



Split Cord Malformation – Illustration and precautions in surgical technique and complication avoidance

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General Note



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ABSTRACT

Descriptions of split cord malformation (SCM) appeared in the medical literature as early as the 17th century (Barkovich, 1990; Bademci, 2006). Several terminologies were in vogue in literature like “diastematomyelia” and “diplomylelia” with a poor distinction. In 1992, Pang and colleagues proposed a unified theory attributing both type I and type II SCM to a single error in embryogenesis: adhesion between the ectoderm

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and endoderm leading to a persistent neurenteric canal (Beardmore et al, 1958; Pang et al, 1992). Increasingly SCM is being recognised as a cause of Tethered Cord Syndrome (TCS). We present an interesting case of SCM in a male infant, who presented to our institute as a part of medical tourism in India and discuss briefly its clinical presentation, illustration of surgical technique and avoidance of complications.

Keywords: Split Cord Malformation, Neural tube Defect, Tethered Cord Syndrome, Diastometamylia, Diplomyelia, Occult Spinal Dysraphism.

1. INTRODUCTION

SCMs are a form of occult spinal dysraphism that share common embryonic etiology and is being increasingly recognised as a cause of TCS. The sine qua non of these developmental anomalies is the presence of a longitudinal cleft within the spinal cord across one or more vertebral segments. Type I malformations (formerly diastematomyelia) are characterized by a bony septum that cleaves the spinal canal in the sagittal plane and a duplicated thecal sac. Type II malformations (formerly diplomyelia) are characterized by a cleft cord within a single dural sac, often tethered by a fibrous midline septum to the adjacent duramater (Beardmore et al, 1958; Pang et al, 1992).

Pang et al. propagated "Unified theory of embryogenesis" to explain the pathogenesis of SCM. The basic error appears to be development and persistence of accessory neurenteric canal (ANC) (Pang et al., 1992). Abnormalities of anterior end of this canal will result in intestinal malformations and of the posterior end in cutaneous malformations, whereas the persistence of the intermediate part causes a split in the notocord and the neural placode. The division of the notocord then leads to segmentation and/or fusion anomalies of vertebral bodies or posterior elements. The division of the placode later on leads to the formation of two hemicords. Presence or absence of precursor meningeal cells in the mesenchyme surrounding ANC will result in type I or II SCM (Table 1). The persistence of the ANC could occasionally interfere with the neurulation process leading to the formation of a meningocele or meningomyelocele, compounding these malformations further as depicted in figure 1 (Pang et al, 1992; Bremer, 1952).

Gupta and Mahapatra have further classified type I SCM into 4 subtypes based on the location of the spur in between the proximal and distal parts of the split and space available above and below the spur (Table 1). They attach prognostic significance and different surgical approaches based on the above classification (Mahapatra, 2005; Gupta 2006).

2. CASE PRESENTATION

1 year old male infant, resident of Nepal, was brought at our OPD by his parents with complaints of gradually progressive painless, soft, palpable swelling in the lower back since birth. This was accompanied by gradually progressive bending of the spine towards the right side along with outward turning of both feet. The patient was unable to stand and walk by 1 year of age. His elder siblings had achieved the locomotor milestones appropriate to their ages. His was a normal, full term, vaginal delivery at home with a normal cry at birth. There was no history of trauma, fever, continued dribbling of urine or urinary retention seizures, radiation exposure or significant drug intake by the mother during antenatal period.

General examination was essentially normal. Local examination revealed a soft, non-tender, non-pulsatile, non-fluctuating, and non expansile, non-transilluminating swelling in the lower back against the level of L3 spine. There was dextrosciosis of spine corresponding to the level of subcutaneous swelling along with bilateral valgus deformity of feet. Neurological examination revealed diminished spontaneous movements of the lower limbs and the patient was unable to bear his weight while attempting to stand. Cranial nerve examination, sensory and motor examination of the upper limbs were normal. Deep Tendon Reflexes (DTRs) in both lower limbs and bilateral planter reflex was absent.

Plain Magnetic Resonance Imaging (MRI) of dorsal, lumbar and sacral region revealed dextrosciosis of spine with occult spinal dysraphism in dorsolumbar region with vertebral segmentation and fusion anomalies. Lipomyelocele was noted in the upper lumbar region with tethered cord and type Ia SCM (Figure 2). Non Contrast Computed Tomography (NCCT) scan of head and workup of other systems along with routine pre-operative investigations did not reveal any abnormality.

