CASE REPORT

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Tethered Cord Syndrome in Patient with Lipomyelomeningocele: A Case Study

I Gusti Ayu Sri Mahendra Dewi [1*], Ni Putu Sriwidyani [1], Desak Made Cittarasmi Saraswati Seputra [2]

[1] Department of Anatomical Pathology, Faculty of Medicine, Udayana University – Sanglah Hospital, Denpasar, Indonesia
[2] Internship Program, Bhakti Rahayu Hospital, Tabanan, Bali, Indonesia

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**Corresponding author:*
I Gusti Ayu Sri Mahendra Dewi
Department of Anatomical Pathology, Faculty of Medicine, Udayana University – Sanglah Hospital, Denpasar, Indonesia
mahendradewi@rocketmail.com

ABSTRACT

Introduction: Tethered Cord Syndrome (TCS) is a condition that occurs due to spinal cord traction caused by the attachment of the spinal cord to the surrounding tissues. The incidence of TCS is quite rare, approximately 0.25 per 1,000 live births.

Case Presentation: We reported a 4-year-old female patient with a lump on her sacral region since birth which enlarged over time. The patient experienced intermittent fever for 1 month before hospital admission and constipation since infancy. Physical examination revealed a lump with a diameter of 6 cm on her sacral region with no neurological abnormality. Ultrasonography examination showed an anechoic lesion with multiple linear structures on supragluteal sinistra with a size of 2.2 x 2.5 x 2.1 cm. MSCT findings showed spina bifida at L5 until S2 level with features of lipomyelomeningocele and tethered cord started from L2 level. Bilateral hydronephrosis, hydroureter, and distended bladder (pine tree appearance) with the possibility of the neurogenic bladder were also found on MSCT examination. The patient underwent surgery to excise the lump and to release the tethered cord. Microscopic findings from excised mass showed a group of neuronal and glial cells, the syncytial sheet pattern of meningothelial cells, and foci of calcification which supported lipomyelomeningocele diagnosis.

Conclusions: Intensive follow-up care and observation must be carried out in children with spinal dysraphism because it may be associated with TCS which may be asymptomatic at an early stage.

INTRODUCTION

Spinal dysraphism is one of the most common birth defects worldwide, caused by abnormalities of the neural tube. Compared to other regions, Asia has the highest incidence of spinal dysraphism which occurs in 81.37 per 100,000 live births [1]. In Indonesia, currently, there is no prevalence data regarding spinal dysraphism cases.

Tethered cord syndrome (TCS) is a neurological disorder associated with spinal dysraphism. TCS is caused by the attachment of the end of the spinal cord to the tissue that surrounds it [2]. This condition may limit the movement of the spinal cord and cause an abnormal stretching of the conus medularis. Low lying conus medularis may also occur due to disproportionate growth between the tethered spinal cord and the vertebrae [3]. The incidence of TCS is quite rare, approximately 0.25 per 1,000 live births [4]. TCS can be found at any range of age, but it is more common in pediatric patients [3].

Signs and symptoms of patients with TCS may vary. The most common clinical manifestation is occult spinal dysraphism that presents as a dimple, fatty lump, hemangioma, or tuft of hair at the back [2,5]. In the pediatric and asymptomatic case, TCS may be suspected if there is an occult spinal dysraphism. Other signs and symptoms include motor weakness, numbness, discrepancy of lower extremities, foot deformity (club feet, pes cavus), scoliosis, back and leg pain, urinary incontinence, recurrent urinary infection, and hydronephrosis due to the neurogenic bladder [2,3,5] although, in many pediatric patients, symptoms often only manifest as irritability [6].

Early diagnosis and treatment are important in TCS cases as untreated patients will experience clinical deterioration over time. In asymptomatic patients, untethering surgery may prevent future deterioration. Meanwhile, in symptomatic patients, it helps to prevent worsening of symptoms and return to optimal function [3,5]. We reported one case of TCS presented with

urinary tract infection and chronic constipation at Sanglah General Hospital Denpasar.

CASE PRESENTATION

A 4-year-old female patient was referred from Siloam Hospital Bali to Sanglah General Hospital with a complaint of a lump on her lower back since birth which enlarged over time. The patient also experienced intermittent fever for 1 month before the initial hospital presentation and constipation that worsened in the past 2 months. There were no growth and developmental delays observed in the patient.

