Case Report

Diastematomyelia : A Case Report with Review of Literature

Kishore PVK[1], Gnaneshwar P[2], Sudha[3], Divya[4]

[1] Consultant Neurosurgeon
[2] Duty Medical Officer
[3] Junior Resident
Department of Anaesthesia
Maheshwara Medical College and Hospital
Medak Dist. - 502 307
[4] Junior Resident
Department of Pulmonology
Maheshwara Medical College and Hospital
Medak Dist. - 502 307
Telangana, India.

CORRESPONDENCE :

[1] Dr.P.V.K.Kishore
Mch (Neuro surg)
Consultant Neurosurgeon
Sree Lakshmi Bhadrakali
Neuro Trauma Hospital
Karimnagar-505 001
Telangana, India.
E-mail:puttetikishore@gmail.com

ABSTRACT

Diastematomyelia is complete or partial splitting of the spinal cord into two hemicords, each with its own central canal and pia surrounding it. We are reporting the clinical features, radiological evidence and surgical outcome of a 3 year old female child who was diagnosed to have Diastematomyelia. In addition, we have presented review of relevant Literature.

Keywords: Diastematomyelia, hemicords

INTRODUCTION

The term Diastematomyelia was first used by OLIVER in 1837 and is derived from Greek "Diastema'' meaning crack and "mielia" which refers to the spinal marrow. Previous to 1950 this infrequent disease was little known and the few diagnosed cases were reported post-mortem.[1]

Split Spinal Cord Malformation[SSCM] can be defined as a form of spinal dysraphism in which any or all of the Spinal Cord,Cauda Equina and Filum Terminale are divided into two lateral parts by a dorsal-ventral bony or fibrous spur.[2]

There are two described types of SSCM namely Type 1 and Type 2. The two hemicords may be separated by a bony or osseocartilagenous spur and be contained in a separate Dural sheaths (SSCM type1) or they may be separated by a fibrous spur and contained in a single Dural sheath (SSCM type2).[3]

Although the precise cause of Neural Tube Defect(NTD) remains unknown. Evidence suggests that many factors including radiation, drugs, malnutrition, chemicals and genetic determinants (mutation in Folate responsive or Folate dependent pathway) may adversely affect normal development of CNS from the time of conception.[4]

CASE REPORT

A 3year old female child presented with tuft of hair at the lumbo-sacral region [Figure 1a]. No other neurological abnormalities detected. Routine laboratory results were normal. Plain X-ray of lumbar sacral vertebra in the frontal projection showed widening of interpedicular distance from L4 to L5 with Spina bifida from L1 to L5.Bony spur noted in the middle of spinal canal [Figure 1b].

CT Scan with 2D Reconstruction showed a bony spur noted at the level of L3 extending from posterior elements to the middle of the vertebral bodies separating the spinal canal into two halves [Figure 1c,1d]

MRI-revealed open spina bifida from L2 to L5 level with dorsal dermoid. Complete focal split spinal cord from L2 to L4 with bony spur inbetween two halfs at L3 level,assosciated with low-lying conus.

