Case Report

An Unusual Case of Split Cord Malformation with Simultaneous Ventral and Dorsal Bony Spur at a Single Site: A Technical Challenge

Ashutosh Agarwal, Amol Raheja, Sachin A. Borkar, Ashok K. Mahapatra

Department of Neurosurgery and Gamma Knife, All India Institute of Medical Sciences, New Delhi, India

INTRODUCTION

Split cord malformation (SCM) is a relatively uncommon condition, wherein the cord is divided into two variable segments with a fibrous or osteocartilaginous septum in between and surrounded by a single or two dural sheaths, respectively. It may present in an asymptomatic child with characteristic neurocutaneous markers or with neurologic, urologic, or orthopedic deficits. Although many cases of composite-type SCMs have been reported in literature,[1,2] SCM with simultaneous ventral and dorsal bony spur at a single site is an extremely rare entity, with only one other case reported so far in the literature.[3] We present a second such case and discuss the relevant surgical implications and possible embryological mechanisms involved in such a complex SCM.

CASE REPORT

A 13-month-old female child presented to us with a skin dimple over the lumbar region with overlying hairy patch without any associated lower limb weakness or urinary complaints. Her physical examination revealed no other abnormalities. On imaging evaluation, magnetic resonance imaging showed a split cord from L2-L5 with broad-based spur at L4-L5 level with single thickened filament tethered at L5-S1 level with syrinx formation in the right hemicord from L1-L5 [Figure 1]. Noncontrast computed tomography showed deficient posterior elements of L5 to S1 with two bony spurs, one arising from the posterior aspect of L4 vertebral body and another from the hypertrophied L4 posterior arch [Figure 2].

The child was taken up for operative intervention, and intraoperatively, two bony spurs were confirmed with soft tissue in between—ventral spur based on the L4 vertebral body and a dorsal spur based on the hypertrophied L4 posterior arch. The cord was divided into two hemicords—right side larger than the left—which were reuniting below the spur to end in a single thickened fatty filament. There was a variable distance of split cords above the spurs suggestive of Type Ic SCM.[4] The spurs were excised with the help of a high-speed diamond drill, and the fatty filament was divided at S1 level. Dural bands

Access this article online

Quick Response Code:  
Website: www.pediatricneurosciences.com  
DOI: 10.4103/jpn.JPN_99_17

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Agarwal A, Raheja A, Borkar SA, Mahapatra AK. An unusual case of split cord malformation with simultaneous ventral and dorsal bony spur at a single site: A technical challenge. J Pediatr Neurosci 2018;13:214-7.
and arachnoid adhesions tethering the cord were released [Figure 3A]. Dura was closed primarily and reinforced with muscle patch and tissue glue. Postoperative hospital stay of the child was uneventful with no new onset of neurological deficits and the postoperative computed-tomography scan demonstrated complete excision of the spurs [Figure 3B].

**DISCUSSION**

According to the classification system proposed by Mahapatra and Gupta,[4] Type I SCM is subdivided into four types with prognostic significance:[5] Type Ia, Type Ib, Type Ic, and Type Id, with the last one having the highest chance of developing postoperative neurological deficits.[5] The present case was a Type Ic SCM as per this classification. Complex SCM with both ventral and dorsal spurs at the same spinal level was diagnosed preoperatively, and accordingly, care was taken to surgically isolate the hypertrophied posterior arch to prevent any undue traction on straddling cord during laminotomy. Once both the spurs were partially drilled extradurally, dura was opened to expose the base of the ventral spur and the tip of the dorsal spur along the L4 vertebral
According to the Unified Theory proposed by Pang et al. on the embryogenesis of SCM, embryogenesis of SCM involves formation of an accessory neurenteric canal rostral to the primitive neurenteric canal. If the precursor cells within the endomesenchymal tract contain the meninx primitiva cells, they form a bony septum and Type I SCM, else a fibrous band giving rise to Type II SCM. Multiple accessory canals may give rise to composite-type SCMs with variable splits at different locations. Generally, in cases of bony septum, a single bony spur usually arises from the posterior surface of the vertebral body, but there have been a few case reports of spurs arising from the posterior arch. Chandra et al. discussed two hypotheses for posterior origin of bony spur, including (1) disconnection of ventral cell mass after dorsal migration of meninx primitiva cells and (2) initial migration of meninx primitiva cells around the hemicords instead of between them to accumulate along the dorsal arch. It is quite unusual to have simultaneous ventral and dorsal bony spurs at the same spinal level. The reported child had soft tissue interposed between the two spurs, emphasizing that there is no single hypothesis that can explain such a unique observation. We believe that such cases are underreported in the literature and the true incidence of such complex SCMs is much higher than documented.

**Conclusion**

Pathogenesis of SCM is still poorly understood and it is a complex multifactorial congenital malformation, which requires further detailed study to elucidate the underlying embryological and pathological mechanisms. It requires detailed imaging evaluation in the preoperative period and utmost intraoperative care so as to prevent new onset deficits in such rare patients.

**Financial support and sponsorship**

Nil.
Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Allawadhi P, Mahapatra AK. An unusual case of spinal dysraphism with four splits including three posterior spurs. Pediatr Neurosurg 2011;47:372-5.
2. Erşahin Y, Demirtaş E, Mutluer S, Tosun AR, Saydam S. Split cord malformations: Report of three unusual cases. Pediatr Neurosurg 1996;24:155-9.
3. Naik V, Mahapatra AK, Gupta C, Suri V. Complex split cord malformation with mediastinal extension of a teratoma and simultaneous ventral and dorsal bony spur splitting the cord. Pediatr Neurosurg 2010;46:368-72.
4. Mahapatra AK, Gupta DK. Split cord malformations: A clinical study of 254 patients and a proposal for a new clinical-imaging classification. J Neurosurg 2005;103:531-6.
5. Borkar SA, Mahapatra AK. Split cord malformations: A two years experience at AIIMS. Asian J Neurosurg 2012;7:56-60.
6. Pang D, Dias MS, Ahab-Barmada M. Split cord malformation: Part I: A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery 1992;31:451-80.
7. Chandra PS, Kamal R, Mahapatra AK. An unusual case of dorsally situated bony spur in a lumbar split cord malformation. Pediatr Neurosurg 1999;31:49-52.
8. Prasad GL, Borkar SA, Satyarthee GD, Mahapatra AK. Split cord malformation with dorsally located bony spur: Report of four cases and review of literature. J Pediatr Neurosci 2012;7:167-70.
9. Akay KM, İzci Y, Baysefer A. Dorsal bony septum: A split cord malformation variant. Pediatr Neurosurg 2002;36:225-8.