Case Report

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Diastematomyelia: A Case Report

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Abstract

Diastematomyeliais a congenital malformation in which a part of the spinal cord is split, usually at the level of the upper lumbar vertebra in the sagittal direction. It results from an abnormal adhesion between ectoderm and endoderm. Females are affected much more commonly than males (3:1).

There is complete or incomplete sagittal division of the spinal cord into two hemi cords due to the presence of an osseous, cartilaginous or fibrous septum in the central portion of the spinal canal.

The course of the disease is progressive as the patients may be asymptomatic to begin with and gradually progress to sensory motor disorder and loss of bowel and bladder control. With modern imaging techniques, various spinal dysraphism can be diagnosed earlier which can aid in improving quality of life of the individual. Treatment depends upon the symptoms. Regular neurological examinations may helps in early detection of progression of the disease and resection is done if required. We are presenting here a case of diastematomyelia, its imaging study and management.

Keywords: Diastematomyelia; Dysraphism; Imaging; Resection.

INTRODUCTION

Diastematomyelia (DSM) is a spinal cord malformation that occurs as a consequence of error during the embryogenic development of spinal cord. DSM is a type of split cord malformation (SCM), in which there is longitudinal split of spinal cord by either cartilage or bone or fibrous septum[1-3] Very few such cases have been reported in adults. In this case report we will discuss an adult patient with a focal DSM who presented with low back pain with bilateral lower extremity numbness and pain.

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CASE PRESENTATION

This case represents a 49 years old female with history of low back pain for 2 years and bilateral lower limb pain and numbness for past 2-3 months. No past history of any trauma, headache, fever or any viral illness. Her bowel and bladder habits were normal. The patient was known case of hypertension and was on regular treatment. Her surgical history was non-contributory. Patient had no addiction of tobacco, alcohol or any other drug. Family history was also non significant.

Her general examination doesn't reveal any abnormality. The neurological examination demonstrated power of grade 4+/5 observed at bilateral hip joints with normal tone. Bilateral straight leg raising test was positive at 45 degree on both sides. Lower extremity had no sensory deficits. Abdominal reflexes were active and symmetrical. Bilateral patellar tendon, achilles, Babinsky reflexes were 2+.

Imaging workup with MRI of the lumbar spine revealed an anomalous development of L4 posterior vertebral elements with abnormal widening of lamina, duplicated spinous process and posterior midline osseous block (Fig. 1). The latter was blocking canal at L4 and causing marked compression of the cal sac and caudaequina roots.

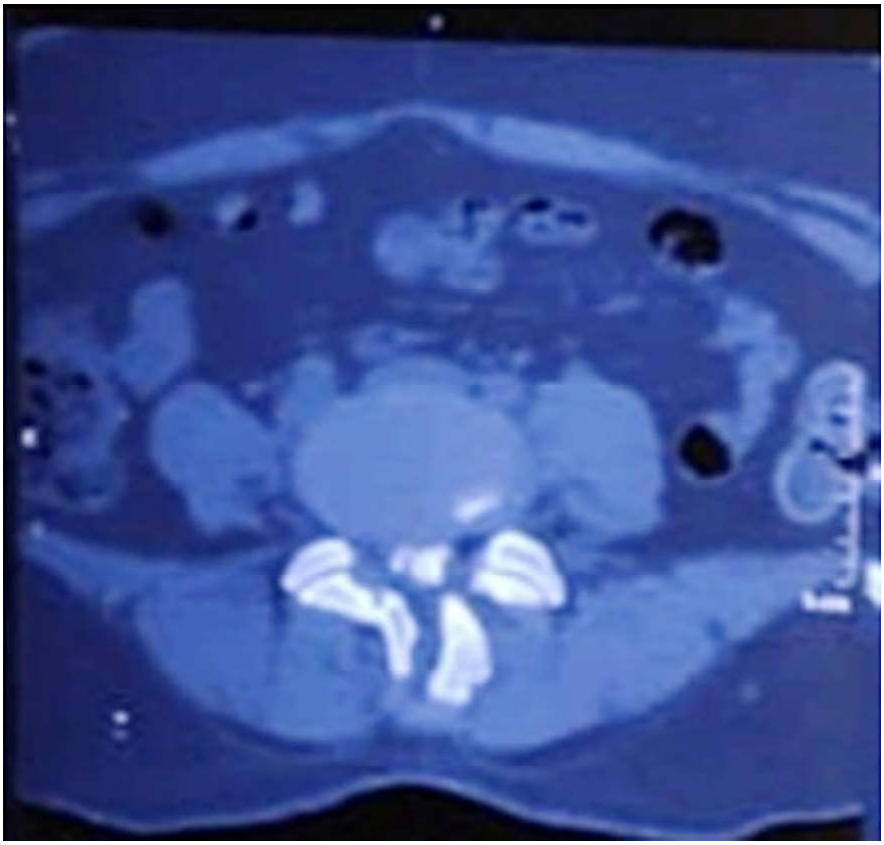

Fig. 1: CT axial scan showing anomalous parts of L4 vertebra along with osseous spur.

