

EC CLINICAL AND EXPERIMENTAL ANATOMY Mini Case Study

Tethered Cord Syndrome Revealing a Duplicated Filum Terminal

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Abstract

Tethered cord syndrome (TCS) is a clinical diagnosis of progressive neurologic aggravation due to the traction on the conus medularis of a low spinal cord. Untethering surgery is effective for most TCS; however, when anatomic variations of spinal cord and filum terminal (FT) exist, regular untethering may lead to a failed outcome. We report a rare case of a duplicated FT in adult with no split cord malformation (SCM).

Keywords: Duplicated Filum Terminal; Non-Split Cord Malformation; Tethered Cord Syndrome

Case Report

A 50 year old woman, consults in our hospital for chronic back pain dating from 2 years ago. She reports an aggravation of the symptomatology during two past months with paroxysmal leg pain.

The neurological examination was normal, including preservation of motor and sensory capacity and absence of sphincter disorder. No cutaneous stigmata were found on her back. No history of neurologic problems during childhood, and no family history of dysraphism.

Thoracic and lumbosacral magnetic resonance imaging (MRI) revealed a low position of the conus medullaris which is attached at L4 level (Figure 1) due to a double and thick FT, without lipoma of FT or SCM. In addition, there are vertebral abnormalities: defect of fusion of the spinal laminas which are sagitalised causing an enlargement of the spinal canal (Figure 2). There are no other malformations such, syringomyelic cavity, myelomeningocele, or other types of dysraphism. We concluded that it is a thick double FT without SCM causing TCS.

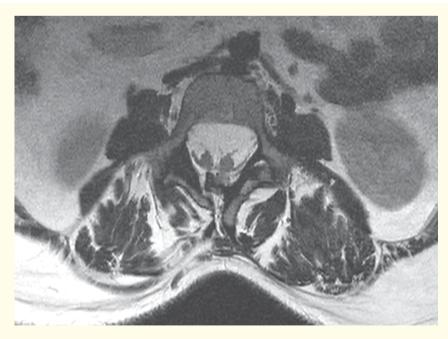


Figure 1: Sagittal section in weighted sequence T2: low insertion of the conus medullaris attached at the L4 level.



Figure 2: Axial section in weighted sequence T2: double FT exceeding 2 mm thickness associated to vertebral malformations such a defect of fusion of the posterior arch.

Discussion

The FT is a small thin filament of connective tissue and ependymal cells that extends inferiorly from the apex of the conus medularis to the sacrum. The FT is continuous with the pia mater and is described as having two sections. The intradural portion is called the FT internum connecting the CM and the distal termination of the dural sac, and the extradural portion is the FT externum, connecting the distal dural sac and the first coccygeal vertebra [1]. The level of the medullary cone is variable, usually located opposite the L1-L2 disc. A situation opposite or above the L2-L3 disc is considered normal, regardless of age [2].

The formation and development of the distal neural tube has largely been studied in 3 phases: caudal neuropore closure, secondary neurulation and retrogressive differentiation. 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 which is the FT. This is in direct continuity with the marginal layer of the primary neural tube, most likely by differentiation from the secondary neurectodermal cells. The mesenchyme surrounding this fibrous layer differentiates to form a meningeal structure, the primitive pia mater. Both structures inserts caudally into the primitive dura mater surrounded be spinal roots and ganglia. The growth and elongation of this primitive FT is caused by an interstitial increase of its constituent fibers. 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. Abnormalities of the FT are classified in the simple closed spinal dysgraphia [3].

A duplicated FT in association with SCM is a well-known entity. According to the unified theory of embryogenesis proposed by Pang., *et al.* 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 [4].

Anomalies of FT are often associated with various malformations, such as SCM, spina bifida, scoliosis, vertebral dysplasia, low-lying conus, thick filum, lipoma, hydromyelia, myelomeningoceles, dermoid cyst, and dermal sinus [2].

A FT malformation can cause tension transmitted to the spinal cord especially to the conus medullaris. Clinically, it manifests itself as a spinal cord syndrome, which causes back pain and neurological deficits in children and requires surgical treatment. Although some patients may remain completely asymptomatic or manifest symptoms until they became adult [1].

In MRI, the double FT appears as two separate films following the conus medullaris. The double FT can be short and hypertrophic with a thickness more than 2 mm on an axial section passing through the L5-S1 disc. Generally, the position of the FT appears in a position lower than L2. The filum and the conus medullaris are moved to the posterior part of the spinal canal and the medullary cone appears in the lower position. MRI can also be used to look for other FT abnormalities, including filum lipoma, neural tube defects and spinal cord malformations [2].

Duplicated FT is uncommon and has been reported in sporadic cases in fetuses and young children. Though double filum without SCMs have been described by Ritz., *et al.* on radiological imaging and Starnoni., *et al.* and indicated that duplicated FT may be a potential reason for the failure of an untethering surgery [3,5].

Conclusion

Duplicated FT with non SCM is a rare cause of TCS in adult. It is necessary to look for duplicated FT in imaging and during surgery in patient having a TCS. It's may be a cause of failed untethering surgery.

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