Findings that shed new light on the possible pathogenesis of a disease or an adverse effect

Widely separated composite split cord malformation

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SUMMARY

We are reporting a rare case of 18-month-old child with split cord malformations (SCMs) at two different levels, one in the upper thoracic (T2) and the other in the lumbar (L2) region. Only few case reports of such wide separation of SCM are available in the published literature.

BACKGROUND

Split cord malformation (SCM) constitutes approximately one-third cases of spinal dysraphism, characterised by longitudinal splitting of spinal cord into two halves with a bony or fibrous spur. Composite types of SCMs are very rare and are characterised by the presence of two different forms of SCM with intervening normal cord in same patient. These malformations are the consequences of two different foci of ectoendodermal adhesions and endomesenchymal tracts, which ultimately lead to development of different SCM types with intervening normal cord.

CASE PRESENTATION

An 18-month-old male child presented with progressively increasing weakness of right foot since birth. Physical examination revealed pigmented skin in upper thoracic and mid-lumbar region with overlying hypertrichosis (figure 1). Tenderness was noted on direct palpation over these areas. Clubfoot was noted on right side and its power was reduced (grade 3/5). Power was normal in rest of the limbs and sphincter functions were normal. Developmental assessment revealed normal milestones and growth parameters were within normal range.

MRI of thoracic spine revealed low-lying spinal cord tethered with the posterior element of L5 vertebra. Longitudinal split of dural sac and spinal cord due to an anterior sagittal osseous spur was noted at L2 level diagnostic of Pang type I SCM. Cord was reuniting above and below this level.

Duplication of the cord with single dural tube was also noted in upper thoracic region extending from D1 to lower border of D3; however, no bony or fibrous spur could be demonstrated suggesting Pang type II SCM. Syrinx was noted in cord from C6 to its lower end. Spina bifida occulta and bony defects due to failure of vertebral formation (butterfly and hemivertebrae) and segmentation (block vertebrae) were noted at multiple levels (figure 2).

The patient was operated under general anaesthesia. Bony septum was excised using microsurgical technique

along with detethering of filum terminale (figure 3). Dura was closed in a watertight manner using fascia lata. Patient was neurologically stable at 6 months of follow-up.

DISCUSSION

The presence of SCMs at multiple levels is a rare occurrence; seen in less than 1% of total cases of SCMs.¹ Only few case reports of such anomalies are available in the published literature.^{2–4} Most of these cases had two spurs at different levels and one case having two different types of spurs at three different levels.⁵ In most of these cases the spurs were not more than three vertebral levels apart. Only one case had widely separated spurs, present at T2 and L3 levels.⁶ The spur is most commonly found in the lower thoracic or the upper lumbar region and only four case reports of septum above T3 are available in the published literature. ‘SCMs’ were earlier known as a part of occult spinal dysraphism which is defined as skin-covered lesions without any exposed neural tissue or visible cystic mass.⁷ This term was coined to include the forms traditionally called ‘diastematomyelia’ and ‘diplomyelia’ which are the variants of same basic malformation, that is, a common odontogenic error. The first form is characterised by two hemicords residing in separate (hence double) dural sacs transfixed by a midline bony spur. Each hemicord is a true hemicord and hence contains only lateral nerve roots and no paramedian roots.

Figure 1 Pigmented skin in the upper thoracic and the mid-lumbar region.

Figure 2 (A) T2-weighted sagittal image of spine showing low-lying cord and bony spur at L2/3 level. (B) T2-weighted axial image showing two hemicords within single dural tube at D1/2 level, (C) reuniting at D4 level (D) single cord is visible below the level of duplication (E) anterior osseous spur at L2/3 level dividing the cord again into two halves. Syringomyelia is evident in all axial images.

The second form, in contrast, contains double cords in single sac with no midline element. Each half is a complete cord and so contains both lateral and paramedian roots.

The recent classification given by Pang consists of type I SCM containing two hemicords within separate dural tubes divided by an osseocartilaginous median septum and type II SCM containing two hemicords within single dural

tube separated by a fibrous median septum. The embryogenesis and genetics of the disorder had been a subject of conjecture but most of these malformations are well explained by the unified theory of embryogenesis which states that the underlying pathology is formation of adhesion between ectoderm and endoderm that eventually leads to the development of an accessory neuroenteric canal. An endomesenchymal tract develops around this

Figure 3 Operative photographs. (A) Before and (B) after excision of osseous spur (arrow). (C) Low-lying thick filum terminale.

canal which bisects the developing notochord and results in the formation of two hemineural plates. The location of this fistulous tract is variable, but it most likely lies rostral to Hensen's node, since the primitive pit marks the end of primary neurulation and most known SCMs involve cord segment rostral to distal conus. If the endomesenchymal tract ultimately leads to the development of a bony septum, it is called type I SCM and if forms a fibrous band then it is known as type II SCM. These neuroenteric canals may be single or multiple, and if multiple, result in one or more types of SCMs at different levels as in our case.⁸

In 1942, Bremer proposed theory of another neuroenteric canal arising from the perforation of the roof of the primitive neuroenteric canal to explain the existence of rare dorsal intestinal fistulas. This unified theory of embryogenesis challenges the hypothesis that this abnormal neuroenteric canal involves accessory openings from the original notochordal canal, as Bremer⁹ proposed. Emura *et al*¹⁰ and Klessinger reported an experimental animal model of SCM produced by surgical induction of a fistula.¹¹ Even though the results of their study supported the hypothesis that SCMs originate from an accessory neuroenteric canal, it is difficult to explain all variations of SCMs. In view of the above, the central doctrine of unified theory which states that the two types of SCM can never overlap needs to be reconsidered.

CONCLUSION

Existence of a type I malformation, immediately next to a type II lesion (composite SCM) in the same patient lends support to unified theory, but separation of these two by more than 10 vertebral levels is an extremely rare embryological event and can be explained only by the presence of another accessory neuroenteric canal, as proposed by Bremer. However, the site of such accessory canals is a matter of open research.

Learning points

- ▶ Patient presenting with spinal dysraphism should undergo radiological evaluation of whole spine to rule out presence of composite type SCMs.
- ▶ Children with tethered cord should undergo preventive surgery to avoid neurological deficit due to cord stretching with increasing age.
- ▶ Strict follow up is necessary in operated cases of spinal dysraphism as chances of re-tethering are quite high in first few years.

Competing interests None.

Patient consent Obtained.

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