation (SCM). Duplicated FT as a cause of TCS but in the absence of SCM has not been previously described to the best of our knowledge.

We report the first case in the literature of a symptomatic double FT but without any radiological evidence of SCM. We highlight the importance of this rare entity as a potential cause of failure after sectioning in TCS.

Case report

A 47-year-old male was evaluated in our clinic for long standing history of low back and leg pain extending

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down to involve the lateral aspect of his thighs and calves bilaterally. Conservative measures failed to improve his symptoms. This gradually worsened over time leading to gait unsteadiness. Neurological examination was essentially normal except for a positive Romberg test. His sphincter tone was preserved and urodynamic assessment was normal. There was no evidence of any cutaneous stigmata. There were no significant neurologic problems during his childhood.

MRI study of his lumbosacral spine showed a dorsal lipoma involving a low lying conus at the level of L3-4 disc. The lipoma was intradural and did not show any evidence of extradural extension. The normal appearing FT could be visualized extending up to the sacrum (Fig. 1). The whole spine MRI did not reveal any evidence of intramedullary syrinx or SCM. In view of persistent progression of his symptomatology it was decided to perform a sectioning of the FT and also look for any adhesions at the level of the lipoma causing tethering.

Following a standard midline posterior approach, an L4-L5 laminectomy was performed bilaterally and dura mater was exposed. The extradural findings were normal. Once the dura was opened, the cauda equina roots were gradually microscopically dissected to identify the FT. Neurophysiological monitoring was used to aid nerve root identification. The conus lipoma was identified which was free from the dura with no evidence of adhesions. The lipoma was gradually traced distally blending into a fibrous band-like structure which was anatomically compatible with and electrophysiologically confirmed as the filum. A second, similar but smaller diameter strand was identified dorsal to this one which also failed to demonstrate responses on stimulation. The structure physically resembled a filum while the nerve roots could be identified as being distinct from this band (Fig. 2). The responses were compared to the normal responses obtained by stimulating the roots.

Both these structures passed caudally and we were unable to visualize their termination with the surgical exposure. Both fila were sectioned, sending a piece of each for pathology (Figs. 3 and 4).

The pathology confirmed the diagnosis of both specimens as fibro-collagen tissues without recognizable nerve structures compatible with fila terminale.

The patient had an uneventful postoperative course. At 11-month follow-up he reported some improvement in back pain with less ataxia on walking. The patient's neurological examination was objectively normal.

Discussion

The FT is a fibrous band, composed of an intradural and extradural portion, extending from the CM to the coccygeal perisoteum, with the intradural portion measuring approximately 4 cm in newborns and 16 cm in adults.¹⁰ Several anatomical studies on human fetuses have showed that the spinal cord until the eighth gestational week is located at the inferior end of

Figure 2 Intraoperative image showing both the fila terminale. (Colour online)

* indicates the conus lipoma.

Figure 1 Preoperative sagittal T2W (A) and T1W (B) MRI showing the low lying conus and the dorsal lipoma (-). Axial T2W (C) image demonstrating the FT seen as a hypointense structure (arrow).

Figure 3 Intraoperative image showing the cut ends of the fila after sectioning. (Colour online)

the dural sac; subsequently the vertebral column gradually extends inferiorly relative to the spinal cord, between the 8th week and the 25th week. The ascending involution of the spinal cord participates in the formation of the FT.¹¹ Pinto *et al.*¹¹ in their anatomic studies indicated that the CM ended at positions ranging from T12 to L3 and in roughly 95% the inferior tip of the CM ended at or above the L2 level. Their results regarding the termination of the FT showed

Figure 4 Line drawing showing the low lying conus, the dorsal lipoma and the duplicated filum. (Colour online)

that the most frequent fusion level was S2. However, in 30% the fila fused above S2.

