Scientific Letter / *Bilimsel Mektup*

DOI: 10.5152/tftrd.2014.71597

Turk J Phys Med Rehab 2014;60:182-3
Türk Fiz Tıp Rehab Derg 2014;60:182-3

Epidermoid Tumor with Tethered Cord

Epidermoid Tümör ve Tethered Kord Birlikteliği

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Dear Editor,

Epidermoid tumors constitute less than 1% of intraspinal tumors. Generally, spinal epidermoid tumors have an intradural extramedullar location and often tend to be located in the lumbosacral region. It is often related to spinal dysraphism or occasionally occurs as a result of repeated lumbar punctures. The appearance of intradural intramedullar epidermoid tumors is extremely rare (1).

Tethered cord syndrome is a disease table characterized by progressive neurological deficits, which occurs as a result of spinal cord traction (2).

A case of tethered cord syndrome in the conus medullaris together with an intradural intramedullar tumor seen in an 8-year-old female patient is presented and discussed here.

An 8-year-old female patient presented at the outpatient clinic with complaints of increasing weakness in the right lower extremity on walking, shortness, and atrophy. The patient had no complaints of pain or numbness. No symptoms of urinary or fecal incontinence or retention were evident. There was no history of trauma, fever, previous surgery, or lumbar puncture. In the examinations made on the patient, the cranial nerve examination was normal, and both upper and lower extremity joint range of movement was normal. In the right lower extremity, there was 3 cm shortness and 1 cm atrophy in the right calf. Motor strength was 5/5 in the left lower extremity muscles and the right lower extremity hip and knee muscles. In the right ankle, strength was 4/5 with dorsiflexion and plantar flexion. On sensory examination, there was hypoesthesia on the right L5-S1. Achilles reflex could not be taken. No pathological finding was evident in routine blood tests. On the lumbar MRI, a mass was observed showing hypointensity on T1WI and hypermix signal intensity on T2WI, heterogeneous contrast on post-contrast T1WI, and limited diffusion on diffusion-weighted imaging (DWI); a mass 4x1.5 cm in size with lobular contours in the conus medullaris located intramedullary and filum terminale was thick and tense (Figure 1). From these findings, a diagnosis was made of epidermoid tumor and tethered cord in the conus medullaris. The recommended surgery resulting from the neurosurgery consultation was refused by the parents.

Epidermoid tumors are often related to developmental anomalies, such as spina bifida, dermal sinus, meningomyelocele, and diastematomyelia. Desquamated cells containing keratohyalin, which is encased within a capsule of well-differentiated stratified squamous epithelium, are formed (1). There are two forms: congenital and developmental. In congenital epidermoid tumors, a benign tumor forms as the result of inclusion of ectodermal tissue during normal tube formation at between 3-5 weeks of embryological development (3). The developmental form often develops associated with the movement of skin tissue during a lumbar puncture (1).

In tethered cord syndrome, traction of the spinal cord, which will form ischemia, causes physiopathological events, which then cause progressive neurological deficits (2). Diagnosis is often made in childhood. In asymptomatic cases, symptoms may start in later years with the carrying of heavy loads, excessive exercise, lithotomy position, childbirth, or spinal trauma. It is known that early diagnosis and treatment of tethered cord syndrome in childhood are important in the prevention of progressive neurological losses (2).

MRI plays an important role in the diagnosis of epidermoid tumors, which are generally seen as hypointense on T1-weighted and hyperintense on T2-weighted MRI. Cerebrospinal

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Received/Geliş Tarihi: March/Mart 2013 Accepted/Kabul Tarihi: November/Kasım 2013

