Split cord malformation

Raj Kumar* and Suyash Singh

Department of Neurosurgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India

Split cord malformation is one of the rare occult dysraphisms. The exact incidence is difficult to quote but in our experience, we find the incidence of 33%, slightly more than reported in other series. With advent of different MRI protocols and increasing diagnostic armamentarium, the incidence continues to rise.

In 1992 Pang revolutionized the concept and nomenclature of split cord malformation. His unified theory of embryogenesis unravelled the long-lasting conundrums in understanding why some septum is bony and other fibrous. After this concept, words like "diplomyelia" and "diastematomyelia" became history. According to Pang's unified theory, persistent accessory neuroenteric canal leads to formation of septum. Actually, it was Bremer et al. [1] who first proposed the term accessory neuroenteric canal. He said that the failure of the ectoderm-endoderm and cleft notochord favours the formation of neuroenteric canal. Later on, Pang added concept that the failure of "anlagen" to integrate in midline and formations of endomesenchymal tract contribute to septum in split cord malformation. Further, incorporation of "menix primitiva" into the tract leads to bony septum and non-incorporation leads to fibrous septum.

Besides type of septum, the surrounding thecal sac and possibility of whether lamina is fused or hypertrophied is to be kept in mind. In SCM type 1 there are two dural sleeves with or without fused lamina. In SCM type 2 there is only one dural sleeve. In our experience of 16 cases, we described that the patients with SCM are rarely associated with meningoymelocoele and their clinical, embryological and prognosis profiles differ from those who are not associated with same. We coined the term "Complex spina bifida" for SCM associated with meningoymelocoele highlighting the fact that they deserve special attention in order to fetch an optimum outcome. Mahapatra et al. [2] has modified the Pang's classification further into type 1a-d for SCM associated with meningoymelocoele and the clinical, embryological and prognosis profiles differ from those who are not associated with same. McLone unified theory further highlights the existence of rare exceptions to pangs embryological hypothesis like dorsal spur. To add on Pang's classification, we proposed a sub classification into type A and B on basis of whether split cord malformation (SCM) is associated with meningoymelocoele or not. This classification highlights the importance of pure or combined occurrence. Pang and Ersahim have also pointed on high incidence of "menix primitive" to extradural space. The main problem of Split cord malformation arises due to tethering of cord either at level of spur or below it. Tethering can be at a single level or multiple level. We defined the entity "spina bifida multiplex" for patients with more than two level tethering. In our series of 41 patients, we showed that these patients of spina bifida multiplex behave in different way and need separate place in classification system because the prognostication and outcome also depends on such associations.

We investigate all patients of spinal dysraphism with cranial and spinal MRI, chest Xray, Ultrasonography, ECHO screening, urolflowmetry and post voidal residual volume as our departmental standard policy wherever possible. CT Spine is done for better bony delineatio for cases of bony split cord malformation. Magnetic resonance imaging is the most common and the investigation of choice for split cord malformations. The spur may be isointense or slightly hyperintense compared to CSF on T1-weighted images. Bony, cartilaginous and fibrous spurs all appear hypointense on T2 weighted images. The osseous spurs can be missed on T1-weighted images.

Surgery is the treatment of choice wherein the bony spur should be excised microsurgically. All the associated tethering lesions like arachnoid bands, adhesions, aberrant nerve roots, thick filum terminale, etc must be thoroughly studied and dealt with. Repairing the dura in most meticulous manner in order to re-construct the normal anatomy is a very important step. Cutting of fibrous septum should be started with distal most point and complete excision is thumb rule. Bony septum is difficult to deal with especially when spur is dividing at certain levels and going high up. We usually extend our laminectomy to have a proper exposure. Mahapatra et al. [2] has also emphasized on use of drills for dorsal spur.

In conclusion, the understanding of embryology, thorough investigations, study of associated lesions, and use of recent concepts

Correspondence to: Raj Kumar, Professor and Head of Department, Department of Neurosurgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India, E-mail: rajkumar1959@gmail.com

Received: March 10, 2017; Accepted: March 20, 2017; Published: March 22, 2017
and classification in dealing split cord malformation are key modulators for better surgical results and may vicissitude the outcome. Early surgical intervention even in asymptomatic presentation has shown to improve long term outcome.

References

1. Bremer JL (1952) Dorsal intestinal fistula; accessory neurenteric canal; diastematomyelia. *AMA Arch Pathol* 54: 53–54

2. Borkar SA, Mahapatra AK (2012) Split cord malformations: A two years’ experience at AIIMS. *Asian J Neurosurg* 7: 56-60. [Crossref]