Diastematomyelia

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Abstract

Diastematomyelia is a rare congenital malformation, accounting for approximately 5% of all congenital spinal anomalies. It is characterized by a longitudinal split of the spinal cord into 2 hemicords and often presents alongside other neural tube defects. Diastematomyelia is classified into 2 types: type 1, which involves a bony or cartilaginous septum separating the hemicords, and type 2, where no septum is present. Clinical manifestations include hypertrichosis, back pain, lower limb motor and sensory deficits, foot deformities, and scoliosis, though some patients may remain asymptomatic. Diagnosis is typically made with CT or MRI. Surgical correction is the standard treatment, even in asymptomatic patients, to prevent future neurological decline.

Keywords: central nervous system, spine, congenital

Case Summary

A child presented for spine imaging due to the presence of a hairy tuft in the mid back. The patient had no neurologic symptoms.

Imaging Findings

Spine MRI (Figure 1) showed a midline bony bar extending from the dorsal aspect of the T10-11-intervertebral disc space and extending to the posterior elements. The bony bar caused a diastematomyelia of the spinal cord extending from the T9-10 intervertebral disc to the level of the T11-12 intervertebral disc.

Diagnosis

Diastematomyelia.

Differential diagnoses included tethered cord syndrome, myelomeningocele, syringomyelia, and lipoma.

Discussion

Diastematomyelia is a congenital malformation characterized by a longitudinal division of the spinal cord into 2 hemicords. It accounts for approximately 5% of all congenital spinal malformations. Classification is based on anatomical features. Type 1 diastematomyelia is the more severe form, in which each hemicord is contained within its own dural sac and separated by an extradural bony or cartilaginous septum. In type 2 diastematomyelia, both hemicords share a single dural sac without an intervening septum.[1,2]

Diastematomyelia is thought to be more common in females.[3] Although the malformation is present at birth, symptoms typically emerge during childhood. Diastematomyelia frequently coexists with other congenital anomalies, including congenital scoliosis, syringomyelia, myelomeningocele, dermoid cysts, hemivertebra, and tethered cord syndrome.[3,4] Common clinical manifestations include hypertrichosis, back pain, lower limb motor and sensory deficits, foot deformities, and scoliosis. Some patients may also experience bowel and bladder dysfunction. Patients with type 2 diastematomyelia are often asymptomatic, with the condition frequently discovered incidentally.[5]

Although there is some evidence suggesting a genetic predisposition, the exact etiology of diastematomyelia remains poorly understood. Both type 1 and 2 diastematomyelia are believed to originate from a shared embryologic error during neurulation, resulting in the formation of a persistent accessory neurenteric canal. This canal creates an abnormal communication between the yolk sac and the amnion, leading to splitting of the neural plate and primitive disk.[6]

Diastematomyelia is typically diagnosed with CT or MRI, with MRI being the preferred modality. CT or CT myelography is useful for assessing spinal canal widening, interpedicular spacing, and the presence of a bony septum separating the hemicords. CT can also evaluate for extra-axial compression from bone anomalies, nerve root compression, or disc protrusions. MRI provides superior soft tissue detail and can determine whether the hemicords are contained within separate dural sacs or share a single