Based on the patient's clinical and radiological workup the patient was taken up for surgery on therapeutic and prophylactic grounds.

General anaesthesia with endotracheal intubation was administered and patient was placed in prone position. After antiseptic painting and draping a vertical midline incision on the back was made centred over the lipoma at L2L3 with extension of incision two level cranial and caudal to the lipoma. The tip of the lipoma in the subcutaneous plane was found piercing the deformed lamina (Figure 3). Subperiosteal dissection of the muscles was done followed by laminectomy one level above and below the attachment of dorsal septum. The lipoma was excised followed by removal of the osseo-cartilagenous septum by bone nibbler and fine kerrison punch till the hard bony base of the spur was reached. Further excision of base of the spur was achieved by high speed diamond drill (figure 4). Lipoma and the excised spur were sent for histopathological analysis. Dura was opened in a curvilinear manner and incision was extended 2 levels cranial and caudal to the attachment of spur. Detethering was performed and free movement of the cord was ensured. This was followed by suturing of the dural sleeves dorsally in a watertight fashion and layered closure of the incision.

No Cerebrospinal Fluid (CSF) leak was noted in the postoperative period with no sensory or motor deterioration as compared to preoperative status. Alternate and complete suture removal was done on postoperative day 7 and 10 respectively. The patient was discharged on postoperative day 10 with advice for physiotherapy and orthopaedic consultation and regular monthly follow up. The patient turned up for follow up 6 months later at our OPD and was able to stand and walk with minimal support. Thereafter, the patient did not turn up and was lost to follow up.