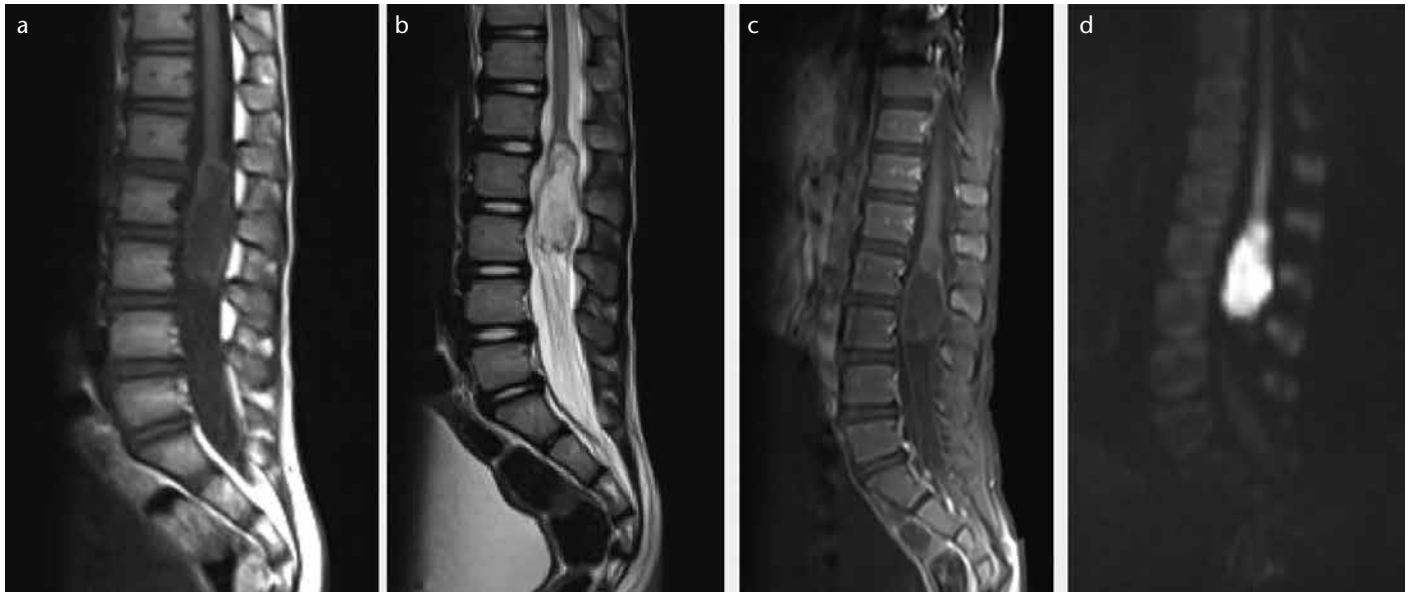

Figure 1. MRI of lumbar spine shows hypointensity on T1WI (a) and hyper-mix signal intensity on T2WI (b), heterogeneous contrast on post-contrast T1WI (c), and limited diffusion on DWI (d). Mass is 4 x1.5 cm in size with lobular contours in the conus medullaris, located intramedullary, and the filum terminale is thick and tense.

fluid (CSF) is at the same intensity. However, dermoid tumors are generally seen as hyperintense on T1, due to the presence of fatty secretions from sebaceous glands and cholesterol, and are hypointense on T2-weighted images (1). Epidermoid tumors do not show contrast. Diffusion-weighted MRI has been proven to be a useful technique for the differentiation of epidermoid tumors from arachnoid cysts, and when epidermoid tumor is considered, MR should be added to the protocol (4).

Epidermoid tumors generally become symptomatic in the 3rd or 4th decade (5). Chemical arachnoiditis may develop because of rupture from epidermoids. Particularly in the presence of dermal sinus, meningitis or spinal abscess may develop. Therefore, delayed diagnosis increases the risk of complications. Surgical treatment is curative (1). To the best of our knowledge, there are a limited number of pediatric cases of epidermoid tumors in the literature.

As the case presented here had no known history of surgery or trauma, the intradural intramedullary location of the lesion was considered to be a congenital epidermoid tumor.

In conclusion, when atrophy in the foot, in particular, or deformity is seen in the lower extremity of a child, spinal cord pathology should be kept in mind. Early diagnosis and treatment are important for the prevention of myelopathy, meningitis, spinal abcess, and arachnoiditis.

Informed Consent: Written informed consent was obtained from patient's parents who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - A.B., N.B.; Design - A.B., A.T.; Supervision - N.B.; Funding - A.B.; Materials - A.B., A.T.; Data Collection and/or Processing - N.B., E.K.; Analysis and/or Interpretation - N.B., E.K.; Literature Review - A.B., A.T.; Writing - A.B.; Critical Review - A.T., N.B.; Other - A.B., A.T., N.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

Hasta Onamı: Yazılı hasta onamı bu olguya katılan hastanın ailesinden alınmıştır.

Hakem değerlendirmesi: Dış bağımsız.

Yazar Katkıları: Fikir - A.B., N.B.; Tasarım - A.B, A.T.; Denetleme - N.B.; Kaynaklar - A.B; Malzemeler - A.B., A.T.; Veri toplanması ve/veya işlemesi - N.B., E.K.; Analiz ve/veya yorum - N.B., E.K.; Literatür taraması - A.B., A.T.; Yazıyı yazan - A.B.; Eleştirel İnceleme - A.T., N.B.; Diğer - A.B., A.T., N.B.

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Finansal Destek: Yazarlar bu çalışma için finansal destek almadıklarını beyan etmişlerdir.

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