SPLIT CORD ASSOCIATION

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ABSTRACT

Split cord malformations (SCMs) are relatively rare forms of occult spinal dysraphism and tethered spinal cord syndrome. The majority of these cases present with neurocutaneous stigmata, we present a case report of split cord malformation associated with diaphragmatic hernia.

Key words: split cord malformation, ventral split in cord, diaphragmatic hernia

Introduction:

Split cord malformations (SCM) are spinal cord defects having double neural tube due to vertical split in the cord. SCM is one of the commonest subset of occult spinal dysraphism (1).

This splitting in the spinal cord is physically associated with an osseous, cartilaginous, or fibrous spur or septum that originates from the vertebral body and can extend to the posterior elements (2, 3).

Case Report:

A new born baby boy, seen at the age of 14 hrs delivered at full term by NVD with birth weight of 3.760 kg to a 29 year old mother with normal antenatal USS.

After birth the baby became tachypneic, grunting, cyanosed, immediately intubated and connected to a mechanical ventilator with normal initial blood gases, pH: 7.324, PO2: 40.2mmHg, PCO2: 46.7 mmHg, HCO3: 23.6mmol/L.

General examination of the baby was normal, no pallor, no dysmorphic features and no skin lesions.

Systemic examination shows decreased air entry on the right side of the chest, scaphoid abdomen, male external genitalia and bilateral impalpable testicles, normal both lower limbs, normal neurological examination.

Chest x-ray revealed bowel gas shadow at the right hemithorax and bifid dorsal spine. (Fig. 1)

The baby underwent surgery and a mid-ileal loop was seen herniating through a defect in the root of the mesentery. The reduction was difficult and some tethering of the loops was found, with intestinal tears, necessitating resection and anastomosis. The defect in the mesentery was repaired. No defect was seen in the diaphragm.

MRI showed spinal dysraphism in the form of upper dorsal scoliosis, spina bifida occulta of C7 down to D6, butterfly D3, D4 and D5 vertebrae, splitting of the upper dorsal cord with no MRI evidence of bony or connective tissues and finally anterior thoracic meningocele (Fig. 2, 3).

The baby had smooth postoperative period and discharged home in good general condition. He had bilateral orchidopexy at the age of 18 months. The parents refused any prophylactic neuro-surgical intervention.

Discussion:

A number of spinal cord malformations involve concomitant anomalies of the gastro-intestinal system, posterior mediastinum and/or retro-peritoneum.

The key to understanding these malformations lies in properly appreciating the embryonic relationship between the developing neural tube and the endoderm during early development (4).

It is now well accepted that all double spinal cord malformations or SCM probably arise from a common embryoge-
netic mechanism (5).

Pang and co-workers have postulated that the participation of certain meningeal precursor cells within an errant midline ecto-endodermal adhesion tract during early gastrulation determines how SCMs can be classified into two radically different types (6).

Type I SCM consists of two hemicylops contained within separated dural tubes, separated by a bone or osteocartilaginous septum that extends from the vertebral body to the neural arches. The rigid median septum is therefore entirely extradural (4,6).

Type II SCM, the hemicylops reside within a common dural tube. There is no rigid median septum (4,6).

These main features of the two types of SCM never overlap, and the classification can be readily made by preoperative neuro-imaging studies (6).

In type I SCMs, the transfixing, blade like bone septum was as deleterious to the hemicylops and the necessity of its removal was seldom disputed. On the other hand it took some convincing to show that the hemicylops in many cases of type II SCM were also severely tethered by taut fibrous bands and septa that are attached to the common dural sac.

Considering the original endomesenchymal tract began as a median structure conjoining the endoderm and ectoderm, traversing all the elements that later form in the spinal canal, it is hardly surprising to find that these mesendyme-derived fibrous bands sometimes retain ventral attachment to the hemicylops.

Indeed, Pang, had found more examples of purely ventral intra-dural bands in type II SCMs, as well as dorso-ventral bands that bisect the entire sagittal plane of the dura, even, he found cases in which the ventral mesenchymal band extended beyond the ventral column to the extra-luminal tissues of the gut (endoderm), resulting in characteristic gastro-intestinal malformations (6).

In fact the above findings completely explain the association of the presence of bowel loops in our case in the right chest due to ventral tethering of the menenchymal bands extending beyond the vertebral column to the extra-luminal tissue of the gut.

Thus, despite the fact that MR imaging and CT myelography often do not demonstrate these thin but medlesome fibrous septa, Pang and others have recommended the surgical exploration of all type II SCMs (7, 8, 9).

To my knowledge this is the second case of SCM type II with diaphragmatic hernia association and the first case of survival (6).

Conclusion:

We recommend prophylactic surgical exploration of cases with Type II SCM as computerized myelography has less than 50% accuracy in revealing the ventral band especially in the presence of intestinal malformation.

Figure 2, 3.

MRI - spinal dysraphism in the form of upper dorsal scoliosis, spina bifida occulta of C7 down to D6, butterfly D3, D4 and D5 vertebrae, splitting of the upper dorsal cord

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**REFERENCES**