

Tethered cord – a case report and literature review

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Abstract

Tethered cord is a congenital syndrome characterized by the spinal cord under tension, being extended and attached to caudal structures, usually being associated with dysraphic lesions. Tethered cord is an anomaly of the caudal cell mass and a secondary neurulation process with a deficit of regression of the caudal eminence. Advancements in ultrasonography allow the condition to be detected from the second or the third trimester, creating the necessary premises for a higher rate of successful treatment. Pain and neurological disfunctions, such as muscular weakness and sphincteric dysfunctions, represent the main symptoms of the condition. Left untreated, tethered cord will lead to progressive neurological decline and irreversible damage to the nerves. The main outcomes of the surgery are the reduction or stoppage of neurological function decline and the improvement of sphincteric control which represent a significant improvement in the quality of life of the patients. We present the case of a newborn with tethered cord diagnosed postnatally, with the aim to emphasize the importance of ultrasonographic use in the prenatal diagnosis of congenital anomalies.

Keywords: tethered cord, neurologic disfunction, spinal development

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Rezumat

Coloana fixată reprezintă un sindrom congenital caracterizat de tensiunea prezentă la nivelul coloanei vertebrale, care este în extensie, având conul medular fixat de structurile caudale. Coloana fixată este o anomalie a procesului de neurulație secundară și a deficitului de resorbție a eminentei caudale. Progresele ultrasonografiei permit detecția prenatală a bolii, din al doilea sau al treilea trimestru de sarcină, ajutând la stabilirea unei conduite terapeutice cu șanse însemnate de succes. Durerea și deficiențele neurologice, cum ar fi slăbiciunea musculară și deficiențele sfincteriene, reprezintă principalele simptome ale bolii. Netratată, coloana fixată evoluează către declin neurologic, adesea cu afectare ireversibilă a structurilor neurologice. Principalele efecte oferite de intervenția chirurgicală de corecție sunt reprezentate de diminuarea durerii, îmbunătățirea controlului sfincterial și prevenirea leziunilor ireversibile, ducând la îmbunătățiri semnificative a calității vieții pacienților. Prezentăm cazul unui nou-născut cu coloană fixată diagnosticat postnatal, cu scopul de a sublinia importanța utilizării ultrasonografice în diagnosticul prenatal al anomaliilor congenitale.

Cuvinte-cheie: coloană fixată, disfuncții neurologice, dezvoltarea coloanei vertebrale

Introduction

Tethered cord is a congenital syndrome in which the spinal cord is extended and attached to structures situated caudally which leads to the high tension in the cord, resulting in a plethora of signs and symptoms. It is usually associated with spinal dysraphic lesions, whether closed or open, and with other orthopedic deformities and motor and sensory neuron disfunctions⁽¹⁻³⁾. Tethered cord is an anomaly of the caudal cell mass and secondary neurulation process where the caudal eminence does not regress as it normally does, leaving a caudal cell mass (retained medullary cord) which stretches from the conus to the dural cul-de-sac⁽²⁾.

It is rarely associated with skin defects, as the primary neurulation and ectoderm development are usually finalized by the time the caudal cell mass starts its process of development. It can be accompanied by lipomeningomyelocoele, myelomeningocoele, myelocystocoele, meningocele, split cord malformation, anorectal malformation, neuro-enteric cyst, and fatty filum terminale⁽³⁾. Spinal development is influenced by the neurulation process

which occurs between 18 and 28 days of gestational life. Beside the neurulation process, the caudal part forms via a process called canalization which occurs from gestational days 28 to 48. In the last five days, the terminal spinal cord starts a process of regression which results in the formation of filum terminale⁽¹⁾. During the late fetal life, the spinal cord (Figure 1) is located between L3 and S5 vertebrae, while at term it is usually located between L1 and L3. Finally, the *conus medullaris* reaches its adult position at 2 years of life which is normally at L2⁽³⁾.

Diagnosis

Prenatal ultrasonography allows the practitioner to diagnose structural anomalies of the fetal spine from the second or the third trimester, providing valuable time and information in order to organize the follow-up plan for the neonate⁽⁴⁾. As more and more pregnant women gain access to early-trimester screenings, the rate at which the spinal defects are diagnosed has increased, and less patients are diagnosed when they reach adulthood^(1,4).