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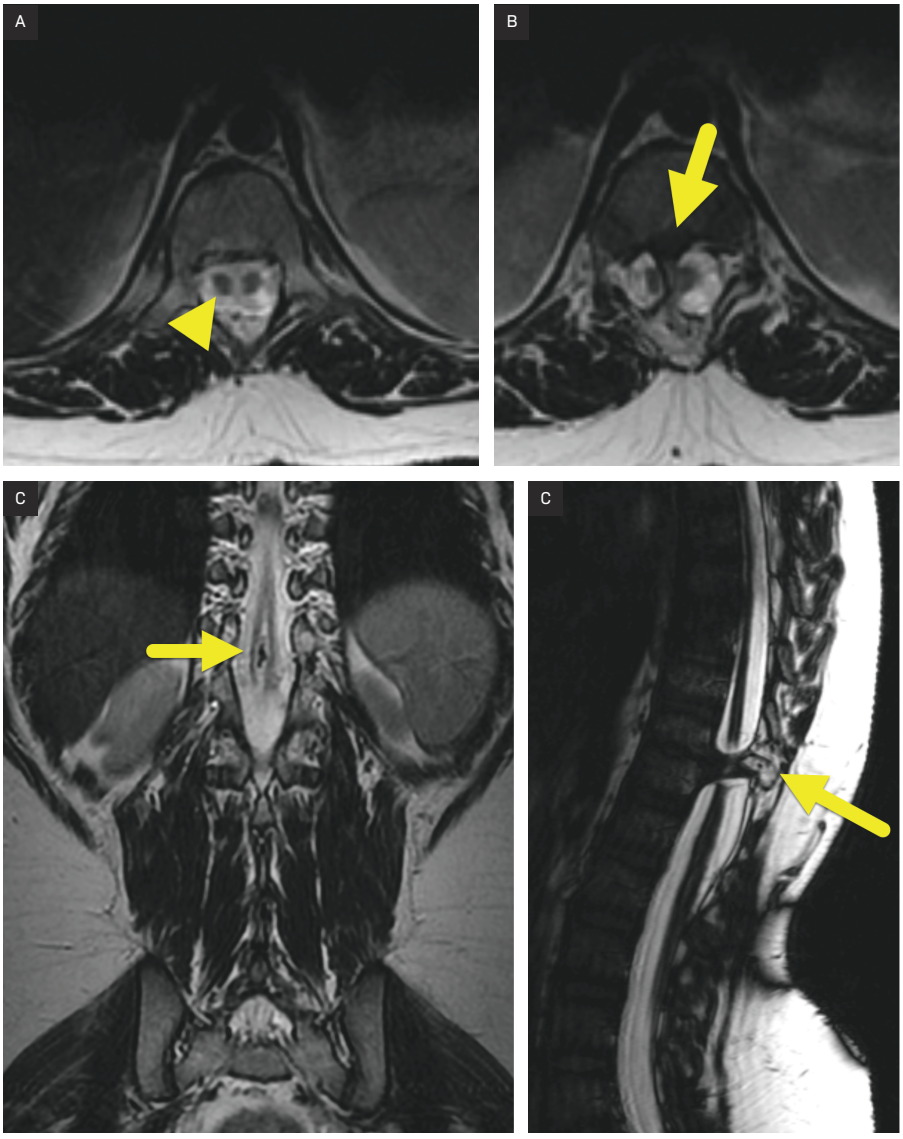
Figure 1. (A, B) Sequential axial T2-weighted images showing a split cord malformation (arrowhead) beginning just above the level of a bony bar (arrow). (C) Coronal T2-weghted image showing the split cord malformation caused by the bony bar (arrow). (D) Sagittal FIESTA image highlighting the extent of the bony bar (arrow).



sac, thereby distinguishing type 1 from type 2 diastematomyelia.[2]

Surgical intervention is the treatment of choice for diastematomyelia. In patients with a type 1 malformation, the osseous or cartilaginous septum separating the hemicords is excised. In both types of diastematomyelia, the dural sac is reconstructed to create a single, unified sac encasing the spinal cord. There is consensus that corrective surgery should be performed even in asymptomatic patients to prevent future neurological deterioration.[3]

Conclusion

Diastematomyelia is a rare congenital malformation, accounting for approximately 5% of all congenital spinal anomalies. It is characterized by a longitudinal split of the spinal cord into 2 hemicords and often presents alongside other neural tube defects. Diastematomyelia is classified into 2 types: type 1, which involves a bony or cartilaginous septum separating the hemicords, and type 2, where no septum is present. Clinical manifestations include hypertrichosis, back pain, lower limb motor and sensory deficits, foot deformities, and scoliosis, though some patients may remain asymptomatic. Diagnosis is typically made with CT or MRI. Surgical correction is the standard treatment, even in asymptomatic patients, to prevent future neurological decline.

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