

Lumbar Diastomatomyelia with Syringomyelia: A Case Report

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ABSTRACT

Diastomatomyelia with syringomyelia is a rare clinical condition where there is a duplication of spinal cord. We recently came across a child who had this anomaly. The case was successfully managed with an uneventful recovery. We describe this patient in detail in addition to reviewing the pertinent literature.

Keywords: *Diastomatomyelia, syringomyelia*

INTRODUCTION

Diastomatomyelia (DM) is a clinical condition in which the spinal cord is split into two portions by either a bony or a fibrous spur. Depending on the type of dural involvement, it is divided into type 1 and type 2. We recently came across a 9-month old child with a lumbar DM with syringomyelia who was managed successfully at our institute. We discuss the management of this patient in detail along with the description of its etiology, embryology, neurological manifestations, relevant investigations, and treatment implications to both patients and parents. To the best of our knowledge, this is the first case report of DM from Nepal in an infant with minimal symptoms.

CASE REPORT

History

This 9-month-old male child was referred to our institute with a history of a vague lump in the lumbar region since birth. There was no history of urinary dribbling or fecal incontinence. The mother had routine antenatal checkup and had normal vaginal delivery of the child at term. The remainder of the history was not contributory.

Physical Examination

General physical examination was normal with a weight of 9 KG. The child was playful. Neurologically, the child moved all limbs actively and symmetrically. Local examination of the back revealed a vague hard prominence in the region of L₁-L₃ vertebrae with no impulse on coughing. Systemic examination of the child was normal.

Management

A magnetic resonance imaging (MRI) of the spine and brain was obtained which showed a constellation of findings at the lumbar region. The cord was split into two halves longitudinally for approximately 3 centimeters at the level of L₁-L₃ vertebra by a large bony septum. A separate dural sleeve covered each split cord. The cords united at the L₃ level. There was associated syrinx of approximately 2 centimeters in length predominantly on the right portion (Figure 1).

After discussing the benefits, risks and alternatives of different modalities of treatment with the family and after obtaining the informed consent, the child underwent surgery. The area of interest was exposed. Intraoperatively, there was gross splaying of posterior vertebral elements of L₁-L₃

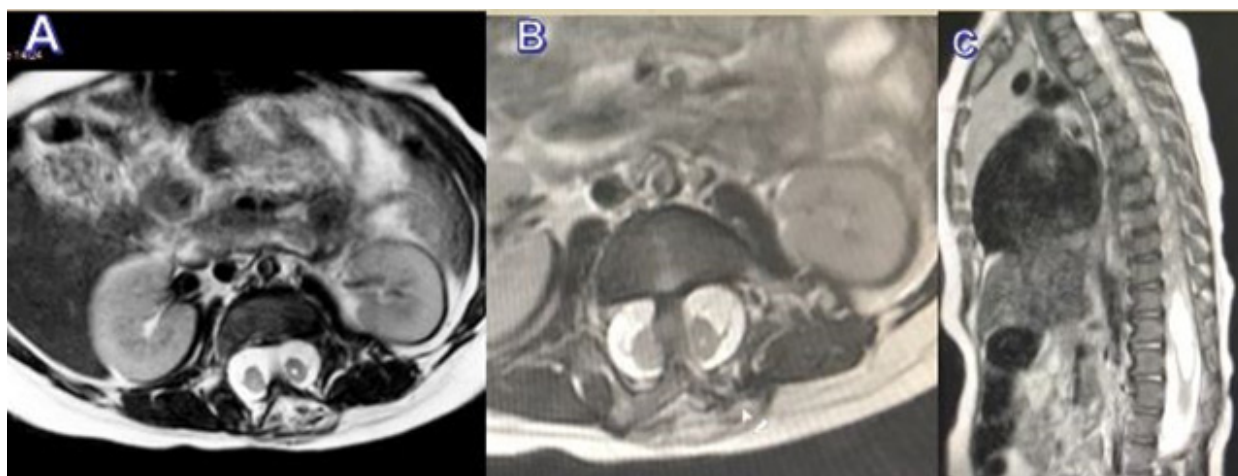


Figure 1. MRI of dorsolumbar spine of the patient: Axial (A, B) and sagittal(C) sections, T2 weighted images, showing split cord with separate dural sleeves (arrow), complete bony septum (double arrows) and prominent syrinx (triple arrows).

vertebrae. A complete midline bony septum separating the two halves of the cord each with a separate dural envelope (Figure 2A) was found. This was resected flush with the posterior part of L₂ vertebral body and dura was cut open posteriorly. After careful mobilization of the split cords (Figure 2B), the two posterior edges of each side of the dura were closed water tight with a delayed absorbable suture enclosing the two halves of the cord inside one dural 'tube' (Figure 2C). No procedure was performed for the associated

syrinx.

The patient tolerated the surgery well. There was no neurological deficit postoperatively. Skin sutures were removed on day 9 and he was discharged home in a stable condition on day 10. On follow up at 6 months, the child had normal growth and development.

