Complex Spinal Dysraphism Complicated by Intramedullary Dermoid

Sedain G[1], Sharma MR[2], Shilpakar S[3]

Abstract

Spinal dysraphism presents in myriad ways. Meningomyeloceles presenting with Split cord malformations are termed as complex dysraphisms. We report a case of myelomeningocele and a split cord with intramedullary lesion making it a more complex type of dysraphism.

Key words: Meningomyeloceles, Split cord, intramedullary lesion

Introduction

Over the past decades, there has been a significant decrease in Spinal dysraphism in developed countries. However, in an underdeveloped country like Nepal it still continues to be a major pediatric neurosurgical problem. Spinal dysraphism presents in various ways ranging from occult to large dorsal lesions.

It is not uncommon to find dysraphism associated with multiple orthopedic abnormalities. The dysraphic spine commonly has tethered spinal cord with variable syrinx. We came across a child who presented with a large mass in the back with normal neurological findings. On MRI scan we found meningomyelocele with a split cord and an intramedullary lesion. She was managed accordingly.

The Case

A six year female child presented to us with a swelling in low back since birth. No similar complaints were there in siblings. She had normal neurology. On examination; she had a cystic skin covered swelling in the upper lumbar region. She had no neurological deficits and no deformities. MRI revealed a meningocele communicating with the intradural space at D12 and additionally split cord and an intramedullary lesion at D9-D11 (Fig 1a, 1b, 1c). The cord was low lying .CT spine revealed a Pang Type 1 split (bony spur) at L2 level (Fig 2).

Excision of the meningocele together with removal of the bony spur was done. The intramedullary dermoid was excised and the dural sacs were converted into a single sac (Fig3). The child developed mild post operative weakness in her left foot which improved with physiotherapy.

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Discussion

The unified theory of embryogenesis proposes that all variant types of SCMs (Split cord malformations) have a common embryogenetic mechanism. Basic to this mechanism is the formation of adhesions between ecto- and endoderm, leading to an accessory neurenteric canal around which condenses an endomesenchymal tract that bisects the developing notochord and causes formation of two hemineural plates[1].

If the dividing septum is bony, it is termed as type I split (formerly Diastematomyelia) and if it is a fibrous septum, it is termed as Type II split (formerly diplomyelia)

Meningomyeloceles are due to failure of fusion of the neural tube during 3rd to 4th week (day 21 to 27) of gestation.