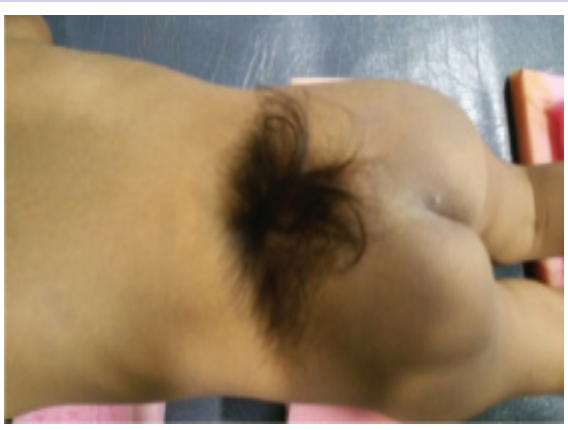

Fig 1a-Tuft of hair in Lumbo-sacral region

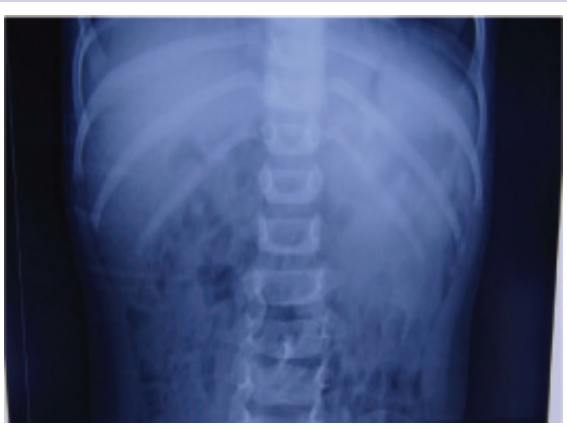

Fig 1b-X-ray showing Spina Bifida and widened interpedicular distance

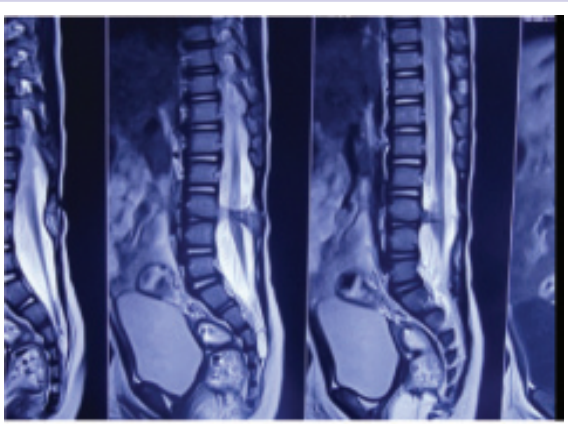

Fig 1c MRI whole spine showing Bony Spur at L3 level

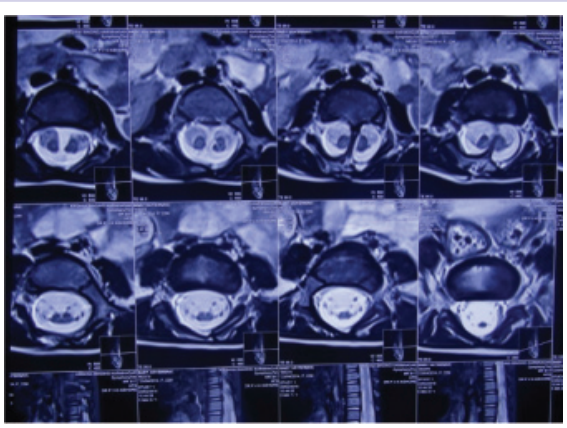

Fig 1d MRI axial sectioning showing two Hemicords with bony spur between them

Surgical Technique

The child was positioned in prone position on bolsters after endotracheal intubation. A midline incision was made from L1 to L5 levels and subperiosteal elevation was performed to expose L2 to L5 lamina. L3 and L4 laminectomy was performed. At the level of L3 a bony spur was noted extending from the anterior surface of the lamina to the posterior surface of the vertebral body dividing the Dural sac into two separate tubes [Figure 2a]. The two Dural tubes were reunited at level of L4. The bony spur was carefully drilled till its anterior attachment to the vertebral body [Figure 2b].

Dural incision was made around the dural cleft in-between the two dural tubes. The part of the dura in the middle which is extending anteriorly in-between the two spinal cords is also excised. We traced the lower lying