DISCUSSION

DM, also known as split cord malformation is a rare congenital spinal cord anomaly

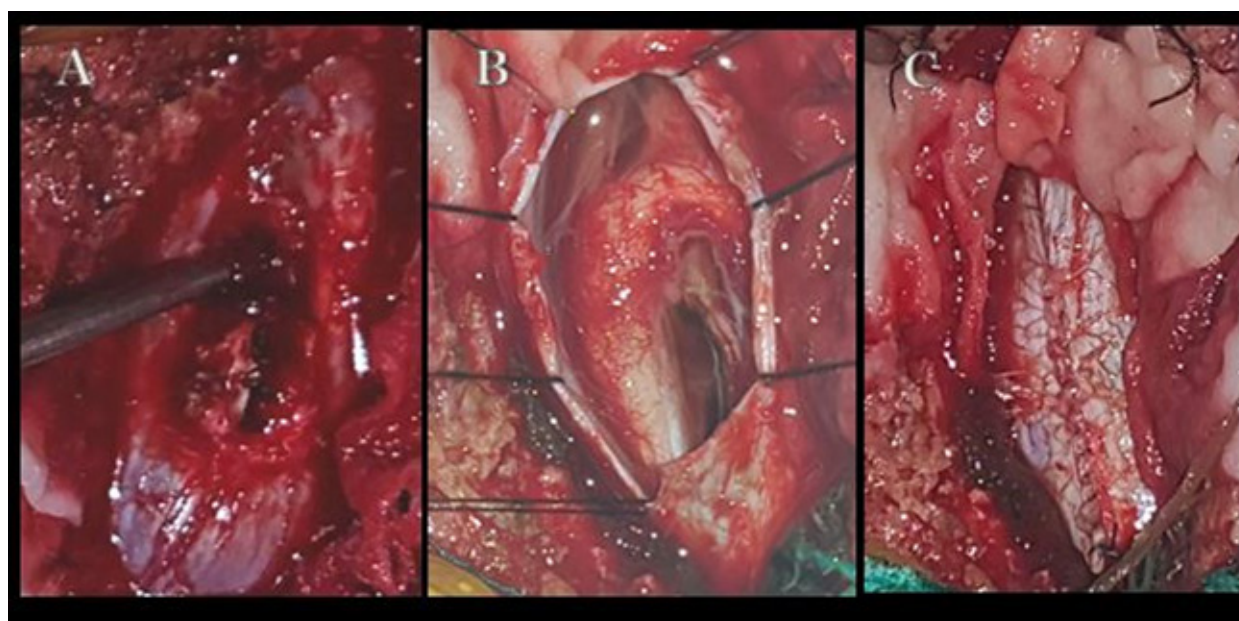


Figure 2. A: Intraoperative picture showing midline bony spur separating the two halves of the cord along with dura (arrow). B: After dural opening two parts of the cord clearly seen (double arrows). C: After Completion of dural closure making a single dural tube enclosing the split portions of the cord (triple arrows).

characterized by “splitting up” of the cord longitudinally for a variable length. It is more common in females. The most common location is lower thoracic and lumbar area. This condition is commonly associated with other anomalies such as congenital kyphoscoliosis, hemivertebra, butterfly vertebra, tethered cord, to name a few.¹

It is broadly divided into two types and both types are equally common.² In type 1, each split portion has separate Dural sleeve whereas in type 2, the split portions are inside one dural tube. Usually, bony spur is the ‘culprit’ for separation but occasionally a fibrous or cartilaginous spur is encountered.

Oliver first described DM in his treatise on disease of the spinal cord in 1837.¹ The credit of first performing the surgery on DM goes to Dr. Hamby in 1936.³ Matson first published the first series of 11 cases of DM in 1950.⁴

Though the exact etiology of this uncommon entity is unknown, it is believed to be due to a congenital disorder of neurulation. It is speculated that an accessory neuroenteric canal develops during the fourth week of life due to the development of adhesions between the embryonic ecto- and endoderm, which initiates the diastatomyelic process.^{1,5}

Symptoms are typically due to early cord traction around the spur. Congenital spinal deformity is the most common clinical manifestation in these patients. Gait abnormality and sphincter disturbances especially bladder are other presenting symptoms.^{1,6,7} Sometimes there are indirect clues to the presence of underlying pathology. One common tell-tale sign is the skin changes in the lumbosacral region (hairy patch, hyperpigmentation, a dermal, etc). All these were conspicuously absent in our patient.

Diagnosis of this condition in patients with subtle symptoms demands a high index of suspicion. Plain radiographs often show different degrees of spinal dysraphism and midline bony spur, if present. However, the investigation of choice is an MRI of the spine

which clinches the diagnosis. To delineate the bony anatomy, a computed tomography can be done, but is optional.

Management generally consists of operation with an aim to resect the bony septum and making a single tube of dura enclosing the split parts together in type 1 abnormality.^{1,8} The surgery is more prophylactic than curative. No extra procedure is recommended if there is associated syrinx present as in our case.⁸ The majority of the patient do well.

CONCLUSION

DM is a congenital malformation due to faulty development of the notochord. Though detected in infancy and childhood, adult presentation is not uncommon. In suspected cases, MRI is the investigation of choice to diagnose the condition. Management is largely surgical with the prophylactic aim of preventing neurological deterioration in the future. One needs to have a high index of suspicion in differential diagnosis in patients with subtle spinal deformity.

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