

# A Case of Split Cord Malformation with Triple Bony Spur in a Single Dural Sleeve—A Classification Dilemma

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## Abstract

Split cord malformation-I (SCM-I) is characterized by the presence of double dural sacs, with rigid extradural bony/cartilaginous spur leading to symmetrical or asymmetrical division of the cord. In split cord malformation-II (SCM-II), there is a single dural sac with a nonrigid fibrous spur and symmetrical division of the cord. SCM-II are slightly more common than SCM-I, constituting around 50 to 60% of SCMs. The authors report a unique case of SCM with triple bony spurs lying both intra- and extradurally in a single dural sleeve. At the time of submission of this report, to the best of authors' knowledge, no case of SCM with triple bony spurs in single dural sleeve has been reported anywhere in the world literature.

## Keywords

- ▶ diastematomyelia
- ▶ bony spur
- ▶ split cord

## Introduction

Split cord malformation (SCM) is an uncommon congenital anomaly in which a segment of the spinal cord is divided into two parts by a fibrous or rigid bony spur. The bony septum usually originates from the vertebral body and passes posteriorly, causing splitting of the cord.

The most widely accepted theory about the embryogenesis of these complex malformations was originally proposed by Bremer in 1952<sup>1</sup> and subsequently modified by Pang et al<sup>2,3</sup> as the “unified theory of embryogenesis.” The basic error appears to be the development and persistence of “accessory neurenteric canal.” According to the theory, these lesions were divided into two types depending on the type of the midline mesenchymal derivative and the dural investment of the hemicords. SCM-I is characterized by the presence of double dural sacs, with rigid extradural bony/cartilaginous spur leading to symmetrical or asymmetrical division of the cord. In SCM-II, there is a single dural sac with a nonrigid fibrous spur and symmetrical division of the cord. SCM-II are slightly more common than SCM-I, constituting around 50 to 60% of SCMs.<sup>2,3</sup>

The authors report a unique case of SCM with triple bony spurs lying both intra- and extradurally in a single dural sleeve.

At the time of submission of this report, to the best of authors' knowledge, no case of SCM with triple bony spurs in single dural sleeve has been reported anywhere in the world literature.

## Case Summary

This was a case of a 3-year-old male child, who presented to the center with complaints of congenital deformity of spine and progressively worsening scoliosis of the spine along with abnormal hairy patch over midline in the back.

Clinically the patient had scoliosis to the right along with hypertrichosis. He, however, had no neurologic deficit and had no lower limb weakness or sphincter disturbance.

## Magnetic Resonance Imaging of Head was Normal

Magnetic resonance imaging (MRI) of the spine dated October 21, 2016 showed scoliosis in dorsal spine to right;

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multiple segmentation anomalies in the dorsal, lumbar, and sacral levels; splitting of the cord from D4 to L2 level; and three bony spurs at D6, D10, and D12 levels (►Fig. 1a, b) along with low-lying tethered cord with conus at the level of lower border of L3 vertebra along with a dorsal dermal sinus at sacral level. Computed tomographic (CT) of the spine showed bony spurs at the level of D6, D10, and D12 vertebrae (►Fig. 1c-f).

The patient was taken up for surgery and exploration. The site of split was explored first after adequately removing the spur and other tethering elements at the site. Filum de-tethering was performed in a single stage. Perioperatively the findings of MRI scan were confirmed in the form of there being a single dural sleeve (►Fig. 2a-c). The bony spurs were intradural for the maximum extent; however, because the bony spurs were attached to both the posterior aspect of the body of vertebrae and to the lamina after piercing out through the dural sleeve both ventrally and dorsally, they were technically deemed to be both intra- and extradural. The postoperative period remained uneventful, and at 4 months after surgery, he had no deficits.