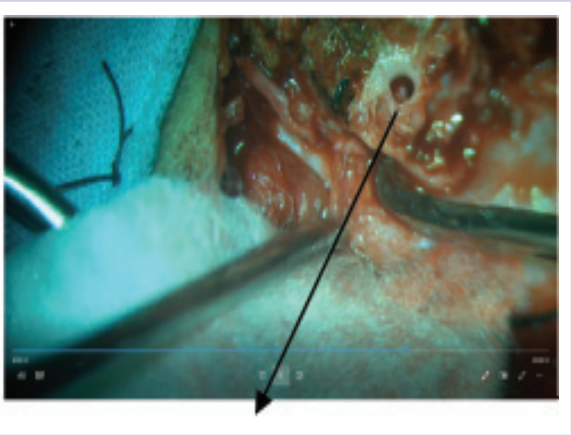

Fig 2a : A-Bony spur

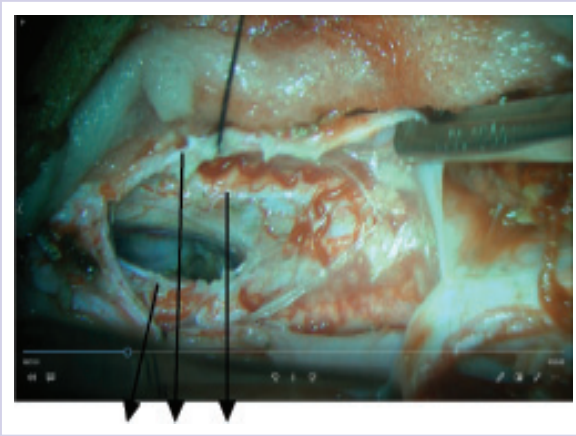

Fig 2b : B-Hemicord 1 C-Dura D-Hemicord 2

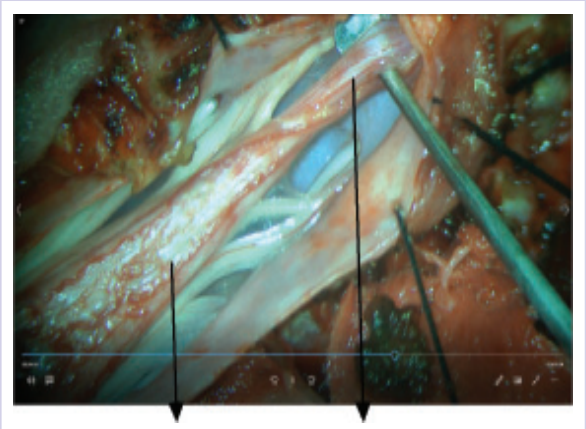

Fig 2c : E-low lying cord F-Filum Terminale

cord inferiorly till we reached the Filum Terminale at the level of L5 and is noted to be grossly thickened [Figure 2c]. The Filum Terminale was now sectioned. We could appreciate upward migration of the spinal after sectioning of the Filum Terminale. The posterior two dural edges was closed water tight and wound was closed in layers. Patient had uneventful post-operative recovery with no neurological deficits.

DISCUSSION

Mahapatra and Gupta sub classified patients with type 1 SSCM as follows

- Type 1a-Bony spur in the centre with equally duplicated cord above and below the spur
- Type 1b-Bony spur at the superior pole with no space above and a large duplicated cord lower down
- Type 1c-Bony spur at the lower end with a large duplicated cord above
- Type 1d-Bony spur straddling the bifurcation with no space above or below the spur[3] Diastematomyelia is assosciated with a number of anomalies. These are Cutaneous anomalies of the back:- Cutaneous Nevi overlying the site of Diastematomyelia occur in 50-75% of patients.[6] Characteristics of this is nevus pilosus, a large batch of long silky hairs situated over the site of cleft.
- Other cutaneous abnormalities include: Dermal sinus, Fibrous band, Lipomas, Hemangiomas, Dimple[7,8] Nevus Pilosus was observed in our case.
- Orthopedic deformities of the foot especially clubfoot are found in half of the patients.[6]
- Spinal column is nearly always grossly abnormal in Diastematomyelia. Laminas are fused or thickened in 90% of the cases, spina bifida of greater severity at L5 and S1, widening of interpedicular distance at the level of Doastematomyelia retaining the medial convexity of the pedicles, and abnormal vertebral body occur in 85% of cases.
- Abnormal vertebral bodies can be Hemivertebra, Block vertebra, Butterfly vertebra, narrowed intervertebral disk space. Scoliosis and Kyphosis occur in 50-60% of the cases. They are related to the bone abnormalities and their incidence and severity increase with age[4,5,8] In our case we noted Spina bifida, irregularly shaped vertebral bodies and bony spur.
- Assosciated other spinal abnormality:-Tethered cord which occurs due to Diastematomyelia itself or assosciated anomalies so that the conus medullaris lies below L2 in 76% of the cases; Filum Terminale is thicker than 2mm in 40-100% of the cases; Hydromylia in 46% of patients and may extend from

the cord above the cleft into one or both hemicords; Meningoceles; Meningomyeloceles; Chiari malformation and intraspinal masses such as Dermoids, Lipomas, Teratomas.[5, 9] Our patient had thickened Filum terminale.