In 2000, Salbacak¹² performed an anatomical study on 123 medically or spontaneously aborted fetuses in order to assess the FT variations in humans. The FT and CM of 78 fetuses were observed to be normal in shape and position and a double FT was observed in 3 fetuses (2.4%) without any evidence of SCM. Rizk *et al.*¹³ reported on 2 cases of duplicated FT without SCM. In both patients, it was an incidental finding wherein the first patient was operated for a radiological diagnosis of fatty filum and the second, for an asymptomatic cutaneous hemangioma. Though the authors mention in their discussion that the diagnoses in both patients were radiologically and pathologically proven, we find that this information contradicts their description of the case reports.

Microscopic observations in early twentieth century still form the present basis of understanding the embryonic caudal neural tube development and the pathophysiology of neural tube defects. The formation and development of the distal neural tube has largely been studied in 3 phases, namely, caudal neuropore closure, secondary neurulation and retrogressive differentiation.¹⁴ During this final process of retrogressive differentiation, apoptosis plays a major role. In conjunction with the degeneration of the caudal neural tube, the secondary neural tube involutes to form a fibrous layer. This is in direct continuity with the marginal layer of the primary neural tube, most likely by differentiation from the secondary neuroectodermal cells. This fibrous tissue constitutes the FT. The mesenchyme surrounding this fibrous layer differentiates to form a meningeal structure, the primitive pia mater. Both structures insert caudally into the primitive dura mater surrounded by spinal roots and ganglia.¹⁴ The growth and elongation of this primitive FT is caused by an interstitial increase of its constituent fibers.¹⁵ Defects during secondary neurulation and/or retrogressive differentiation may play a major role in the development of anomalies such as double FT, fibrolipoma of the FT, and congenital tumors.¹⁴

Histologically the FT consists of fibrous connective tissue and vascular layers. However, other types of tissue are scattered within this band and play significant roles in many of its pathologies. Tarlov¹⁰ in 1938 described the differences in the intradural and extradural anatomy of the FT. The intradural portion consists of collagen fibers, fine reticulin and a few elastic fibers. This connective sheath becomes more prominent caudally. Other types of cells which also can be identified include astrocytes, oligodendrocytes, microglia

and ependymal cells. Nerve fiber tracts of the CM extend caudally within the intradural portion of the filum especially in its upper half. At the site of transition between the intradural and extradural portions, the filum becomes surrounded by arachnoid and dural layers whose collagen stroma contains thin-walled blood vessels. On the back of the coccyx the extradural portion of the filum fuses with the dense fibrous tissue of the periosteum.

A duplicated FT in association with two distinct hemicords is a well-known entity. According to the unified theory of embryogenesis proposed by Pang *et al.*,¹⁶ an abnormal fistula called the neurenteric fistula forms between the ectoderm and endoderm at the time of neurenteric closure. This canal develops into an endomesenchymal tract that splits the notochord and the overlying neural plate which is in the process of forming the neural tube. The final characteristics of the SCM depend on the further development of the endomesenchymal tract.

An extensive review of literature did not reveal any cases of duplicated FT in the absence of SCM as a cause of TCS. Though double filum without SCMs have been described by Ritz *et al.*¹³ on radiological imaging and Salbacak *et al.*¹² on fetal cadaveric study, our report highlights this finding as a cause of clinical symptoms. It is difficult to explain duplication of the FT based on embryology for SCMs since the development of the spinal cord and FT is distinct. Though we cannot hypothesize the exact embryological basis for this occurrence in the absence of a true SCM, we think that this might also represent a variant where the endomesenchymal tract causes distal splitting at a level rostral to the primitive pit. Though this hypothesis is an extension of the unified theory, it requires validation at an embryological level.

Conclusion

This article highlights a rare case of duplicated FT without evidence of SCM presenting clinically as TCS. It may often be unrecognized on preoperative imaging, such as in our case, and thus may be a cause of treatment failure. It supports the consideration of exploratory surgery in patients with symptomatic TCS, particularly with neurological deficits, in the absence of definite radiologic abnormalities. The presence of a double filum should be considered and looked for during the exploratory dissection of the cauda equina. The importance of sectioning both the fila cannot be overemphasized since logically this is necessary for relieving the traction on the cord. Meticulous dissection and direct visual

examination of the cauda equina, with direct intraoperative nerve stimulation may be the only way of identifying this anomaly.

Disclaimer statements

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Ethics approval None.

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