Based on the medical history, although the patient's lump was present since birth, her parents refused to do further examination at that time because there were no other complaints. They were advised to do a regular check-up to monitor their daughter's condition as progressive neurological deterioration might occur. However, because of the parents' lack of understanding and perception that the patient's complaint was normal at her age, the patient was taken to the hospital after experiencing severe complaints.

On physical examination, a round, soft, mobile, and non-tender lump was found on the sacral region. The lump was 6 cm in diameter. Neurological examinations were within normal limits. Based on laboratory examination, leukocytosis (15,17x103/µl) was found. Urinary tract infection was present based on laboratory examination.

Ultrasonography was performed as occult spinal dysraphism was suspected. Ultrasonography evidenced a spina bifida at the sacrum level with lipomyelomeningocele features and tethered cord (**Figure 1**). As tethered cord was found, thoracolumbar Multi Sliced Computed Tomography (MSCT) examination was done to determine the position of the tethered cord. MSCT revealed spina bifida at L5 until S2 level with features of lipomyelomeningocele, and it also showed that the spinal cord was fixed on the dorsal aspect of the spinal canal (tethered cord) started from L2 level. Distended bladder, bilateral hydronephrosis, and hydroureter were also found on MSCT (**Figure 2**).

Surgery under general anesthesia to release tethered cord and total excision of the lump were performed in this case. The excised lump was sent to Anatomical Pathology Laboratory. Histopathology findings showed that the specimen was composed of collagen connective

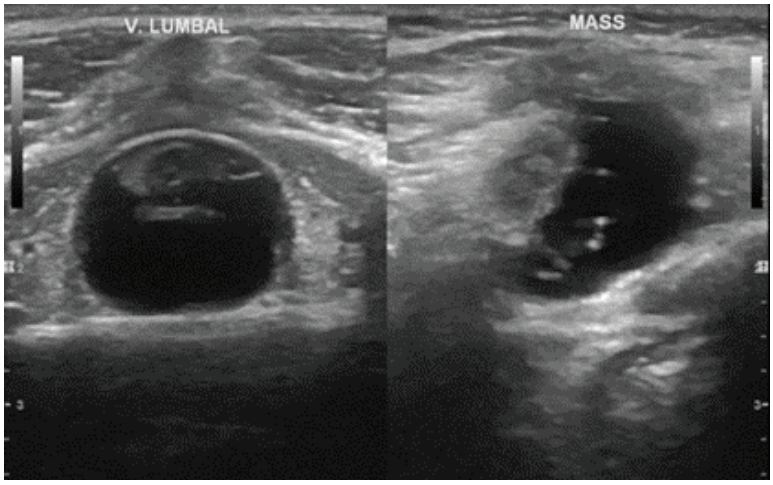

Figure 1. USG showed an anechoic lesion with multiple linear structure on supragluteal sinistra with a size of 2.2x2.5x2.1 cm. The conus medularis is fixed on the dorsal aspect of the lumbar spinal canal.

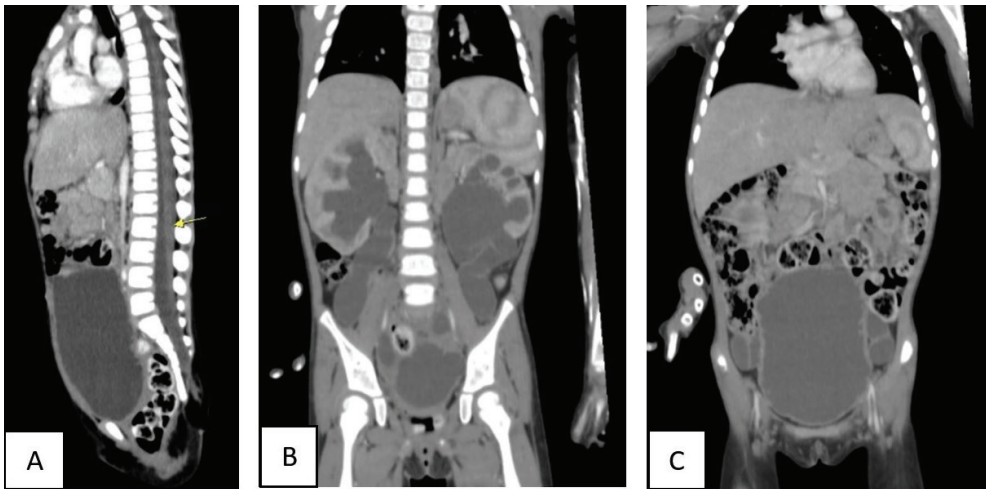

Figure 2. Multi Sliced Computed Tomography (MSCT) examination revealed (A) Tethering of cord (sagittal view), (B) Bilateral hydronephrosis and hydroureter (axial view) and (C) Distended bladder (pine tree appearance) possibly caused by neurogenic bladder.

