CASE REPORT

Tethered spinal cord syndrome in a 14-year-old teen with coexisting Tarlov cysts

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ABSTRACT

Introduction: Filum terminale (FT) is a thin fibrous band connecting the conus medullaris with the coccyx. Tethered spinal cord syndrome (TSCS) is caused by pressure exerted on the spinal cord by a tense inelastic filum terminale. It is subdivided into congenital/primary and acquired/secondary. Congenital TSCS is usually diagnosed at birth and early childhood. In older children, the FT (that was not tethered from birth) is elongated in time by coexisting myelomeningocele, lipomyelomeningocele, split cord malformation, tumor, trauma, surgery or Tarlov cysts (meningeal or perineural cysts around the sacral nerve roots).

Case presentation: We present the case of a 14-year-old boy who developed parieto-occipital headache, cervicalgia, dizziness, vomiting, and urinary/fecal incontinence after an injury of the lumbar and sacral spine. The imaging results (MRI and MR myelography) were consistent with lumbar-sacral subarachnoid space distension. Immediate surgery was performed: large Tarlov cysts putting pressure on the FT were identified and surgical untethering of the FT was performed. The postoperative course was smooth: symptoms remitted within 48 hours.

Conclusion: The injury of the lumbar and sacral spine in our patient caused an accumulation of cerebral fluid in pre-existing Tarlov cysts, which put pressure and tension on the FT. This caused neurologic symptoms as a first clinical manifestation at the age of 14. Immediate surgical intervention was necessary to prevent permanent damage.

Keywords: tethered spinal cord syndrome, Tarlov cysts, childhood

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INTRODUCTION

Tethered spinal cord syndrome (TSCS) as a divergent clinical entity is characterized by signs and symptoms caused by extreme tension applied on the spinal cord. TSCS may present at any age, and clinical manifestations vary according to the underlying pathological

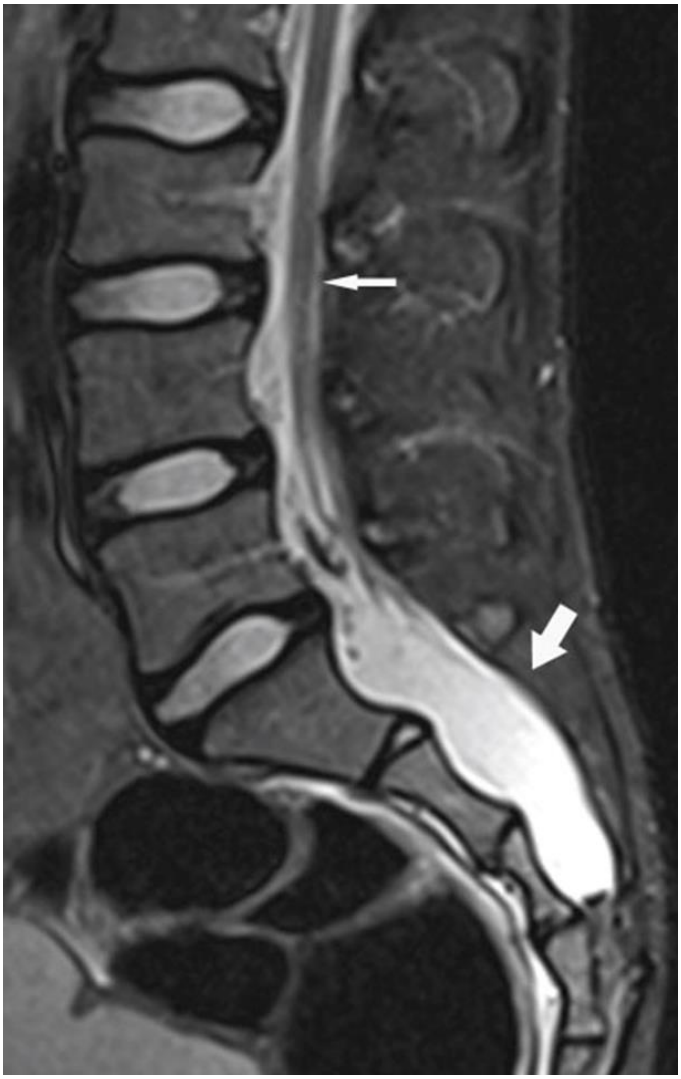

Figure 1. Sagittal STIR MR image shows meningocele (arrow). Conus medullaris (small arrow) is low-lying at the L4 level, and the filum terminale appears thickened.

