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BRIEF COMMUNICATION

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Split cord malformation: three unusual cases of composite split cord malformation

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Abstract *Background:* All split cord malformations (SCMs) arise from one basic fault, i.e. the formation of an accessory neurenteric canal between yolk sac and amnion through the midline embryonic disc that splits the notochord and neural plate. Multiple accessory neurenteric canals may lead to two or more non-contiguous SCMs, also known as composite SCM. *Case histories:* We present here three cases of composite SCM. First, a 2 1/2-year-old boy had type II SCMs at two different levels with normal cord between them. In case 2, a 16-month-old girl revealed a type I SCM at levels L2–3 and a type II SCM at level L5. The third case had a combination of both these types of SCMs at levels L1–3. Only very few cases of composite SCM have been reported in the literature. *Conclusions:* These three cases also support the unified theory proposed by Pang et al.

Keywords Double cord formation · Split cord malformation · Diplomyelia · Diastematomyelia · Spinal dysraphism · Scoliosis

Introduction

Split cord malformations (SCMs) are congenital anomalies of the spinal cord in which the cord is split over a portion of its length to form a double neural tube [7]. Dachling Pang [5, 7], in 1992, advocated a new classification, recommending the term 'split cord malformation' for all double spinal cords, and also proposed a unified theory of embryogenesis to explain various components of these malformations. In this classification all SCMs show a common element of incomplete spinal cord duplication and differ only in the character of the associated mesenchymal structures. Type I consists of two hemicords, each contained with in its own dural sac and separated from its counterpart by a rigid bony or fibrocartilagenous spur covered by the dural sheath [5, 7]. Type II also consists of two hemicords, but they are contained in a single dural sheath and separated by a finer, more delicate intradural fibrous midline septum. Composite SCMs are very rare and result from two separate loci of ectoendodermal adhesions and endomesenchymal tracts [6] leading to development of SCMs with intervening normal cord between them in the same patient. Only very few cases of composite SCM have been reported in the literature [2, 6]. We present three cases of composite SCM having both type I or type II SCM or a combination of both.

Case reports

Case 1

A 2 1/2 -year-old boy presented with a hyperpigmented patch on his back and severe kyphoscoliosis in the dorsal region, with increased muscle tone in both lower limbs; there was no history of bladder, bowel or gait disturbances. Plain X-ray revealed bifid lamina at the lower thoracic vertebrae. MRI revealed the dorsolumbar kyphoscoliosis, with a type II SCM at T-7, below which the cord unified, and another type II SCM at T10–11(Fig. 1). The intervening cord segment had a small syrinx. Cervical spine evaluation revealed a type I ACM, but no syrinx was present at that level. Head MRI was normal. A T-7 to L-3 laminoplasty was done with the help of Midas Rex S-4 drill with foot plate attachment. At level T-7 a type II SCM was found with a thin fibrous septum, while at T10–11 no tethering lesion was found although the cord was split. The intervening cord segment was normal. The filum terminale was cut.

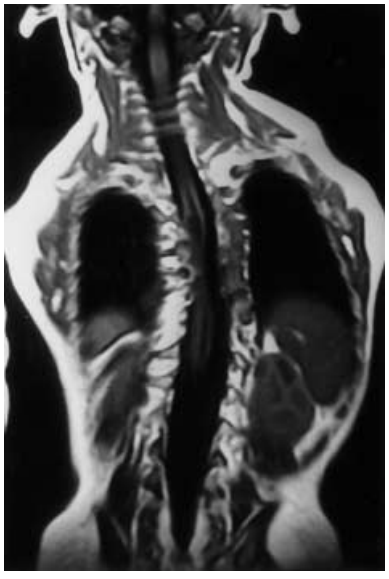

Fig. 1 T1-weighted coronal image showing composite split cord malformation (SCM) at two sites (case 1)

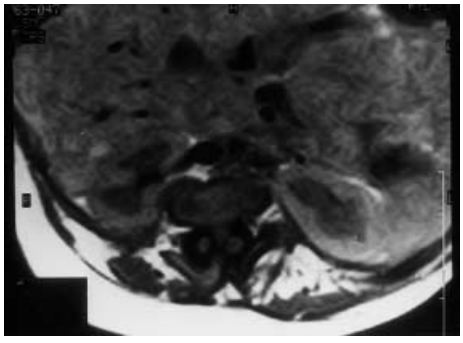

Fig. 2 Axial T1-weighted image showing SCM (case 2)

Case 2

A 16-month-old girl, who had been born at full term by a normal vaginal delivery at home, presented with scoliosis in the lower thoracic region, with the convexity towards the left. Developmental mile stones had been normal so far, and there was no neurological deficit. MRI revealed scoliosis in the lower thoracic region, splitting of the spinal cord at L1–3 with a bony spur at the same level, and low-lying conus at L-5 (Fig. 2). A skin incision was made from L-1 to S-1 (spinous processes). L1–3 laminectomy was performed; the bony spur was excised and unification of the dura was done. At L-5 there was a single dural sac, but two hemicords and a fibrous median septum were seen. This fibrous septum was attached to the median aspect of both hemicords. The fibrous septum was removed and the thick filum was cut. Between L3 and L5 the cord was normal.