Short focal split cord malformation diastematomyelia opposite L2-L3 levels with division of cord into two equal halves at L2 and further reunion of these symmetric hemi cords at L3 level. These findings indicate associated low lying cord with teethered filum (Fig. 2, 3). The rest of the lumbar vertebrae were developmentally normal, with normal alignment and with no signs of spondylitis.

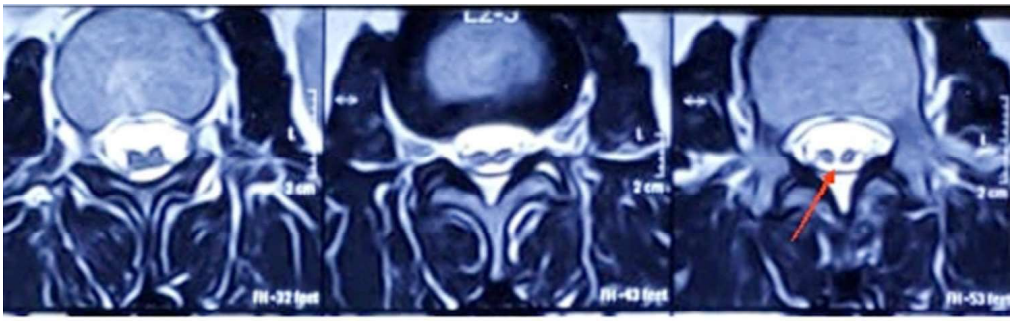

Fig. 2 & 3: MRI axial scan showing double cord (Diastematomyelia)

During the hospital stay neurophysician, neurosurgeon, physiotherapist and pain management specialists were consulted. Acute inflammatory demyelinating polyneuropathy (AIDP), acute transverse myelitis, chronic inflammatory demyelinating polyneuropathy (CIDP), acute cerebral vascular accident, or infectious etiologies were ruled out after extensive workup. Due to duplicated spinous process and posterior midline osseous block at L4 which was causing marked compression of the cal sac and caudaequina roots, L4 laminectomy was done along with excision of osseous spur and deteethering of cord.

The patient participated in physiotherapy and pain management during her hospital stay. Further outpatient follow-up at our neurosurgery OPD for an electromyogram (EMG) and nerve conduction study (NCS) was scheduled for further evaluation.

DISCUSSION

Diastematomyelia is characterized by longitudinal "splitting up" of the spinal cord into two. Though it is a rare congenital disorder, its prevalence was predicted 16% in pediatric patients in the study conducted by Shen *et al.*[4] Females are affected more than males. Prevalence of DSM in the general adult population is not known.[5] There is a complete or incomplete sagittal division of the spinal cord into two hemi cords by an osseous (bone), cartilaginous or fibrous septum. In our patient, DSM was a coincidental finding when the patient presented to the hospital for worsening lower backache and bilateral lower extremity numbness. DSM is broadly defined into two types. Type one is characterized by the duplication of an embryogenicdural tube with hemi cords while

type two has a single embryogenicdural tube with a divided spinal cord.[6,7] DSM has been found associated with intra and extra dural findings such as hypertrichosis, hemangioma, vertebral scoliosis, hydromyelia, syringomyelia, dermoid, lipoma, chiari malformation and spina bifida.[6-8] DSM may be isolated or associated with other vertebral body segmental anomalies.[9]

It usually occurs between D9 and S1 levels.[10,11] Cervical diastematomyelia is a rare of all.[12,13] In 50% of the patients, the hemi-cords are contained in separate dural sac and bony or fibrous spurs are usually found between the two sacs.[14]

This anomaly can be diagnosed in early mid trimester by prenatal ultrasound. It appears as an extra posterior echogenic focus between the foetal spinal laminae with splaying of the posterior elements on ultrasound. Antenatal diagnosis hence allows an early surgical intervention and a favourable outcome.[9,15]

Plain radiographs can recognize bony malformations and dysplasia and if required or clinically indicated, they can be further confirmed by CT scan. However, MRI is the technique of choice for dysraphism and will also allow adequate analysis of the spinal deformities.[16]

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