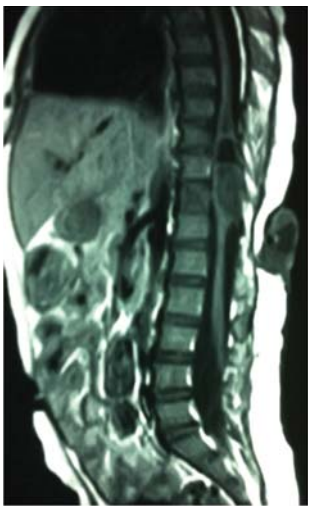

Fig 1a

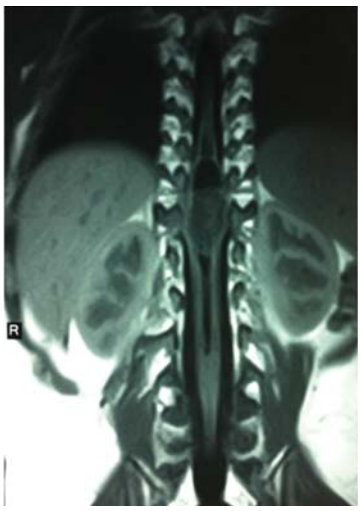

Fig 1b

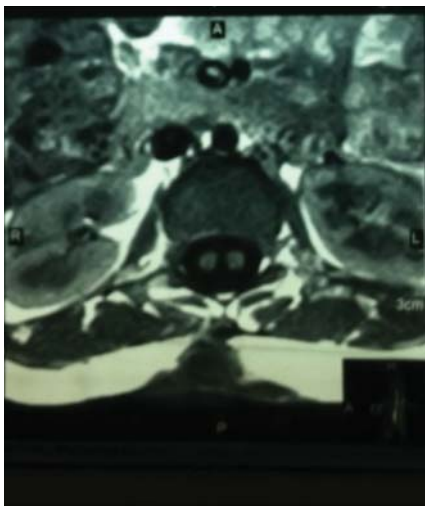

Fig 1c

Fig 1a: Sagittal T1W MRI images showing intramedullary lesion with meningomyelocele and low lying tethered cord
Fig 1b: Coronal image showing intramedullary lesion with split cord below it
Fig 1c: Axial image depicting the double cord forming an owl eye appearance

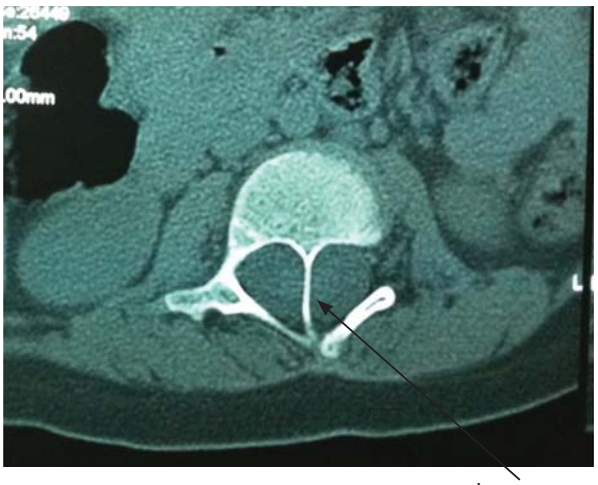

Fig 2: CT axial image depicting the split in spinal canal due to a bony spur (Pang Type 1 SCM)

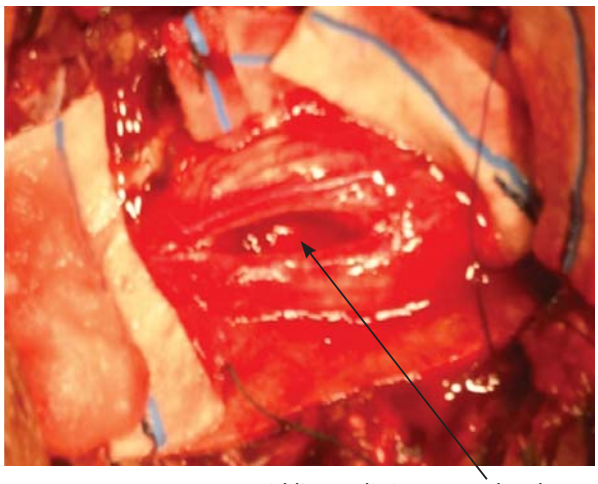

Fig 3: Intra operative photograph showing double dural sacs

The dermoid cysts are developmental in origin and arise from the nests of embryonic ectoderm which get buried or trapped under the lines of fusion of the ectodermal folds in the developing embryo. Combination of all these type of embryological errors together are not very common.

Dorsal cord is the most common site for SCM in 38.8% patients, followed by lumbar region in 28.6%, dorsolumbar in 22.4% lumbosacral in 4.1% and cervicodorsal region in 6.1% cases[2]. In 28.6% patients SCM is found at 2 to 3 level higher than the associated MMC sac, whereas in 12.2% children SCM and MMC are encountered at the same level[2].

The presence of combination of spilt cord and meningomyelocele has been coined as complex spina bifida[3]. In a series of 106 patients with split cord 16 had meningomelocele(15%)[3]. Borkar et al reported 6 MMC in their series of 53(11%) split cord malformations[4]. Similarly, Ansari et al reported 10% association between split cord and MMC[5].

Conclusion

Complex spinal dysraphism should always be suspected in a patient with meningomyeloceles. Microsurgical management with de-tethering and formation of a water tight single dural tube is the goal of surgery.

References

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