

Case Report

An unusual case of two spinal level diastematomyelia with cord tethering

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ABSTRACT

Diastematomyelia at two spinal levels is a rare disease. And also, a dorsal and lumbosacral spur along with cord tethering and spina bifida has been rarely reported in the literature. Diastematomyelia is a rare congenital anomaly that results in the “splitting” of the spinal cord in a longitudinal direction. It occurs in the presence of an osseous (bone), cartilaginous, or fibrous septum in the central portion of the spinal cord which then produces a complete or incomplete sagittal division of the spinal cord into two hemicords. When the split does not reunite distally to the spur, the condition is referred to as a diplomyelia, or true duplication of the spinal cord. In this study, we will report a 16 months' old girl who was diagnosed with spina bifida during her intrauterine life and underwent division of bony spur at D10 to D11 and L4 to L5 and detethering of cord with intra-operative monitoring on day at 1 year and 6 months' of age.

Keywords: Cord tethering, Diastematomyelia, Spina bifida, Spur

INTRODUCTION

Split cord malformations (SCMs) are rare congenital anomalies in which the cord is split over a portion of its length to form a double neural tube in a single dural sac.^[1,2] SCMs are associated with a split of the spinal column, spinal bony spurs, myeloceles, myelomeningoceles, lipomas, and dermal sinuses.^[3-5] Here, we are presenting one such rare case.

CASE REPORT

The patient's mother was diagnosed as having a fetus with spinal dysraphism (spina bifida and horse shoe kidney) at her 6 months transabdominal ultrasound. She was delivered by spontaneous vaginal delivery at 37 weeks of pregnancy with Apgar score of 8/1 and 9/5, respectively.

On examination, this female patient was awake and alert. The patient had no apparent pallor, jaundice, edema, cyanosis, or clubbing. Vitals were: Pulse – 142 beats/min, blood pressure – 95/65 mmHg, respiratory rate – 28 breaths/min, and temperature – 36.6°C. Systemic examination revealed soft and non-tender abdomen; cardiovascular system examination – S1+S2+0; respiratory system examination – chest clear and normal vesicular breathing; higher mental functions were intact, Glasgow Comma Scale was 15/15, cranial nerves were intact; vision was intact, pupils were bilaterally equal and reactive; Motor – bulk and tone were normal in both limbs, moving all four limbs; plantars – bilateral downward going and sensory functions were intact; there were no cerebellar signs.

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She has a shield chest, with spinal dysraphism and spina bifida with tuft of hair at the back (lumbar swelling with intact skin). She was followed in neurosurgery clinic during her 1st year of life. MRI whole spine was advised, and it was followed by surgical removal of osseocartilagenous spur along with detethering of cord [Figures 1 and 2].

She had a right tip toe walking on gait assessment at 16 months of age on her second clinical visit. She underwent division of bony spur at D10-D11 and L4-L5 and detethering of spinal cord with intra-operative monitoring.

Intraoperatively, bony spur at D 10-D11 penetrating the dura was found. Another bony spur at L4-L5 penetrating the dura and dividing the cord into two with main thecal sac at the left side was also found.

Patient was kept in prone position with reverse trendelenberg posture. Post-operative recovery was uneventful and she was discharged home after 2 days. She developed non-significant hematuria which was self-resolving.

DISCUSSION

SCM is a rare spinal anomaly that refers to a longitudinal division of the spinal cord into two divided hemicords. This malformation has been reported to be associated with a split of the spinal column, spinal bony spurs, myeloceles, myelomeningoceles, lipomas, and dermal sinus. SCMs are often located in the lumbar and thoracolumbar regions. The incidence rates for cervical and cervicothoracic locations as 3% and 1%, respectively, have been reported. Symptoms related to congenital anomalies occur most commonly in childhood, so SCMs were initially regarded as a pediatric problem; but in many patients, the diagnosis is not established until symptoms manifest in adulthood.^[4,5]

Embryologically, the spinal cord is formed by the integration of two paramedian notochords along the midline, any defect in this integration leads to spur formation.

SCMs are classified as one of two types, according to the unified theory – in Type I SCM, the hemicords are always invested with individual dural sacs and the medial walls of the sacs always ensheath a rigid (bony or cartilaginous) midline spur; and in Type II SCM, the hemi-cords are always within a single dural sac and the midline septum is always composed of non-rigid fibrous or fibrovascular tissues. Keeping all these points in consideration, our case would be classified as Type I.

An awareness of the presence of associated congenital anomalies is surgically important to improve post-operative outcomes and to determine surgical priorities.^[6,7] For example, if the SCM is accompanied by a thickened filum terminale, the septum should first be excised to release the cord.

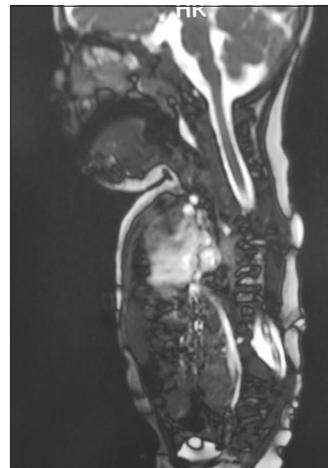


Figure 1: Sagittal view of spinal cord.

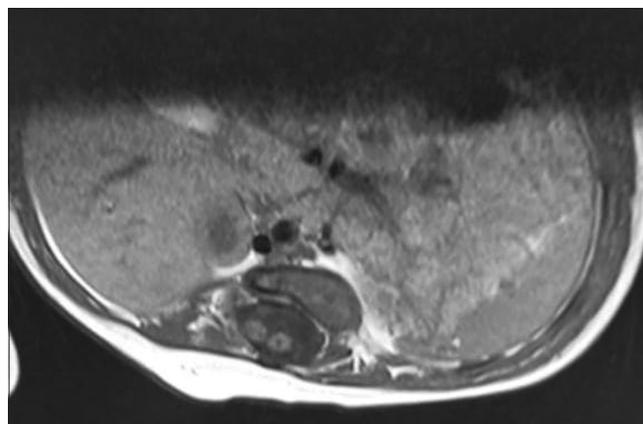


Figure 2: Demonstrating axial view of spinal cord. Cord division and syrinx.

CONCLUSION

Two spinal level diastematomyelia with cord tethering and spina bifida is very rare. Our patient had all four findings.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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