After birth, most of the times, the simple examination and observation of a neonate or infant are enough to detect or at least to suspect a problem, while imagistic tools are required to properly diagnose the condition and to establish the medical conduct and treatment^(5,6).

Even though MRI is the gold standard in detecting any anomalies of the spinal cord, advancements in the ultrasonography have allowed it to become a reliable tool in detecting and diagnosing the disease early enough to prevent the irreversible damage to the spine^(6,7). Khoury and Sepulveda reported a higher incidence of tethered cord or other spinal anomalies in pregnant women with folic acid deficiency, gestational diabetes, obesity or in long-term users of antiseizure drugs like carbamazepine or valproate^(2,4).

Case presentation

We present the case of a male, term neonate, born at 38 weeks of gestational age. The pregnancy was affirmatively uneventful, and the baby was born by caesarean section. At the clinical examination, there was identified a stigmata in the lumbar region – red macula, 1-2 cm in diameter, situated over the spine (Figure 2 a and b).

A spinal ultrasound was performed immediately and identified a syrinx situated in the thoraco-lumbar region (T9-L1) – Figure 3 a-c. There was also observed a diminished mobility of the spinal cord with respiration and heart pulsations. An MRI was performed on the third day of life and the syrinx has been identified with the same characteristics as the ultrasound (Figures 4 and 5).

The baby was followed by the pediatric neurologist (clinical examination and ultrasound evaluation) and neurosurgeon until 3 months of age, when the decision was taken to operate the tethered cord.

After the procedure, the child was followed-up until the age of 3 years old. The control MRI showed a partial resolution of the lesion (Figure 5). The motor milestones were in the normal range for the age (head control at 2 months old, independent walking at 12 months old, normal fine motor and cognitive-language development for the age). The only problem observed was a delayed control of the bladder, probably due to a neurogenic bladder, that resolved by the age of 3 years old.

Discussion

If left untreated, tethered cord will lead to progressive neurological decline. The patient may experience trouble walking or running as a result of progressive motor dysfunction, sensory deficits, as well as problems with bladder control or, less usual, bowel control. Pain is an occasionally expressed symptom, most usually located in the lower back or perineum. The most frequent signs and symptoms found in teenagers and adults are scoliosis, pain, muscular weakness and sphincter dysfunctions⁽¹⁾.

The major goal of the treatment is preserving the neurological function and prevent further irreversible damage that may occur if the treatment is delayed. It is recommended to perform the surgery as early as possible during the lifetime. The main outcomes of the

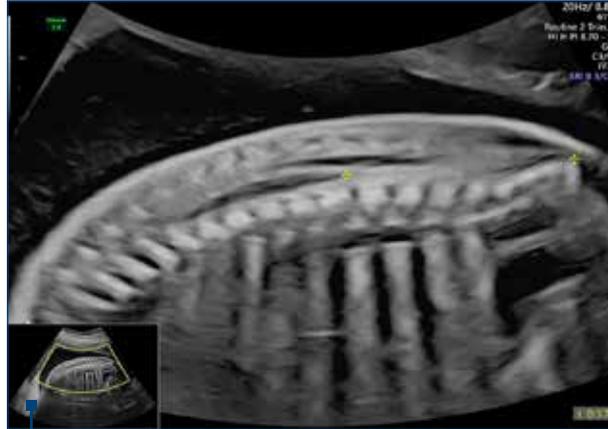


Figure 1. Two-dimensional ultrasound measurement of the conus medullaris. The identification in the midsagittal plane of the triangular conus medullaris (personal collection of dr. R. Bohilțea)



Figure 2 a) and b). Infant with tethered cord – physical examination (personal collection of dr. A.I. Toma)

