

THE ASSOCIATION OF TETHERED CORD, SYRINGOMYELIA, DIASTOMETAMYELIA, SPINAL EPIDERMOID, SPINAL LIPOMA AND DERMAL SINUS TRACT IN A CHILD

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We report on a 5-year-old boy presenting with tethered cord, diastometamyelia, spinal dysraphism, terminal lipoma, spinal epidermoid, and dermal sinus tract with CT, conventional MRI, and diffusion-weighted MRI findings. To the best of our knowledge, our case has the property to be the first case in the literature showing the association of these pathologies all together.

Keyword: Spinal cord, developmental defect.

Tethered cord is a clinical syndrome which frequently is associated with a short and thick filum terminale. Orthopedic deformities are related to the lower settlement of the conus medullaris. While myelomeningocele, lipomyelomeningocele, and diastometamyelia may cause tense or short filum terminale, dermal sinus tract, tumor, hemangioma or scar tissue may cause tethered cord syndrome (1-3). In this case, diastometamyelia, spinal dysraphism, terminal lipoma, spinal epidermoid cyst and dermal sinus tract were all observed concomitant with tethered cord and we aimed to present the computerized tomography (CT), conventional magnetic resonance imaging (MRI) and diffusion-weighted MRI findings.

Case presentation

A 5-year-old boy was referred to our hospital with the complaints of complete abasia and incontinence. On physical examination, motor strength in the upper extremity was complete, deep tendon reflex in the lower extremity was decreased, and he could not walk without any but-tress. The patient had a sinus tract at sacral level. The anamnesis taken from the parents revealed that the patient was the ninth child of the 31-year-old mother. His two siblings died just after they were born. No consanguineous marriage was reported in the family. A spinal MRI and a lumbosacral CT examination were carried out. The patient was examined with 1.5 Tesla (Siemens symphony, Erlangen, Germany) MRI T1-weighted turbo spin echo (TSE) sagittal, T2-weighted TSE sagittal-

axial, T2-weighted coronal images and sagittal and axial diffusion-weighted images were obtained.

Spinal MRI examination showed spinal cord ending at S1 level. At the level of L5-S1 vertebrae a 22×15 mm intracanal cystic lesion showing restricted diffusion consistent with an epidermoid cyst was observed (Fig. 1A-C). On sagittal images syringohydromyelia all along the spinal cord (Fig. 1A, B), and on coronal and axial images diastometamyelia (Fig. 1D, E) at lower thoracic and lumbar levels were demonstrated. Besides, at the level of S1-2 vertebrae, a 15mm sized intradural lipoma and an associated dorsal dermal sinus tract were visualised (Fig. 1A, B, F, G). CT examination demonstrated a spina bifida at the posterior elements of S1 vertebra (Fig. 1H).

Discussion

In 1953, Garceau first asserted that the short filum terminale causes spinal cord traction. In 1976, Hoffman et al. used the term tethered spinal cord (1-3). In 1981, Yamada et al defined the term tethered cord syndrome (4). In the 8th week of the gestation, spinal cord reaches the bottom length of the spinal channel. During the remaining time of the gestation, bone components develop faster than the spinal cord. Therefore the cord reaches T12-L2 level which is the normal position of conus medullaris (5). If an obstruction or a mechanical pull occurs due to any intradermal anomaly (e.g. terminal lipoma, diastematomyelia, dermal sinus, etc.), this rise is not completed (4).

Surgical intervention is advocated even for the asymptomatic cases. Although the time and choice for the treatment of tethered cord remain controversial, treatments applied before 6 months reduce the probable deficits to a minimal level compared to treatments applied after 4 years. The therapy to be preferred is early surgical procedure. Aim of the therapy is to release the spinal cord by pulling it apart from its abnormal links. If no intervention is applied, irrecoverable neurological deficits will develop in 90% of the patients (5-8).

Female/male ratio in tethered cord syndrome is 2/1. While it is observed more commonly in females, no data is found in the literature for the frequency of this rare syndrome. The most frequently encountered symptom is the motor deficit in the lower extremities and it is seen in 75% of the patients. Lower extremity pain is seen in 40% of the pediatric patients and urological problems are seen in 35% of all age groups (5, 9-12). Our patient had the complaint of abasia. Neurological deficits become more apparent as the child grows up especially between the ages of 5 and 15 (13).

The most frequently reported causes of tethered cord are split spinal cord syndrome of 17.0%-38.2% and lipomyelomeningocele with 19.1%-94.1%, short and thick phylum terminale with 5.8%-55.8% and the adhesences which appear subsequent to the myelomeningocele operation with 5.4%-17.9%. A rare cause is the spinal epidermoid cysts. A few causes are asserted for the formation of spinal epidermoid cysts. Lumbar puncture, trauma and congenital causes are determined in the etiology (13-17). Epidermoid cysts result from the association of anarchic movement of ectodermal cells during the early fetal period and defect in the closure of the neural tube. In our literature review, we