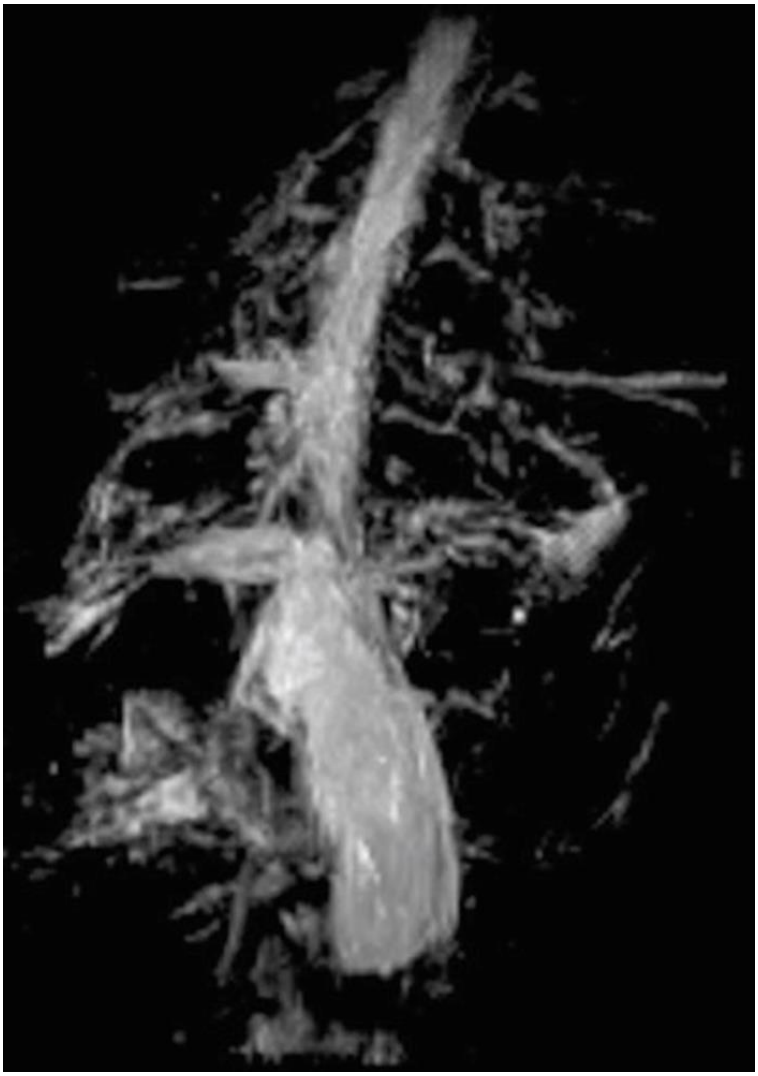

Figure 2. MR myelography shows marked dural ectasia.

condition and age, with pain, cutaneous signs, orthopedic deformities, and neurological deficits being the most common. Progressive or new onset of symptoms of TSCS mandates surgical untethering, aiming to release the tethering structure and thus the chronic tension on the cord [1].

The normal Filum Terminale (FT) is a viscoelastic band that stabilizes and protects the distal spinal cord from traction. Loss of the FT's elasticity has been described to be crucial for the development of tethered spinal cord syndrome (TSCS) [2].

The current case report presents an interesting case of TSCS due to abnormal FT and its surgical therapy.