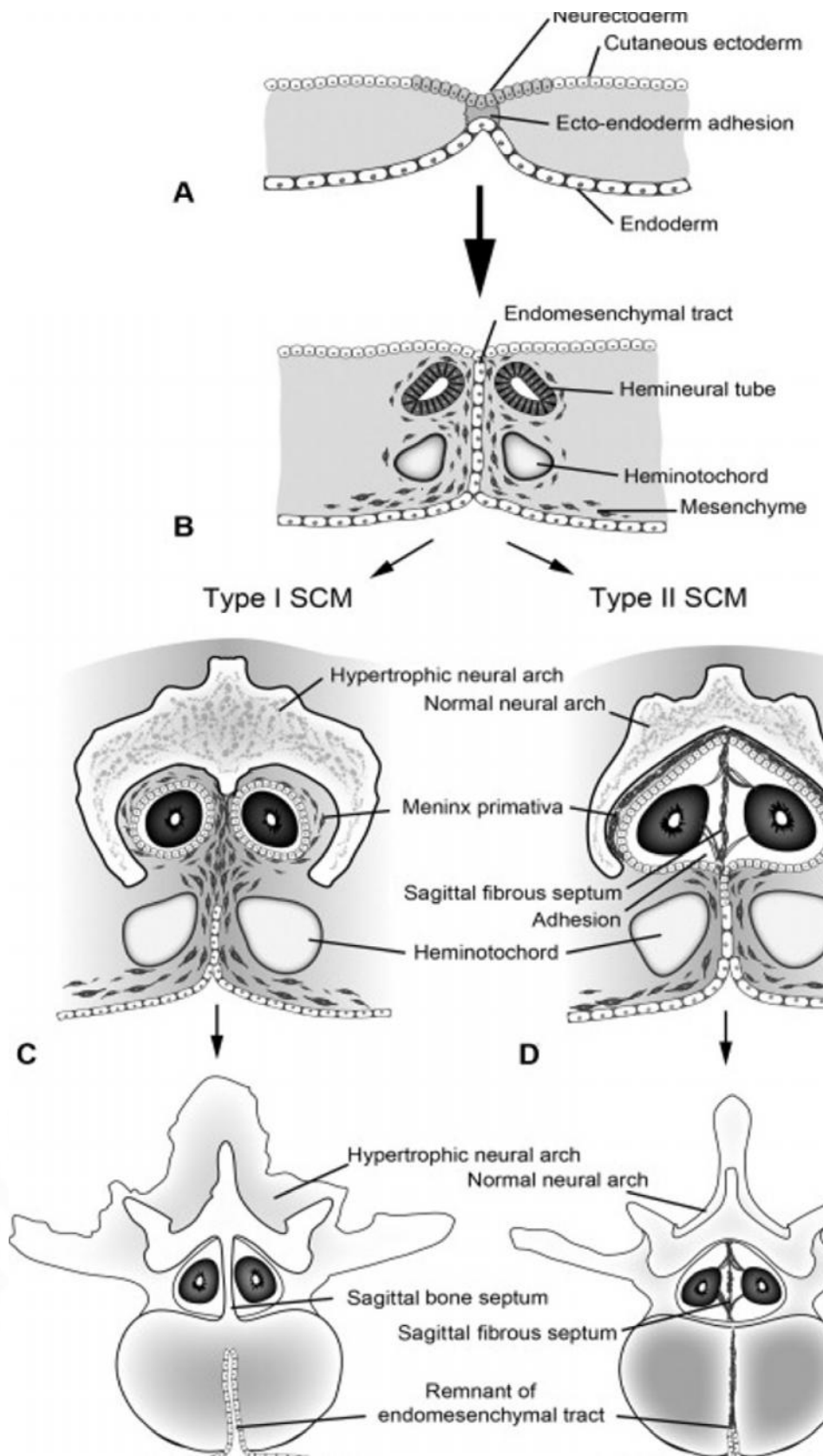


Figure 1 Embryogenesis of SCM

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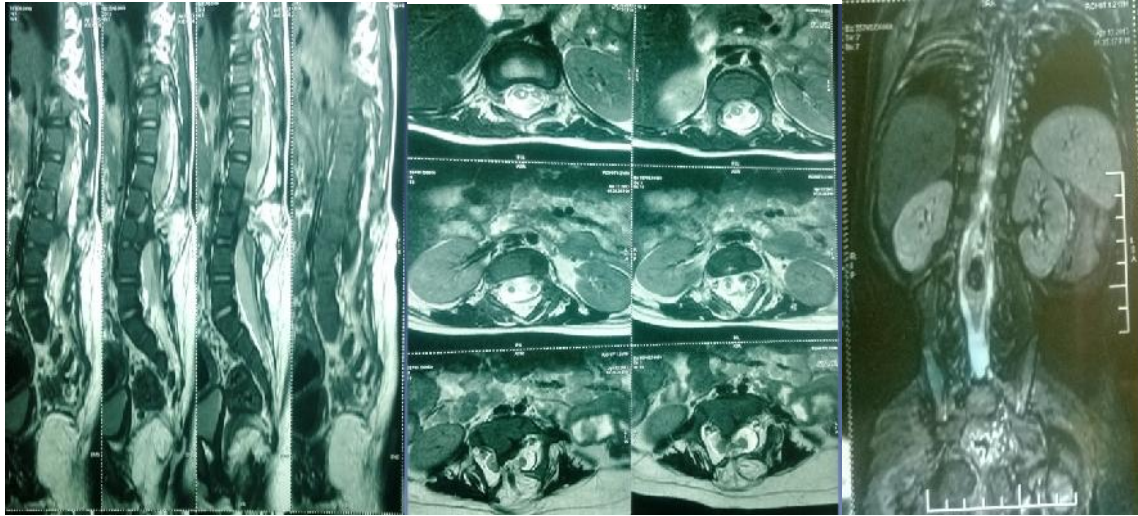