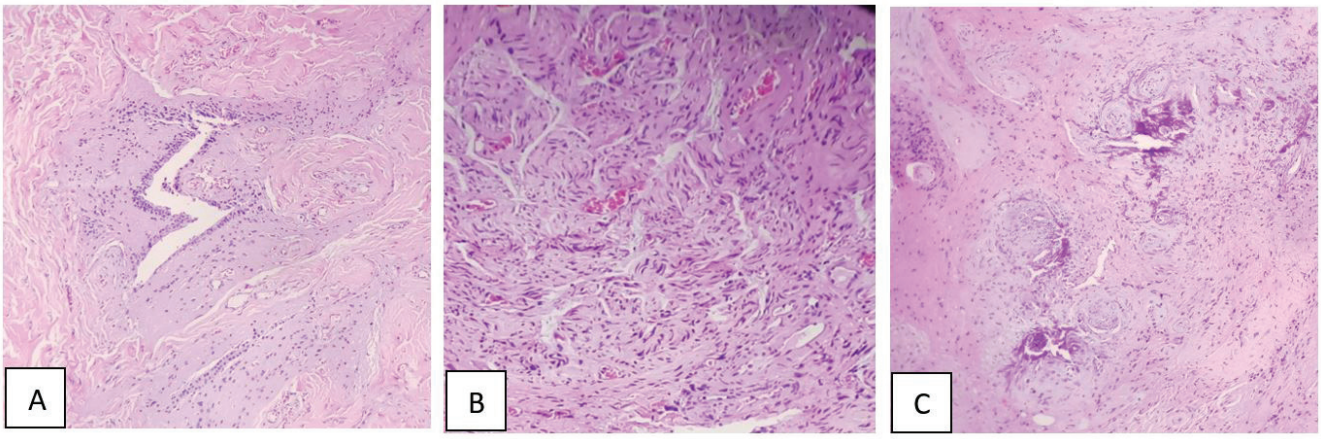

Figure 3. Microscopic finding shows (A) group of neuronal and glial cells, (B) meningothelial cells forming syncytial sheet pattern, and (C) foci of calcification (400x, H&E).

tissue and mature lipomatous tissue, syncytial sheet pattern of meningothelial cells, groups of neuronal and glial cells, and foci of calcification (**Figure 3**).

Based on clinical presentation, laboratory examination, imaging, and histopathological findings, the patient was diagnosed with lipomyelomenigocele, urinary tract infections, bilateral hydronephrosis, neurogenic bladder, and chronic constipation secondary to a tethered cord. There was no observed surgical complication, and the patient was discharged 3 days post-operation.

DISCUSSION

Spinal dysraphism is a congenital defect of the spinal cord that resulted from incomplete fusion of the spinal column. The prevalence of spina bifida varies depending on the region. Spinal dysraphism occurs in 1.87 and 6.25 per 1,000 live births in Malaysia and China, respectively. In Indonesia, there are no reports on the prevalence of spinal dysraphism. However, a study conducted at Yasri Hospital, Jakarta, found that spina bidifa constituted for 40% of congenital abnormalities [7–9].

The pathomechanisms of spinal dysraphism remain poorly understood, but some risk factors such as genetic predisposition, folic acid deficiency, vitamin B12 deficiency, and maternal obesity are related to spinal dysraphism. Folic acid may be associated with spinal dysraphism through its role in several processes including methylation, synthesis of DNA, RNA, and some nucleotide. During pregnancy, rapid cell replication and development of neural tubes require folic acid to facilitate DNA synthesis and methylation processes. If the need for folic acid is not met, disturbance of cellular replication and development of the neural folds may occur [10].