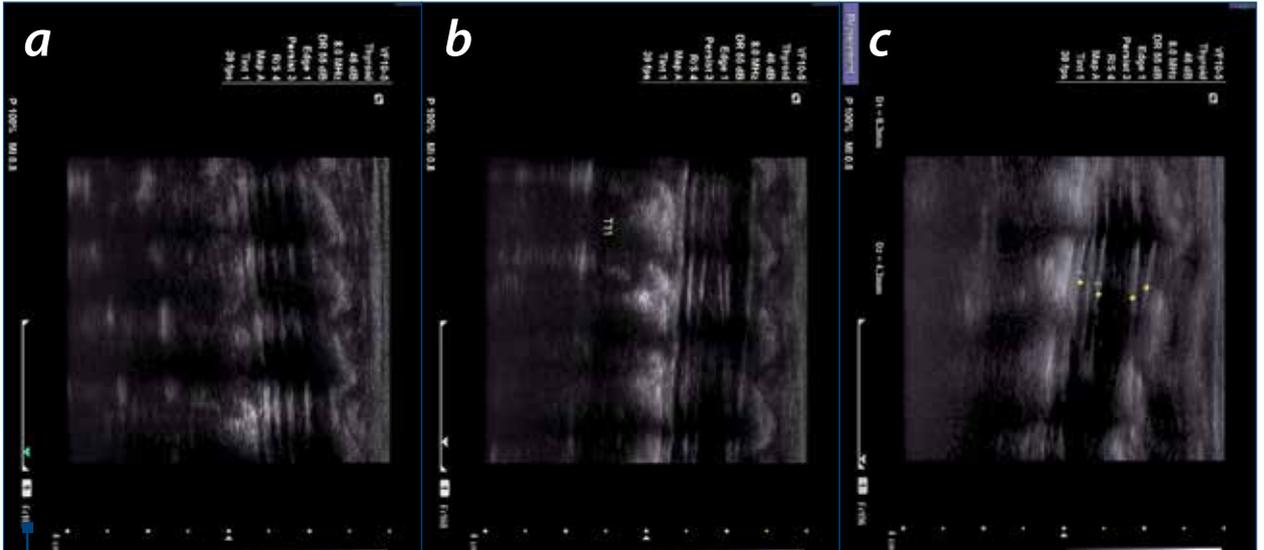


Figure 3. a), b) and c). Successive sections of the spinal cord (personal collection of dr. A.I. Toma)

surgery are reduction of pain (improvement reported by 70% of patients), stoppage of neurological function decline (achieved in around 90% of patients), and improvement in sphincteric control⁽¹⁾. Retethering of the cord is a frequent complication following surgery. Lew et al. reported a retethering rate of 5-50%, depending

of the procedure followed. Repeated surgeries for untethering can be performed at similar success rates as the initial surgery⁽¹⁾.

Tethered cord is a severe condition that could lead to cord rupture and subsequent paralysis if not detected in time⁽⁸⁾.



Figure 4. MRI section of tethered cord (personal collection of dr. A.I. Toma)



Figure 5. Sagittal MRI section of tethered cord (personal collection of dr. A.I. Toma)

Spinal ultrasound is indicated for the examination of the spinal cord in the neonates and small infants (until 3-6 months of age)^(6,7,9). The method takes advantage of the fact that the posterior aspect of the spine is incompletely ossified, allowing the visualization of the spinal structure^(9,10). Spinal ultrasound is indicated in the cases of paraspinous stigmata^(8,10,11). The diagnostic value of the spinal ultrasound has been found to be comparable with the MRI in the case of occult spinal dysraphisms.

In the present case, the physician identified immediately the stigmata and promptly indicated the ultrasound examination. The spinal ultrasound identified the anomaly which was confirmed by the MRI. The early identification of the pathology allowed the early neurosurgery consult and the close monitoring of the patient. The operation was decided and performed before the occurrence of complications.

Conclusions

Tethered cord is a condition with significant influence on the quality of life of patients, as a result of the tension in the spinal cord which generates sensitive and motor dysfunctions. Tethered cord is usually associated with dysraphism, as well as other congenital syndromes. Surgery represents the main treatment which leads to a stabilization of the neurological functions, reduction of pain and improvement of the sphincteric control.

We consider that the early use of spinal ultrasound in all the cases in which an occult dysraphism is suspected could identify eventual anomalies in time and offer a good window of opportunity for the follow-up of the patient and the timely decision about the surgical care, thus assuring the best prognosis of the patient. ■

Conflicts of interests: The authors declare no conflict of interests.

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