

Case 2069: Type I split cord malformation: CT and MR findings

Subspecialty: Neuroradiology

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Patient:

Age: 47 year(s)

Sex: female

Clinical Summary

Patient presenting with a history of right leg pain.

Clinical History and Imaging Procedures

The patient presented with right leg pain. Physical examination showed scoliosis and neurological examination showed left leg sensory loss at the L5 level. There was no history of bladder or bowel disturbance. Gait disturbances (claudication) were present. The patient underwent lumbar CT and spine MR examinations. CT showed bifid lamina at the L4 level associated with the presence of a thin osseous septum of the spinal canal. Spine MR examination showed the presence of a double spinal cord each contained within its own dural tube at the L4 level and a low lying conus medullaris at the L5 level. The presence of a small syringomyelia was found in the dorsal tract.

Discussion

Split cord malformations (SCMs) are rare spinal anomalies and their classification is still a matter of debate. In 1992 Pang et al. proposed a "unified theory" attempting to explain the embryogenetic mechanism as a basic error occurring around the time when the primitive neuroenteric canal closes. The new classification recommended the term SCMs for all double spinal cords. In this classification all SCMs show a common finding of incomplete spinal cord duplication and differ only in the nature of the median septum and the number of dural tubes. SCM type I consists of two hemicords, each contained within its own dural tube and separated by a rigid osseocartilaginous median septum. SCM type II consists of two hemicords contained in a single dural tube separated by a fibrous median septum. Most SCMs are found in the lumbar tract; however, they can also be found in the cervical or, less frequently, in the dorsal tract. Symptoms of SCMs include progressive sensorimotor loss and pain. Neurological deterioration in SCMs is independent of the lesion type or location. Scoliosis, spina bifida, vertebral anomaly and subcutaneous manifestations are also observed with SCMs. Hypertrichosis represents one of the best predictors of an underlying SCM, being found in 56% of SCM patients. Neurosurgical therapy includes release of the tethered hemicords by eliminating the bone septum, dural sleeves and fibrous septa. The natural history of these lesions supports an early surgical approach to untether the spinal cord before neurological deterioration begins.

Final Diagnosis

Type I split cord malformation

Figure 1: Axial CT



Figure 1a

Axial CT imaging shows the presence of a bony septum at the L4 level dividing the spinal canal into two asymmetric parts (arrow).

Figure 2: Axial T1-weighted MR image

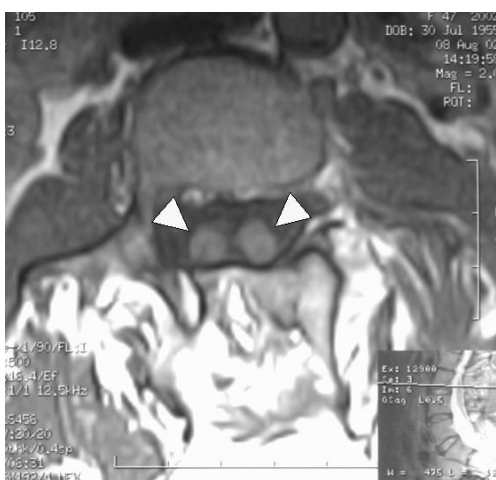


Figure 2a

Axial T1-weighted MR image performed at the same level as Figure 1 shows two distinct hemicords contained within their own dural tubes (arrowheads).

Figure 3: Axial T1-weighted image



Figure 3a

Axial T1-weighted MR image performed at the L5 level shows the low lying conus medullaris tethered posteriorly in the spinal canal (arrow).

Figure 4: Coronal T2-weighted image

Figure 4: Coronal T2-weighted image



Figure 4a

Coronal T2-weighted MR image shows a small syrinx at the dorsal tract (arrow). Note the presence of lumbar scoliosis.

Figure 5: Coronal T2-weighted images

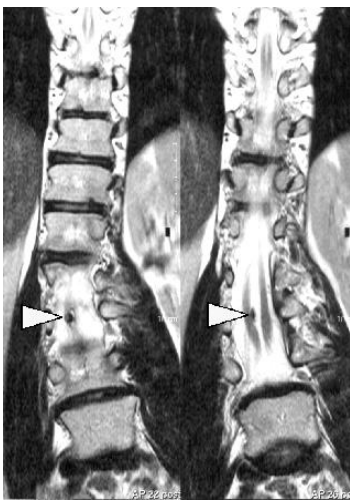


Figure 5a

Coronal T2-weighted MR contiguous images clearly show the presence of the septum (arrowheads).

Figure 6: Sagittal T2-weighted image



Figure 6a

Sagittal T2-weighted MR image shows the low lying tethered conus medullaris (arrowhead).

MeSH:

[C10.500.680] Neural Tube Defects

Congenital malformations of the central nervous system and adjacent structures related to defective neural tube closure during the first trimester of pregnancy generally occurring between days 18-29 of gestation. Ectodermal and mesodermal malformations (mainly involving the skull and vertebrae) may occur as a result of defects of neural tube closure. (From Joynt, Clinical Neurology, 1992, Ch55, pp31-41)

[C10.500.680.800] Spinal Dysraphism

Congenital defects of closure of one or more vertebral arches, which may be associated with malformations of the spinal cord, nerve roots, congenital fibrous bands, lipomas, and congenital cysts. These malformations range from mild (e.g., SPINA BIFIDA OCCULTA) to severe, including rachischisis where there is complete failure of neural tube and spinal cord fusion, resulting in exposure of the spinal cord at the surface. Spinal dysraphism includes all forms of spina bifida. The open form is called SPINA BIFIDA CYSTICA and the closed form is SPINA BIFIDA OCCULTA. (From Joynt, Clinical Neurology, 1992, Ch55, p34)

References:

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