- Intramedullary tumours assosciated with Diastematomyelia have been rarely described.[10]

IMAGING OPTIONS

Plain X-ray is often the first modality of Investigation and both AP and Lateral films are needed. It shows the vertebral anomalies and the bony spur in 50% of the cases. Sometimes the spur may be obscured by vertebral anomalies.[6, 8]

Prenatal ultrasound diagnosis of this anomaly is usually possible in the early mid third-trimester. An extra posterior echogenic focus between the foetal spinal laminae is seen with splaying of the posterior elements, thus allowing an early surgical intervention and a favourable prognosis.[11,12]

CT unmistakably demonstrates the bony spur. The cartilaginous and soft tissue spurs are well seen with CT-Myelography. It clarifies the Diastematomyelia and assosciated spinal anomalies and accurately demonstrates vertebral anomalies. Reconstruction will help to see full extent of the spur and cord clefting.[8]

MRI with its multiplanar capabilities and superior soft tissue contrast generally allows adequate analysis of the spinal deformities and is a technique of choice for dysraphism in general.[5]

CONCLUSION

Only on meticulous history and physical examination supported by radiological investigation and interpretation such rare diseases can be diagnosed such as one presented here.

CONFLICT OF INTEREST :

The authors declared no conflict of interest.

FUNDING : None

REFERENCES

1. Pang D, Dias MS, Ahab-Barmada M. Split cord malformation, part I: A unified theory of embryogenesis for double spinal cord malformations. *Neurosurgery.* 1992; 31(3):451-80.

2. Schijman E. Split spinal cord malformations: report of 22 cases and review of the literature. *Childs Nerv Syst.* 2003; 19(2):96-103.

3. Mahapatra AK, Gupta DK. Split cord Malformations: A Clinical study of 254 patients and a proposal for a new Clinico-Imaging classification. *J Neurosurg Pediatr.* 2005; 103(6):531-6.

4. Kinsman SL, Johnston MV. Congenital anomalies of CNS. In: Kliegman RM, Stanton BMD, Geme JS, Schor N, Behrman RE, editors. Nelson text book of Pediatrics. 19th ed. London: WB. Saunders Company; 2011:1883-993.

5. Singh H, Maurya V, Saini M. Images-Diastematomyelia. *Ind J Radiol Imag.* 2000;10(4):1-5.

6. Naidich TP, Zimmerman RA, McLone DG et al. *Congenital malformation of spine and spinalcord.* In: C. Manelfe: Imaging of the spine and spinal cord. Raven Press Ltd. New York; 1992:685-94.

7. Unsinn KM, Geley T, Freund MC, Gassner I. US of the Spinal Cord in Newborns: Spectrum of Normal Finding, Variants, Congenital Anomalies, and Acquired Diseases. *Radiographics.* 2000; 20(4):923-38.

8. Hawkins TD. The Abnormal spine. In Duboulay H. *Textbook of radiological diagnosis.* Ed Vol 1, London: HK Lewis and Co Ltd; 1984:472-585.

9. McClelland RR, Marsh DG. Double Diastematomyelia. *Radiology.* 1977; 123(2):123-378.

10. McMaster MJ. Occult intraspinal anomalies and congenital scoliosis. *J Bone Joint Surg Am.* 1984; 64(4):588-601.[PubMed]

11. Allen LM, Silverman RK. Prenatal ultrasound evaluation of fetal diastematomyelia: two cases of Type I split cord malformation. *Ultrasound Obstet Gynecol.* 2000; 15(1):78-82. [PubMed]

12. Bannizavon Bazan U, Krastel A, Lohkamp FW. Diastematomyelia - a harmless finding or cause of late neurological disturbance. *Z Orthop Ihre Grenzgeb.* 1978; 116(1):72-80. [PubMed]