**Discussion**

SCM-I is characterized by the presence of double dural sacs, with rigid extradural bony/cartilaginous spur leading into symmetrical or asymmetrical division of the cord. In SCM-II, there is a single dural sac with a nonrigid fibrous spur and symmetrical division of the cord. Among all the types of SCM, the composite-type SCM is very rare and results from two separate foci of ecto-endodermal adhesions and endomesenchymal tracts<sup>3</sup> leading to development of different SCM types with intervening normal cord in the same patient.<sup>4</sup> According to a classification system proposed by Mahapatra and Borkar,<sup>5</sup> SCM-I has been subclassified as follows:

- Type Ia: Bony spur in the center with equally duplicated cord above and below the spur
- Type Ib: Bony spur at the superior pole with no space above and a large duplicated cord lower down

- Type Ic: Bony spur at the lower pole with a large duplicated cord above
- Type Id: Bony spur straddling the bifurcation with no space above or below the spur

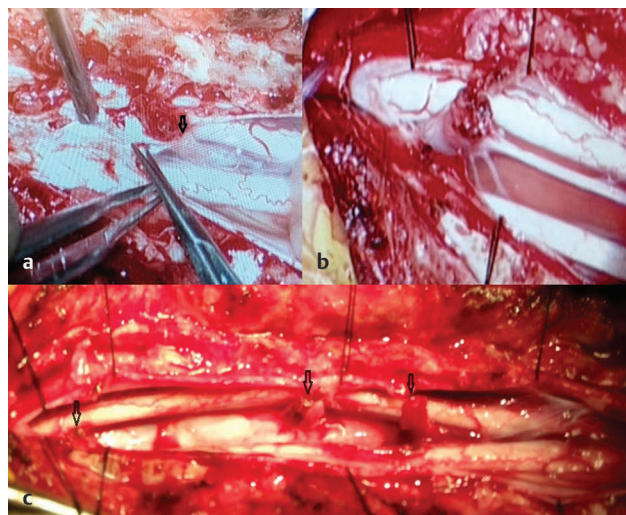
Only a few cases of composite-type SCM have been reported in the literature.

In this report, the authors present a case of case of SCM with triple bony spur in a single dural sleeve and multiple associated bony anomalies.

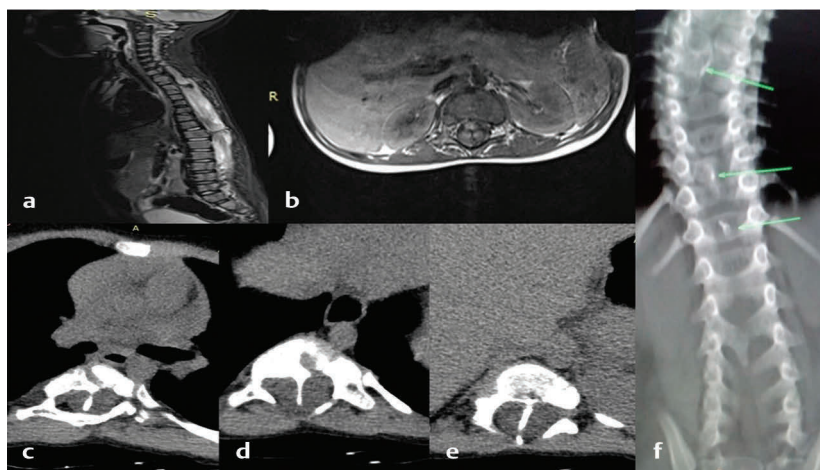
This case does not fit into any of the previously mentioned classification system.

**Conclusion**

The authors report the first documented case of “SCM with triple bony spurs in a single dural sleeve.” This case does not fit into any of the existing classification system of SCM



**Fig. 2** (a) Intraoperative image showing single dural sleeve—one spur visible (arrow). (b) Intraoperative image showing single dural sleeve—one spur visible at another level. (c) Intraoperative image showing single dural sleeve—all three spurs (arrows) visible.



**Fig. 1** (a) Sagittal section whole spine MRI image. (b) Axial section MRI of the spine showing bony spur. (c-e) Axial section NCCT of the spine showing bony spur at three levels. (f) Coronal section NCCT spine showing bony spur—DV6, DV10, DV12 (arrows).

although the clinical-radiologic findings in this patient seem to substantiate the multiple accessory neuroenteric canal theory in the development of this unique type of SCM.

**Conflict of Interest**

None.

**Acknowledgments**

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