Figure 2 Sagittal, axial and coronal sections of plain MRI

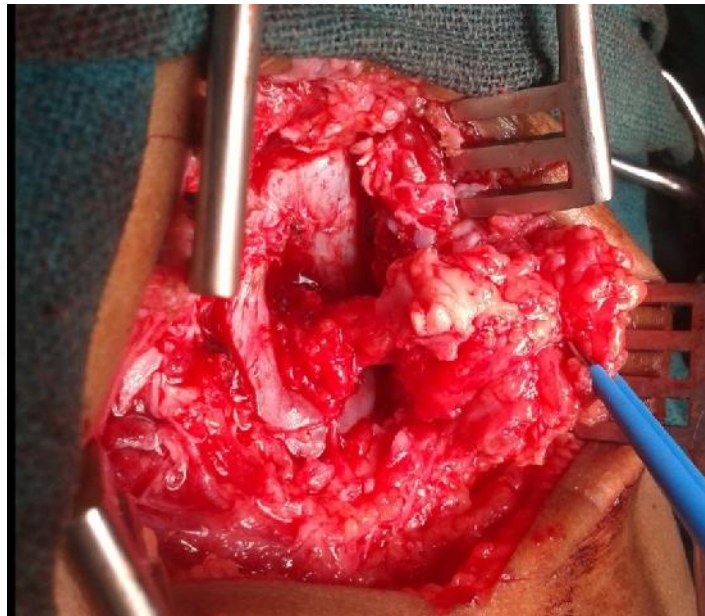


Figure 3 Subcutaneous lipoma, dorsal spur and split cord in view



Figure 4 Lipoma and spur has been excised. Split cord in view

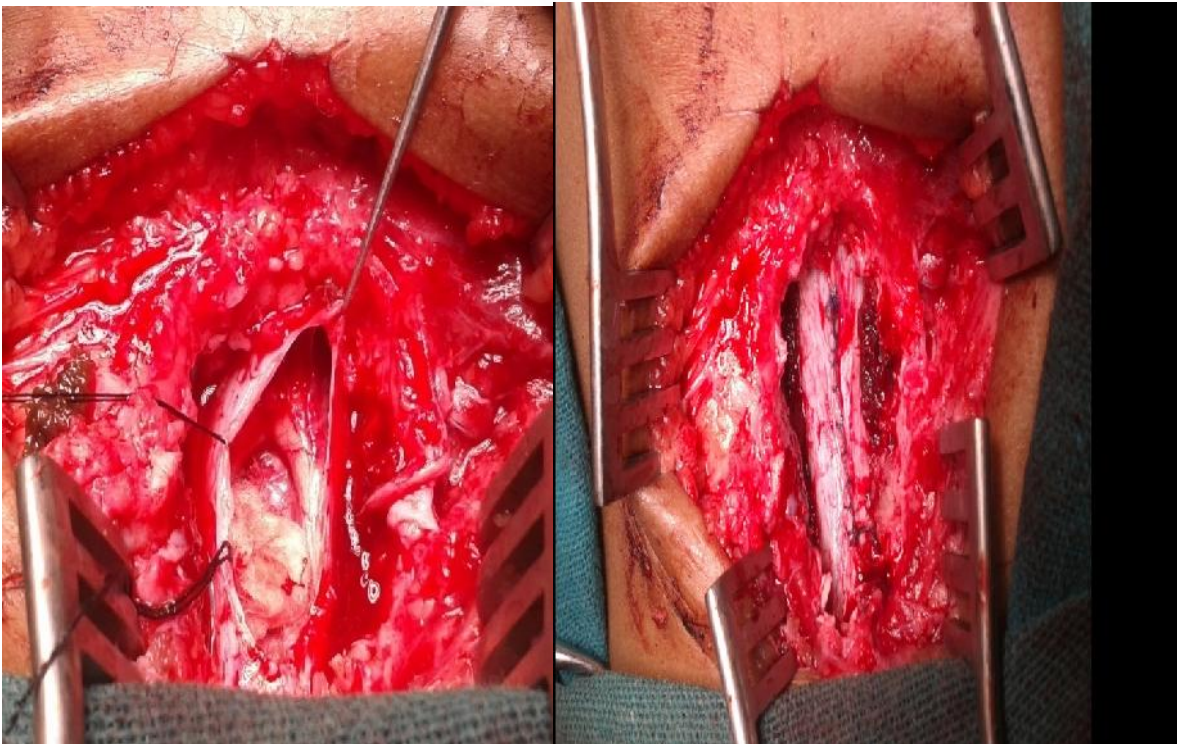


Figure 5 Curvilinear durotomy followed by watertight dorsal dural closure

Table 1 Distinction between SCM type 1 and 2

TYPE 1 SCM	TYPE 2 SCM
Two hemicords with its own central canal and surrounding piamater	Two hemicords in a common central canal
Separate dural tubes	Single dural tube
Rigid osseocartilaginous median septum	Non rigid fibrous septum
Spinal deformities common	Usually no spinal deformity
Orthopaedic deformities common	Uncommon
SUBCLASSIFICATION OF TYPE 1 SCM	
Type 1a	Bony spur in the centre with equally duplicated cord above and below the spur
Type 1b	Bony spur at superior pole with no space above and a large duplicated cord lower down
Type 1c	Bony spur at lower pole with a large duplicated cord above
Type 1d	Bony spur straddling the bifurcation with no space above or below the spur