Providing folic acid supplementation during pregnancy is a way to prevent spinal dysraphism. However, despite the Indonesian national program for folic acid supplementation that has been conducted, the effectiveness of the program is still low due to low compliance. Approximately 20% of pregnant women did not consume any folic acid tablet, and only 30% of pregnant women consumed more than 90 tablets of folic acid as recommended. In addition, low access to care, inadequate supply, low attendance of antenatal care visits, and delay of ANC initiation also contribute to the low consumption of folic acid [11,12].

Spinal dysraphism has been associated with several comorbidities. One of the important comorbidities is TCS, a rare condition due to attachment of the spinal cord to surrounding tissues within the spinal canal. Although congenital tethered cord presents at birth, the symptoms do not appear immediately, but progress slowly and mostly appear during childhood. This happens because, during childhood, the spinal column growth is faster than the spinal cord growth. As the growth progresses, the spinal cord will ascend freely. If there is an abnormal adhesion of the spinal cord, it will stretch the spinal cord, and hypoxia of conus medularis will slowly occur [13].

Patients with tethered cord syndrome can experience a variety of symptoms including cutaneous, neurologic, orthopedic, and urologic abnormality. Closed spinal dysraphism presented as cutaneous mass and urinary tract infections secondary to neurogenic bladder were found to be the most common clinical presentation in 59% and 18% patients, respectively [2]. In this case, the patients presented with a mass in the sacral region and symptoms of urinary tract infection, which corresponds to common findings in TCS based on literature. We can say that recognition of TCS was delayed because the diagnosis was made after the patient experienced severe symptoms.

The absence of innervation to the bladder in the neurogenic bladder may be caused by embryogenesis malformation or secondary to TCS. This condition will result in poor bladder control. Failure to control bladder emptying may cause recurrent urinary tract infections, built-up pressure in the bladder, reflux of urine to

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the ureter and kidney, hydronephrosis, and renal damage [5].

In cases with TCS suspicion, a radiological examination should be performed to confirm the diagnosis and to detect urologic consequences of TCS such as distended bladder and hydronephrosis [2]. Imaging modality that can be used includes ultrasonography, CT scan, and MRI. Ultrasound is the simplest modality and is considered the first-line imaging modality to screen TCS. However, ultrasound results are often operator-dependent [6].

Magnetic Resonance Imaging (MRI) is still considered the standard diagnostic tool to diagnose TCS. However, MRI is expensive and time-consuming, and pediatric patients often require sedation to remain still during the examination and avoid motion artifacts. MRI is used to visualize the location of conus medularis and the thickness of filum terminale. Conus medularis below L2 and filum terminale thickness more than 2 mm are typical findings in TCS. MRI also may visualize the cause of tethering. Although not widely used, MSCT may also be used to identify TCS [3,6]. In our case, we could not perform MRI due to our hospital limitation of imaging modality. Therefore, we performed MSCT. MSCT has several disadvantages compared to MRI. MSCT examination cannot determine the cause of tethered cord because MSCT is inferior to MRI in the capability to visualize soft tissue structure. It is also difficult to evaluate the spinal cord using MSCT, making it hard to determine the spinal cord's position accurately which may affect surgical planning [14].

Untethering surgery was done on this patient. Surgery is the main treatment option for TCS. Incision at the back region is performed under general anesthesia. Laminectomy is usually done for easier access to the spinal cord and untethering is subsequently carried out by releasing all attached connecting tissue from the spinal cord and conus medularis [15]. Clinical improvement occurs in 85–95% of patients who undergo surgery. Although not all impairment can be recovered by surgery, immediate surgery will undeniably result in better improvement and decrease the chance of further decline [16].

Histopathological examination is important to confirm the type of spinal dysraphism. In our case, histopathological examination revealed lipomyelomeningocele features. Previous studies found that the most frequent spinal dysraphism associated with tethered cord syndrome is lipomyelomeningocele (34.3%) which is consistent with our findings [3].

CONCLUSIONS

TCS is a neurological disorder caused by the attachment of the end of the spinal cord to the tissue that surrounds it. TCS is frequently related to occult spinal dysraphism, especially lipomyelomeningocele. Regular follow-up and tight observation of patient symptoms are required in the case of spinal dysraphism as symptoms may not be present at the beginning. Late recognition of symptoms and long duration of tethering may affect the outcome of TCS.

DECLARATIONS

Competing of Interest

The authors declare no competing interest in this study.

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