Case Report

Diastematomyelia with hemimyelomeningocele: An exceptional and complex spinal dysraphism

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ABSTRACT

Variations in split cord malformation (SCM) have been described earlier. However, a true hemimyelomeningocele (HMM) as only congenital malformation is extremely rare and is reported infrequently in published literature. We are reporting the case of a 3-month-old girl child who presented with a swelling on the lower back since birth. Magnetic resonance imaging revealed a type 1 SCM with right hemicord forming a HMM. Precise diagnosis and thorough anatomical detail of dysraphism is essential for optimal, individualized neurosurgical management.

Key words: Diastematomyelia, hemimyelomeningocele, spinal dysraphism, split cord malformation type 1

Case Report

A 3-month-old girl child presented to us with a history of swelling on back since birth. She had a history of full term normal vaginal delivery with no significant family history. Her developmental milestones were normal for age. On neurological examination there was complete flaccid paralysis of right lower extremity with normal anal tone.

On local examination, swelling was soft, cystic, and tensile with positive fluid thrill. Neural elements were visible on translucency examination. An initial diagnosis of a myelomeningocele was made and patient was referred for magnetic resonance imaging (MRI) of neuraxis.

MRI spine revealed a bony spur at D11 level dividing the spinal cord into two halves suggesting Pang type 1 split cord malformation (SCM) or diastematomyelia. Right hemicord, along with meninges and cerebrospinal fluid was herniating through the defect in posterior element of same vertebra forming a large hemimeningomyelocele [Figures 1 and 2]. Both hemicords were reuniting below the level of herniation.

Based on clinical examination and MRI a final diagnosis of SCM type 1 with right hemimyelomeningocele (HMM) was made and patient was prepared for surgery.

Surgery was carried out through a mid-line skin incision. Excision of bony spur along with microneurosurgical dissection and meticulous layered closure of myelomeningocele was achieved. Patient was discharged on eighth postoperative day without any additional neurological deficit.

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**Discussion**

According to Pang’s unified theory of embryogenesis,[1] the whole spectrum of split cord syndrome originates from one basic ontogenic error occurring around the time of primitive neurenteric canal closure. This basic error is the formation of “accessory neurenteric canal” between the amniotic cavity and the yolk sac. This abnormal fistulous connection bisects the notochord and the neural plate on approximately postovulatory day 18. Each hemineural plate undergoes neurulation in order to its own heminotochord. Final appearance of matured SCM depends on the ability of the embryo to heal around this abnormal fistulous connection aka endomesenchymal tract. According to Pang et al., formation of endomesenchymal tract before postovulatory day 21 gives rise to SCM type 2 or diplomyelia, whereas formation of this tract after postovulatory day 30 gives rise to SCM type 1 or diastematomyelia. The formation of this tract between postovulatory day 21 and 30 gives rise to composite/mixed SCM.[1,2]

Each hemineural plate undergoes neurulation in which, the neural folds converge and fuse each other in relation to their respective hemi-notochords. This process of primary neurulation occurs between postovulatory days 22–28. In case of neurulation defect on one side, HMM or hemimeningocele may occur. Our patient has hemimeningocele on the right side.

Myelomeningoceles are frequently associated with Chiari malformation[3] diastematomyelia (up to 45% in some reported series)[4] and other congenital vertebral defects such as spondylocostal dysostosis,[5] Jarcho-Levin syndrome, Klippel-Feil anomaly and a wide range of syndromic malformation complexes, but true hemimeningocele, where a single hemicord fails to neurulate is rare.[6,7] The neurological deficit in children with HMM is less severe than compared to myelomeningocele patients and is limited only to the side of exposed hemicord. Our patient has flaccid paralysis on the right side.

**Conclusion**

This case report highlights the importance of detailed imaging of complete neuraxis in cases of spinal dysraphism for optimal neurosurgical management. Second the rarity of this
condition suggests that formation of the endomesenchymal tract and neurulation defect are two separate embryological events, and their coexistence at the same level is only by chance though more inputs are warranted in this regard.

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Conflicts of interest
There are no conflicts of interest.

References