CASE PRESENTATION

We present the case of a 14-year-old teen who is an amateur parkour athlete. He fell on

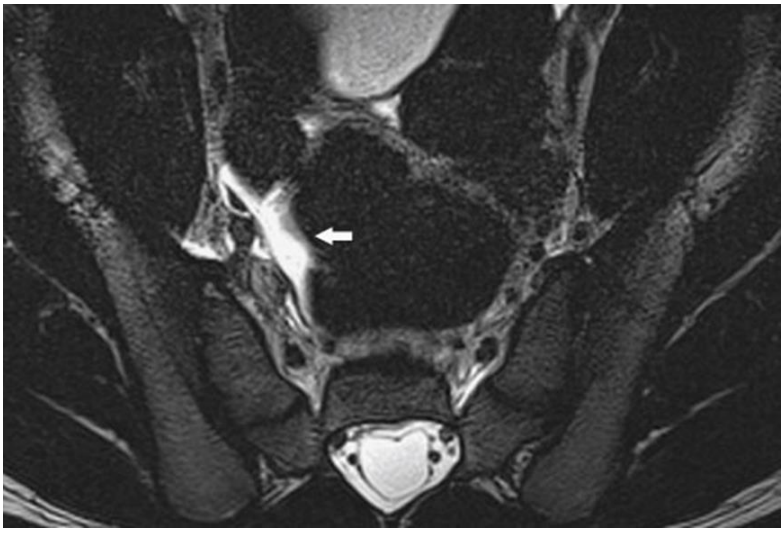

Figure 3. Axial T2-weighted MR image shows widened spinal canal with CSF leak along the cauda equina in the pelvis.

the ground while climbing a wall and hurt his sacral and coccyx area. From that moment he developed parieto - occipital headache, cervicalgia, dizziness, and vomiting. These symptoms intensified when he moved from a supine to a standing position. On the second day of hospitalization, he developed urinary and fecal incontinence.

His MRI and MR myelography (Figures 1,2,3) demonstrated a low-lying conus medullaris and dural ectasia. There was leakage of CSF tracing the cauda equina, in the pelvis. Immediate surgery was performed.

An incision was made at the level of the S5 vertebra into the subcutaneous tissue, muscle fascia, paravertebral muscles, and dura mater. The filum terminale was identified in a posterior position regarding the cauda equina and showed increased elasticity. Large Tarlov cysts were putting pressure on the FT and as a result, surgical untethering of the FT was performed (Figure 4).

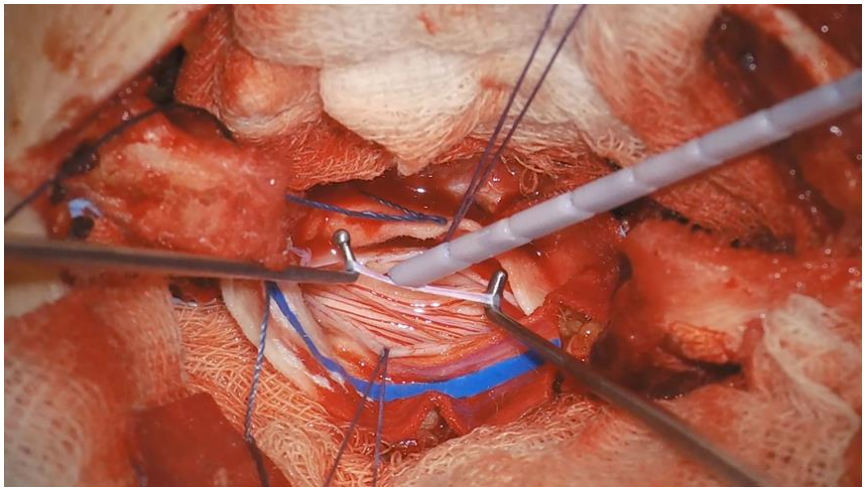

Figure 4. The filum terminale is checked via bipolar neurostimulation prior to its dissection.