Case 3

A 2 1/2-year-old boy presented with hypertrichosis over the lower back and kyphoscoliosis; he had no neurological deficit. MRI scan revealed splitting of spinal cord at the L2–3 level, with a bony spur and low lying conus medullaris at L-4 level. D12–L4 laminectomy was performed. Intraoperatively no bony spur was found, and there was a single dural sac. After dural incision, two hemicords and a fibrous median septum were seen at L1–3. The fibrous septum was dissected and removed. After removal of the fibrous septum a small bony spur arising from the posterior surface of body of vertebra at the same level was seen, which was invaginating into the dural sheath from the anterior aspect so that it looked like two dural sheaths anteriorly and only one posteriorly (Fig. 3). The dural sheath was incised and the bony spur was removed. The hemicords were reunited one vertebral level below the septum. The thickened filum was sectioned.

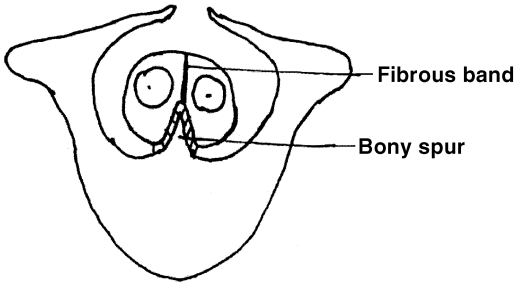

Fig. 3 Line diagram showing bony spur and fibrous band at one side (case 3)

The postoperative period was uneventful in all three patients.

Discussion

SCMs are uncommon congenital anomalies of the spinal cord and its covering. In 1837 the term ‘diastematomyelia’ was first used by Ollivier [4] to describe an abnormality of spinal cord in which the dura is separated by a bony spur or rigid fibrous bands to create two sleeves, each containing a portion of spinal cord, divided into two parts sagittally. Bruce et al. [1] used the term diastematomyelia to describe a spinal cord split by a midline bony spur, reserving the term ‘diplomyelia’ for a true doubling of the spinal cord without a spur. Feller and Stenberg [3] first implicated a notochordal cleft caused by persistence of a midline cell rest as the cause of diastematomyelia.

Pang [6] advocated a new classification, recommending the term ‘split cord malformation’ for all double spinal cords. They also described the unified theory, which explains the embryogenetic mechanisms of all variants of SCMs. The basic error, according to this, is the formation of an “accessory neurenteric canal” between yolk sac and amnion through the midline embryonic disc, which is subsequently invested with mesenchyme to form an endomesenchymal tract that splits the notocord and neural plate. The location of this abnormal fistula is variable, but it must be rostral to the primitive neurenteric canal, because the primitive pit into which the normal neurenteric canal opens ultimately comes to lie opposite the coccyx. In type I SCM, the precursor (meninx pri-

mativa) cells within the endomesenchymal tract may form a bony spur in the midline, attached to the bone of developing vertebral column, while in type II SCM the mesenchymal tract forms a thin fibrous septum in the space between the hemicords.

Multiple accessory neurenteric canals may form two or more noncontiguous SCMs. One or both types of SCM at different levels have been reported in the same patients [2, 6], as we have seen in our cases 1 and 2, owing to the formation of two mesenchymal tracts at two separate levels. In case 3, the endomesenchymal tract near the ventral aspect of hemicords admixed with the meninx primativa cells and formed a small bony spur and the double dural sacs surrounding the anterior aspect of hemicords, since the meninx primitiva cells first appear close to the ventral aspect of neural tube. A portion of endomesenchymal tract dorsal to the hemicords did not involve meninx primitiva cells and formed a single dural sac and a fibrous septum. The presence of an endomesenchymal tract caused by an accessory neurenteric canal may explain the embryogenetic mechanisms of both types of malformation at the same level in case 3 (Fig. 1). Ersahin et al. [2] have also reported a similar case, with both types of SCMs at the same level.

These three cases are good examples supporting the unified theory proposed by Pang et al. We also think that all variant types of SCMs have a common embryogenetic mechanism differing only in severity and that the formation of an accessory canal is the essential step in all SCMs.

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