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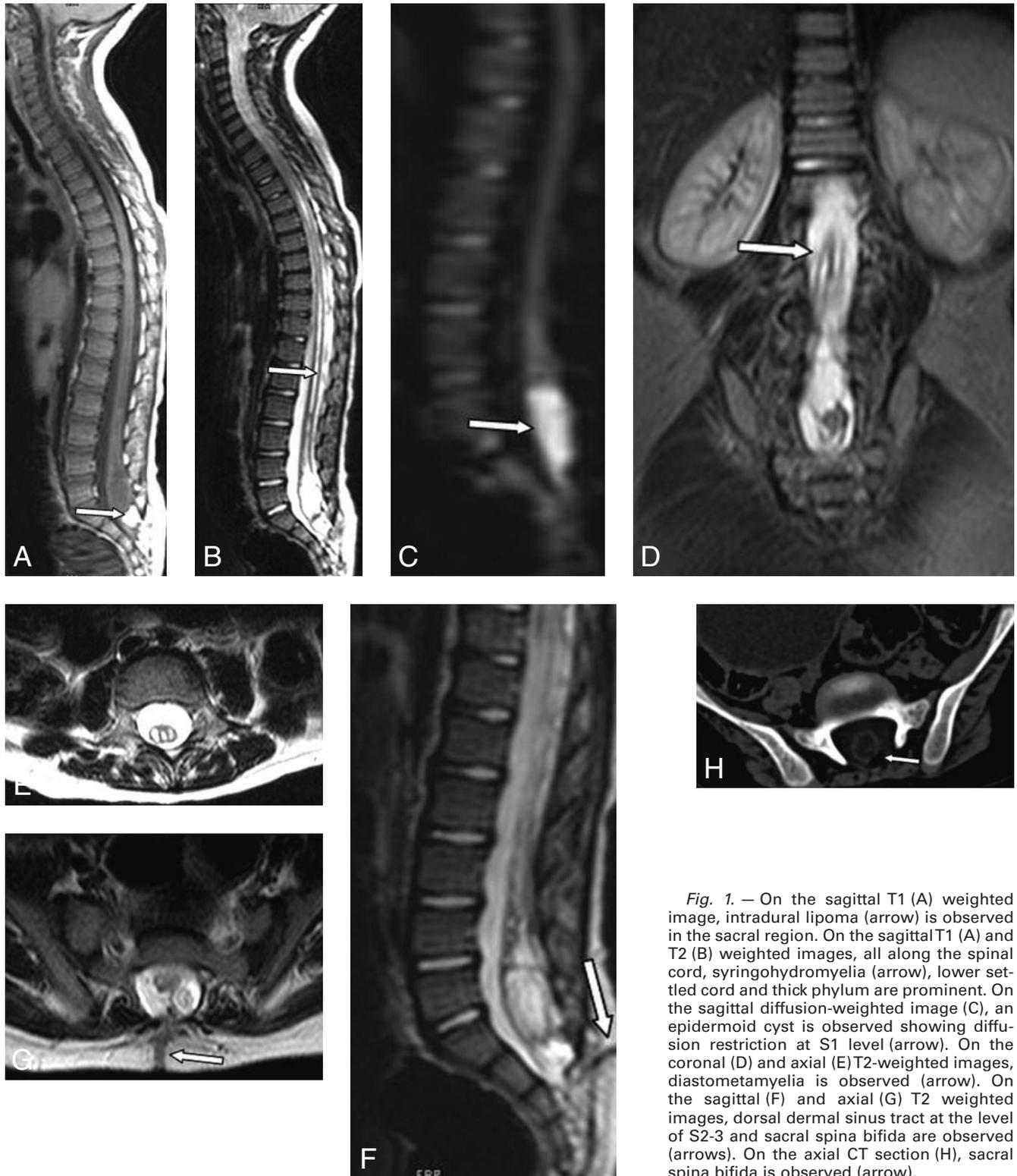


Fig. 1. – On the sagittal T1 (A) weighted image, intradural lipoma (arrow) is observed in the sacral region. On the sagittal T1 (A) and T2 (B) weighted images, all along the spinal cord, syringohydromyelia (arrow), lower settled cord and thick phylum are prominent. On the sagittal diffusion-weighted image (C), an epidermoid cyst is observed showing diffusion restriction at S1 level (arrow). On the coronal (D) and axial (E) T2-weighted images, diastematomyelia is observed (arrow). On the sagittal (F) and axial (G) T2 weighted images, dorsal dermal sinus tract at the level of S2-3 and sacral spina bifida are observed (arrows). On the axial CT section (H), sacral spina bifida is observed (arrow).

came across a few spinal epidermoid cyst cases which accompany the tethered cord (17). Our case associates epidermoid cyst, diastematomyelia, syringohydromyelia, dermal sinus tract as well as terminal lipoma along with tethered cord. Referring to the literature, among

30% of the patients who had spinal lipoma and tethered cord, at least one of the supplementary pathologies (dermoid tumor, hydromyelia, thick phylum, and diastematomyelia) was observed.

Since the epidermoid cysts associated with tethered cord are fre-

quently isointense on conventional MRI, they can be missed easily. In these cases, additional diffusion-MRI should be done as epidermoid cysts characteristically exhibit restricted diffusion (17). In our case, the epidermoid cyst which cannot be clearly distinguished from the CSF on con-

ventional MRI sequences flared up on diffusion-weighted images and the fact the epidermoid cyst was detected by means of diffusion-MRI suggested that the spinal diffusion-MRI would be beneficial as a routine examination method in the tethered cord cases.

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