The postoperative course was smooth: both urinary and fecal disturbances as well as the rest of his symptoms remitted within 48 hours. The patient is still observed for possible reappearance of neurologic symptoms, due to the pre-existing Tarlov cysts that were not surgically removed in this surgery. A follow-up imaging test after six months was recommended.

DISCUSSION

Filum terminale (FT) or spinal ligament is a fibrous band connecting the conus medullaris with the coccyx. Luschka was the first to describe two distinct parts of filum terminale: the filum terminale internum (FTI), connecting the conus medullaris and the dural sac, and the filum terminale externum (FTE), connecting the dural sac and the periosteum of the coccyx. FTE seems to house neural tissue, whereas FTI consists of fibrous connective tissue. Therefore, describing the FT as a fibrous band is probably an oversimplification that disregards its complex histology and embryology [3].

The vertebral level of the CM-FTI and the FTI-DS-FTE junction is L1 and L2 respectively. The FT has elastic properties and protects the spinal cord from mechanical stress, together with the dentate ligaments. Normal FT diameter is 1,1-1,2mm. FT diameter is > 2mm is considered abnormal. During stretch tests, an abnormal FT shows 10% elasticity, while a normal FT has 50% elasticity.

Because the FT has received little attention in literature, relatively little is known about its width, length, shape, and position relative to the spinal column. Even less has been published regarding the morphology of its transition zones. To the best of our knowledge, there is no clear description of the transition of FTI to FTE. Historically, the change in type I and type III collagen ratio has been proposed as a cause for the loss of elasticity of the FT and the tethered cord.

Apart from the anatomic interest, the clinical significance of FT in Tethered Cord Syndrome is obvious. It was first described in 1953, although its current name was given by Hoffman in 1976. TSCS is subdivided into congenital and acquired. Congenital TSCS is usually diagnosed at birth and early childhood.

In many cases, it is associated with spinal dysraphism [4]. It has also been identified in children (age range 2-14 years) with Rubinstein-Taybi Syndrome (RTS) [5]. In older children, the filum terminale (that was not tethered from birth) is elongated by coexisting myelomeningocele, lipomyelomeningocele, split cord malformation, tumor, trauma, surgery [6- 8] or Tarlov cysts.

Tarlov cysts (meningeal or perineural cysts) were originally described on autopsy findings in 1938 by the American neurosurgeon Dr. Isadore Tarlov. They occur around nerve roots anywhere in the spine, but most frequently around sacral nerve roots, at the S2 level, due to increased hydrostatic pressure in the spinal cord and due to trauma [9, 10]. They are usually filled with cerebral fluid but can also be hemorrhagic if there is any complication. Although usually asymptomatic, they can present with symptoms such as chronic pain in

the sacral or coccyx area, leg weakness, bladder and bowel dysfunction, sexual dysfunction, or any other acute symptoms in case of a rupture or hemorrhage [11].

The injury of the lumbar and sacral spine in our patient caused an accumulation of cerebral fluid in pre-existing Tarlov cysts, which put pressure and tension on the FT and caused neurologic symptoms as the first clinical manifestation at the age of 14. Immediate surgical intervention was necessary to prevent permanent damage. In case of recurrent symptoms due to an increase in the size of Tarlov cysts, a second surgery may be necessary.

To this day, there is no clear consensus about the best surgical strategy to use when needed. Various strategies are discussed such as posterior decompression, cyst wall resection, CT-guided needle aspiration with intralesional fluid injection, and shunting. Neurological deficit is less likely to remain after surgery, but pain may insist as a symptom [12].

CONCLUSION

In conclusion, a TSCS diagnosis is exceptionally difficult to be made, especially during teenage and adult life, because it is a rare entity, and it presents with a variety of symptoms. Fecal and urinary incontinence should be a red flag for the pediatrician, who is usually the first person to encounter the patient and must refer them to a specialized doctor for neurological assessment and further imaging.

