Split Cord Malformation: Presentation, management and surgical outcome

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Disclosure

The authors declare that the content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.
Background

• Split cord malformation (SCM), also known as diastematomyelia, is a rare anomaly characterized by a split along the midline of the cord, which divides it into 2 symmetric or nonsymmetric entities.

• SCM surgical indications and outcomes are still debatable, the signs and symptoms are generally nonspecific and is commonly associated with other anomalies and deficits.

• In this study we reviewed and analyzed data from all patients with SCM who have presented at our center for the last 20 years, including presentation management and outcomes of surgically treated patients.
Methodology

We retrospectively searched the hospital database.

At KAMC-Riyadh, SA

For patients diagnosed with SCM between 1998 and 2018.

Descriptive statistics were used to present categorical data as percentages and frequencies.

Ethical Considerations: Approval was obtained from institutional review board at King Abdullah International Medical Research Center, Riyadh, Saudi Arabia.
Results:
I. Baseline characteristics

- A total of 25 patients were included in this series.
- The mean age of patients at the time of diagnosis was 4.4 years.
- Most (n = 21, 84%) were female.
- Types of split were almost equally distributed: 12 (48%) were type I and 13 (52%) were type II.
Results:
II. Main presenting signs and symptoms

• Approximately half of the patients 14 (56%) presented with neurologic manifestations. Eleven (44%) had a bladder dysfunction at time of presentation.

• Skin stigmata were remarkable in only 12 patients (48%), 9 of whom had type II SCM. Tuft of hair and skin dimple were the most frequently encountered hallmarks.

• A total of 19 patients (69%) in this series had associated congenital spine deformities, represented mainly by scoliosis.

• Foot deformities (flat foot, pes valgus, cavuvarus, and club foot) were observed in 7 patients (28%).
Results

III. Radiologic and surgical findings

• Whole spine magnetic resonance imaging (MRI) was performed for all patients.
• The thoracolumbar level was the most common level of split, seen in 9 patients (36%).
• A cervical-level split was seen in only 1 patient. Syrinx was evident in 14 patients (56%).
• A total of 16 (64%) had coexisting spinal cord lesions (myelomeningocele, lipomyelomeningocele, and meningocele).
• Brain imaging showed Arnold-Chiari malformation type 2 in 4 patients (16%).
Results

III. Surgical outcomes and complications

- In this series, the mean difference between age at diagnosis and age at correction was 7 months.
- A total of 18 patients (72%) underwent surgical correction. All patients underwent intraoperative neurophysiologic monitoring.
- Postoperative complications were minimal. Cerebrospinal fluid (CSF) leakages were noted in 2 patients, transient urinary retention in 1 patient, and transient unilateral leg paresis in 1 patient.
- Most of the patients (n =15, 83%) were discharged within 19 days after surgery.
- The overall long-term outcomes were good. A total of 81% showed improvement (defined by either complete or partial symptom resolution, symptom stability, and/or hydronephrosis resolution).
- Over the long-term follow-up, none of the patients developed new urologic or neurologic deficits. Regrowth of bony spur was evident in only 1 patient.
Discussion

- SCM accounts for approximately 5% of all congenital spinal disorders.
- Skin stigmata frequently reported in approximately half or more than half of patients with SCM.
- Neurologic manifestations and bladder dysfunction are common among patients with SCM.
- Congenital scoliosis is the most commonly reported spinal deformity associated with SCM. According to the literature, the incidence of congenital scoliosis among patients with SCM ranges between 26% and 84%. Conversely, the rate of SCM among patients with congenital scoliosis varies from 5% to 16%.
- Coexisting spinal cord lesions among patients with SCM are not uncommon.
- In the literature, the overall reported early postoperative complications were minimal. The use of intraoperative neurophysiological monitoring in SCM corrective surgery were largely determined by its availability. The surgical indications are debatable. However, the majority considered prophylactic surgery for patients with SCM type I and surgical treatment for symptomatic patients of both types.
Conclusions:

Most patients with SCM present during childhood. Postoperative complications after SCM corrective surgery are generally minimal, and the overall outcomes are good. Outcomes, mainly including partial or complete symptomatic improvement and/or symptom stability and hydronephrosis resolution, were favorable. Early surgical intervention in all patients with type I SCM and symptomatic patients with type II SCM might be associated with better postoperative outcomes. We believe that intraoperative monitoring can reduce the rates of postoperative neurologic complications.