

ORIGINAL ARTICLES

Split cord malformation type I: Technical notes on the subject of a rare variant of a rare malformation

Bruno Lourenço Costa, José Augusto Costa, Gustavo Bento Soares, Carla Domingos, Marcos Barbosa

Department of Neurosurgery, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal.

Correspondence: Bruno Lourenço Costa. Address: Centro Hospitalar e Universitário de Coimbra, Department of Neurosurgery, Praceta Professor Mota Pinto, 3000-075 Coimbra, Portugal. Email: brunolourencocosta@gmail.com

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Abstract

Background: Split cord malformation type I is a rare form of occult spinal dysraphism. A median septum arising from vertebral arches is an exceedingly rare variant with only a few cases described.

Methods: Description of clinical case and surgical technique.

Results: We describe the case of a neurologically intact 2-year-old boy who was diagnosed with a split cord malformation type I. Radiological examination revealed an osseous septum in the vertebral canal arising from L2 and L3 arches and separating two independent hemicords. Septum removal, cord partial untethering and duramater reconstruction were performed without associated morbidity. The patient has been followed by a multidisciplinary team for five years. A subtle and non progressive left foot deformity was observed. We review practical aspects of the surgical treatment of this malformation.

Conclusion: The diagnosis of associated vertebral malformations and tethering lesions is critical. Paramedian laminectomy, subperiosteal spur dissection and watertight reconstruction of posterior duramater represent important technical variations related to the posterior origin of the median septum.

Keywords

Diastematomyelia, Occult dysraphism, Split cord malformation, Surgical technique

1 Introduction

Split cord malformation (SCM) type I, also known as diastematomyelia, is an exceedingly rare form of occult spinal dysraphism, characterized by the presence of a rigid osseocartilaginous median septum in the spinal canal that sagittally separates two hemicords with individual meninges^[3]. This congenital malformation is typically found at the lumbar region varying the longitudinal extension of vertebral column/spinal cord involvement. An osseocartilaginous septum arising from posterior arches is particularly uncommon and implies changes in the surgical technique. Other regional vertebral malformations and other anchoring lesions throughout the neuroaxis frequently coexist and must be accurately recognized.

2 Case description

A previously healthy 2-year-old male presented with cutaneous nevi and a painless lump located in the median lumbar region. General and neurologic examinations were found to be normal. Spine imaging revealed a split cord malformation type I characterized by a median osseous septum arising from the posterior arches of L2 and L3 (see Figure 1A, 1B and 1C) as well as two independent hemicords (see Figure 1E and 1F). Syringomyelia T11-L2 and low-lying conus medularis (L4-L5 disk level) coexisted (see Figure 1D). MRI did not show other tethering anomalies throughout the neuroaxis. Urodynamic testing showed normal bladder function. Uneventful L2-L3 laminectomy, osseous septum removal and posterior meningeal reconstruction were performed. The patient has been monitored by an interdisciplinary spina bifida team for 5 years regarding spinal/orthopedic deformities and neurological/urodynamic decline. A subtle and non progressive lateral deviation in the left foot was noted three years after the surgery. Low-lying conus medularis, ventral adhesion of the two hemicords and syringomyelia remained unaltered during the follow-up period.