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ΠΑΡΟΥΣΙΑΣΗ ΠΕΡΙΣΤΑΤΙΚΟΥ

Σύνδρομο καθήλωσης τελικού νηματίου νωτιαίου μυελού (tethered spinal cord syndrome) σε έφηβο 14 ετών με συνυπάρχουσες κύστεις Tarlov

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ΠΕΡΙΛΗΨΗ

Εισαγωγή: Το τελικό νημάτιο είναι μια λεπτή ταινία συνδετικού ιστού που συνδέει τον μυελικό κώνο με τον κόκκυγα. Το σύνδρομο καθηλωμένου τελικού νηματίου (TSCS) οφείλεται σε πίεση που ασκείται στο νωτιαίο μυελό από τεταμένο, ανελαστικό τελικό νημάτιο. Διακρίνεται σε συγγενές και επίκτητο. Το συγγενές

διαγιγνώσκεται συνήθως στη γέννηση και την πρώιμη παιδική ηλικία. Σε μεγαλύτερες ηλικίες, το τελικό νημάτιο που δεν είναι καθηλωμένο εκ γενετής, διατείνεται και επιμηκύνεται προϊόντος του χρόνου από συνυπάρχουσα μυελομηνιγγοκήλη, λιπομυελομηνιγγοκήλη, διαστηματομυελία, όγκους, τραυματισμούς, χειρουργικές επεμβάσεις στην περιοχή ή κύστεις Tarlov (μηνιγγικές ή περινευρικές κύστεις που βρίσκονται στις ρίζες των νεύρων στο ιερό οστό).

Παρουσίαση περιστατικού: Παρουσιάζεται περίπτωση έφηβου αγοριού, ηλικίας 14 ετών, που μετά από κάκωση οσφυοϊεράς μοίρας σπονδυλικής στήλης (ΟΙΜΣΣ) εμφάνισε κεφαλαλγία βρεγματοϊνιακής εντόπισης, αυχεναλγία, ζάλη, εμέτους και σφικτηριακές διαταραχές εντέρου/κύστης. Τα ευρήματα του απεικονιστικού ελέγχου (MRI και μαγνητική μυελογραφία) ήταν συμβατά με διάταση του υπαραχνοειδούς χώρου ΟΙΜΣΣ. Διενεργήθηκε άμεσα χειρουργική αποκατάσταση: αναγνωρίστηκαν ευμεγέθεις κύστεις Tarlov που πίεζαν το τελικό νημάτιο. Έγινε διατομή αυτού. Η μετεγχειρητική πορεία υπήρξε ομαλή: τα συμπτώματα υποχώρησαν πλήρως εντός 48ώρου.

Συμπεράσματα: Στον ασθενή μας, η κάκωση ΟΙΜΣΣ προκάλεσε συσσώρευση ΕΝΥ στις προϋπάρχουσες κύστεις Tarlov, που με τη σειρά τους πίεσαν και διέτειναν το τελικό νημάτιο, με αποτέλεσμα την εκδήλωση συμπτωματολογίας από το νευρικό σύστημα για πρώτη φορά στην ηλικία των 14 ετών. Η έγκαιρη χειρουργική παρέμβαση ήταν επιβεβλημένη για αποφυγή μόνιμων βλαβών.

Λέξεις ευρετηρίου: σύνδρομο καθήλωσης τελικού νηματίου νωτιαίου μυελού, κύστεις Tarlov, παιδική ηλικία

Β Γκέτση, Δ. Σαββίδου, Η. Κυροχρήστου, Γ. Μαρκογιαννάκης, Δ. Θεοδώρου, Η. Λώλος, Μ. Παπασάββα. Σύνδρομο καθήλωσης τελικού νηματίου νωτιαίου μυελού (tethered spinal cord syndrome) σε έφηβο 14 ετών με συνυπάρχουσες κύστεις Tarlov. Επιστημονικά Χρονικά 2023; 28(1): 122-128

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