3. DISCUSSION

Patients with SCM may be asymptomatic, with only cutaneous stigmata of their spinal dysraphism (Anderson, 1994). Our case had a subcutaneous lipoma in the lumbar region, reported in 2-25% of cases (Anderson, 1994; ASPN Pediatric Neurosurgery, 2001) and presented with motor weakness of both lower limbs, reported in 21% of patients. Bilateral talipesequinovarus present in our case is reported to occur in 17-20% of patients (Anderson, 1994; ASPN Pediatric Neurosurgery, 2001).

Location of SCM in our case was lumbar and the same is reported to be the commonest site in literature followed by dorsolumbar region (Freeman, 1967; Ersakin, 2002). Rarely split may be seen in cervical and sacral region (Andro et al., 2009). Spur causing split in the cord are largely ventral in location, as in our case. However, rarely, dorsally situated spurs have also been reported (Chandra et al., 1999).

Plain MRI of the spine is the investigation of choice which shows tethering of the cord, position and thickening of the filum and associated anomalies. Mahapatra recommends screening of entire spine by MRI rather than limiting to the region of split with cutaneous stigmata as cases of multisite dysraphism, though rare, may otherwise be missed. Alongside, Plain MRI of Craniovertebral Junction (CVJ) may be done to exclude Chiari malformation. NCCT spine and CT myelography along with plain X Rays may be required to assess the scoliotic component local vertebral anomalies of the disease (Venkatraman, 2005).

Treatment of SCM encompasses multidisciplinary approach including neurosurgery, orthopaedics, physiotherapy and occupation therapy. The goal of surgery is removal of the fibrous or bony septum, resection of any other local spinal cord attachments causing tethering, exploration for associated tethering-related anomalies such as dorsal tethering bands or thick filum, which can be seen in the majority of patients, and excision of spur along with detethering of the cord. Opinion varies in regards to fusion of scoliotic deformity with some neurosurgeons opting to correct the spinal deformity in the same setting while others opt for neurological correction first followed by correction of bony deformities of the spine subsequently (Venkatraman, 2005; Mahapatra, 2011). We preferred cord detethering first with plans to excise and fuse the scoliotic segment in subsequent follow ups.

Few points are mentioned in order to avoid complications during the procedure.

1. As mentioned above, screening of entire spine may be advisable to exclude the possibility of multisite dysraphism or tethered elements above and below leading to failure in improvement/progression of symptoms after single site correction.
2. Careful intraoperative assessment of the attachment of spur is to be done as rarely the spur may be dorsal which increases the chances of cord laceration when excised indiscriminately.
3. Laminectomies one level above and below the attachment of spur should be achieved followed by careful excision of the spur to minimise damage to the cord.

4. Fine kerrison punch may be preferred while excising the tip of the spur, specially in pediatric patients. Drill can be used to carefully shave off the base of the spur as the base is firmly attached to the vertebral body.
5. In case drill is used, care must be taken to avoid cottonoids getting tangled in the drill bit causing traction or avulsion of the roots or cord.
6. Aggressive traction over the cord or roots is to be avoided while drilling the base of spur.
7. Watertight dural and layered wound closure is ideal for minimising post-operative CSF leak.

4. RESULT

Our patient turned up 6 months post surgery for follow up and was able to stand and walk with minimal support. Degrees of scoliosis, valgus deformity were unchanged compared to the pre-operative state. He was subsequently lost to follow up before spinal and corrective/rehabilitative orthopaedic interventions could be made.

5. CONCLUSION

SCM is a rare pathology. It becomes particularly challenging in present scenario due to lack of awareness and late presentation as most cases reach the neurosurgeon well after neurological deficits have set in. Associated anomalies make the overall management challenging with respect to therapeutic and rehabilitative interventions. Detethering of the cord is attempted even in asymptomatic cases as progressive neurological deterioration is the norm in the absence of surgery or may deteriorate suddenly due to trivial or accidental trauma or stress.

DISCLOSURE STATEMENT

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