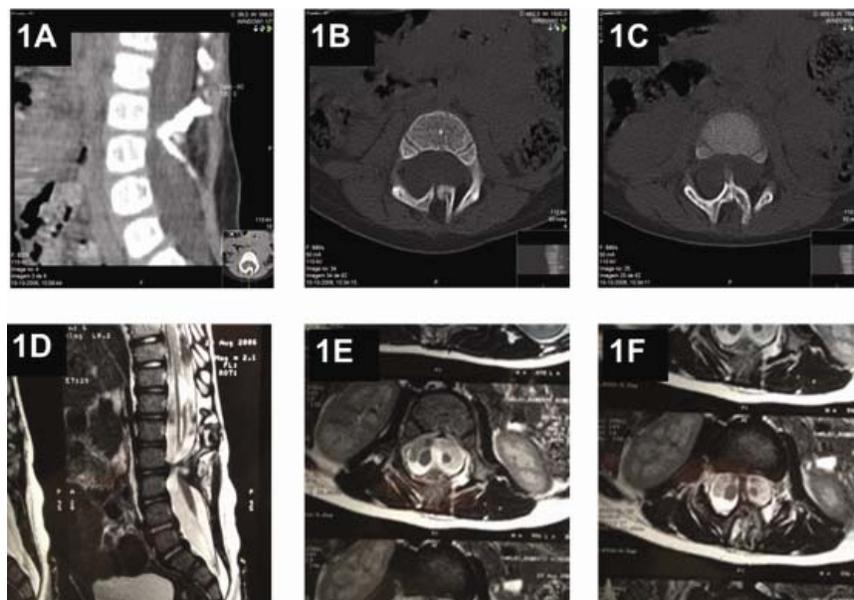


Figure 1. Non contrast-enhanced spine CT scan, sagittal (A), axial at L2 level (B) and axial at L3 level (C) images showing a median osseous septum arising from posterior arches of L2 and L3. Deformity and bony malformations were absent except for hypertrophic, bifid and fused neural arches. Spine MRI scan, T2-weighted sagittal (A), axial at L2 level (B) and axial at L3 level (C) images showing two hemicords with independent dural sleeves and subarachnoid spaces as well as syringomyelia at T11-L2 levels and low-lying conus medularis (L4-L5 disk). MRI did not show other tethering anomalies throughout the neuroaxis.

3 Surgical technique

The patient was positioned prone. A posterior midline approach exposed the posterior arches of T12-L3 (see Figure 2A). Special care was taken to avoid durotomy and neural injury given the existence of posterior arch defects. Paramedian laminectomy of L2 and L3, preserving the spinous processes, midline laminas and insertion of the spur was performed. Once the spur was isolated and exposed, subperiosteal dissection separated the duramater bilaterally. Once freed, the spur was removed with rongeur (see Figure 2B and 2C). The duramater was sharply open in the midline above and below the split and at duplicated dural sleeves (see Figure 2D). Subarachnoid space was carefully inspected and adhesive bands attaching the hemicords to the duramater were divided. The two hemicords were firmly adherent to the ventral duramater. Dissection was not performed given the risk of neural injury. The redundant duramater was resected and the normal configuration of the dural sac was restored with uninterrupted watertight suture (polypropylene 4/0). Fasciae,

subcutaneous plane and skin were closed with interrupted suture. The patient was kept flat for two days in order to avoid CSF fistulas.

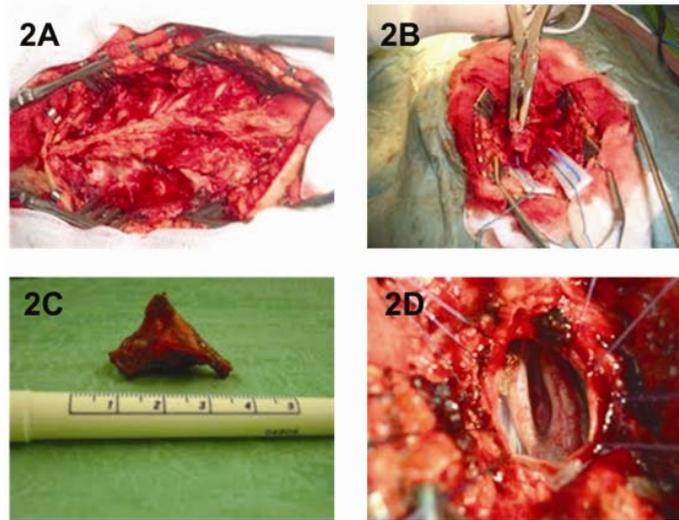


Figure 2. Intraoperative photographs. See surgical technique

4 Discussion

Surgical treatment of SCM type I is advocated to prevent, improve or stabilize symptoms and neurological deficits. The main goal is to remove the spur and to untether the spinal cord by operative management of both the SCM and any associated tethering anomalies. Outcomes are more favorable in asymptomatic patients [2, 4]. Complications associated with repair of SCM include CSF leak, wound infection and new neurologic deficits [1, 2, 4-7].

The criticality of diagnosis of associated vertebral malformations and associated tethering lesions must be emphasized. Paramedian laminectomy, subperiosteal spur dissection and watertight reconstruction of posterior duramater represent important technical variations related to the posterior origin of the median septum. Firm ventral adhesion of the two hemicords impaired complete untethering. The patient was neurologically intact and surgery was not associated with deterioration. However, subtle foot deformity diagnosed when the patient was 5 years-old deserves careful monitoring.

5 Conclusions

Split cord malformation type I is a rare form of occult spinal dysraphism. An osseocartilaginous septum arising from posterior arches is an exceedingly rare variant. Subtle technical aspects enhance the safety and effectiveness of its surgical treatment. Long-term follow-up by an interdisciplinary spina bifida team is mandatory in order to manage spinal/orthopedic deformities and persistent/recurrent cord tethering.

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