Case Report

Imaging findings in diastematomyelia: A rare type of spinal dysraphism

Ramakrishna Narra[1], Chaitanya Kadiyala[2], Prathibha Gadde[2], Bhimeswara Rao Pasupuleti[3]

From [1]Professor, [2]Junior Resident, [3]Professor, Department of Radio-diagnosis, Katuri Medical College and Hospital, Guntur, Andhra Pradesh, India

ABSTRACT

Diastematomyelia, also termed as split cord malformation (SCM), is a rare congenital spinal dysraphism in which there is an abnormal sagittal clefting of the spinal canal that causes the cord to split into two hemicords by a fibrous, osseous, or cartilaginous septum. Various vertebral segmental anomalies such as hemivertebrae, butterfly vertebrae, and scoliosis are seen in association with diastematomyelia. Most of the diagnosed cases of diastematomyelia are children under the age of 7 years. Such a spinal anomaly is less frequently diagnosed in adults with female preponderance. We reported a case of diastematomyelia that was diagnosed by magnetic resonance imaging (MRI) in a 23-year-old woman who presented with chronic back pain for 7 years. MRI is the imaging modality of choice for establishing the diagnosis and exploring any associated malformations.

Key words: Diastematomyelia, Magnetic resonance imaging, Spinal dysraphism, Split cord malformation

Diastematomyelia is a rare congenital spinal cord anomaly that belongs to a group of occult spinal dysraphisms. It occurs as a result of an abnormal notochord development between the 15th and 18th days of gestation [1-3]. Several cases of diastematomyelia were reported in the literature in the prenatal period by means of ultrasonography that is confirmed by magnetic resonance imaging (MRI) [4]. Its diagnosis in the later age groups is rare as seen in our case. It shows a female predilection accounting for about 80–90% and it most commonly occurs within the dorsolumbar spine (85%). In a few cases, it may be confined to either the lumbar spine or to the thoracic spine. Reported cases of cervical or multilevel diastematomyelia were also reported [4]. Here, we report the case of diastematomyelia which was diagnosed based on the imaging findings in a woman at the age of 23 years which is an unusual age for the presentation.

CASE REPORT

A 23-year-old female presented with a complaint of chronic dull back pain for 7 years which is radiating to the bilateral lower limbs, aggravated on work, and relieved with rest and usage of NSAIDs. She had a history of primary infertility. The patient had no other relevant symptoms.

On examination, the vitals were stable. On physical examination, an external spine deformity (scoliosis) was noticed with primary curvature toward the left at the thoracolumbar region and with positive Adam's forward bend test. Small swelling in the lumbar region was noticed. There was no evidence of any abnormal hair growth at the level of the spine, foot deformities, and leg shortening.

In view of persistent pain, an MRI of the whole spine was performed with a 1.5T scanner. The study revealed a split spinal cord due to the presence of an osseous septum from the T5–T9 level. On T2-weighted (T2W) coronal and axial images, the hypointense bony septum was present in the midline extending from the vertebral body anteriorly to abnormally thickened neural arch posteriorly at the T8 level dividing the spinal canal into two separate canals including two hemicords in separate dural sacs (Figs. 1 and 2). The hemicords were again united below the level of the cleft. On T2W axial MR images, both dural sacs showed the posterior defects with herniation of meninges and spinal nerves (Figs. 3a-c). The hemicord on the right showed syringohydromyelia (Fig. 3d). Computed tomography (CT) with bone window settings clearly demonstrated the bony spur (Fig. 4). A final diagnosis of Pang type-I Diastematomyelia was considered. The patient was treated conservatively and follow-up was advised regularly.

DISCUSSION

Dysraphic lesions constitute a broad group of developmental abnormalities affecting the spinal cord and canal [5]. They are generally diagnosed in the neonatal period and may also present in older children or adults [5]. As per the literature,

Access this article online	
Received - 23 November 2020 Initial Review - 08 December 2020 Accepted - 23 December 2020	Quick Response code
DOI: 10.32677/IJCR.2020.v06.i12.014	

Correspondence to: Chaitanya Kadiyala, Department of Radio-Diagnosis, Katuri Medical College and Hospital, Chinakondrupadu, Guntur - 522 019, Andhra Pradesh, India. E-mail: chaitanyakmbbs@gmail.com

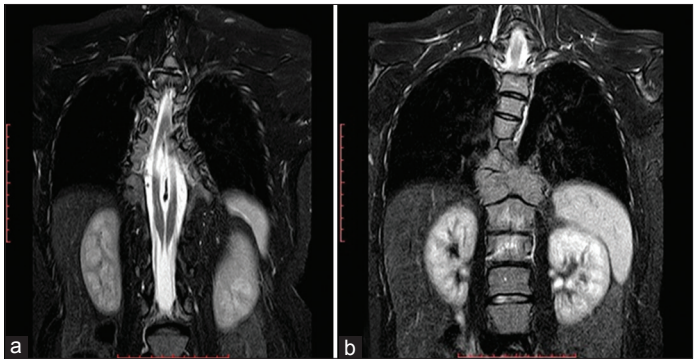

Figure 1: (a) Coronal T2W images demonstrating a sagittal cleft spitting the spinal cord into two hemicords due to the presence of an osseous septum, (b) vertebral anomalies (hemivertebrae with rudimentary intervertebral discs) at multiple levels are also noted

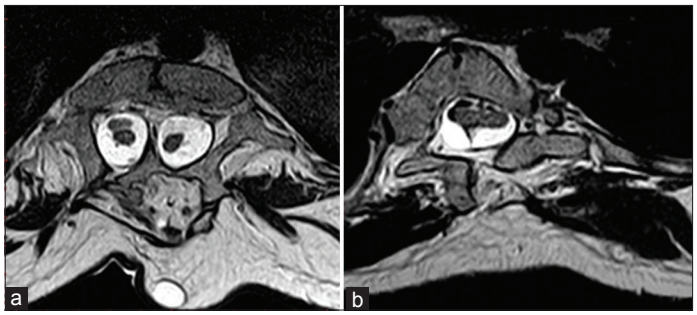

Figure 2: (a) Axial T2W image showing two hemicords in separate dural sacs separated by a bony spur. A bifid spinous process is also noted; (b) Axial T2W MR image showing two hemicords united below the level of cleft

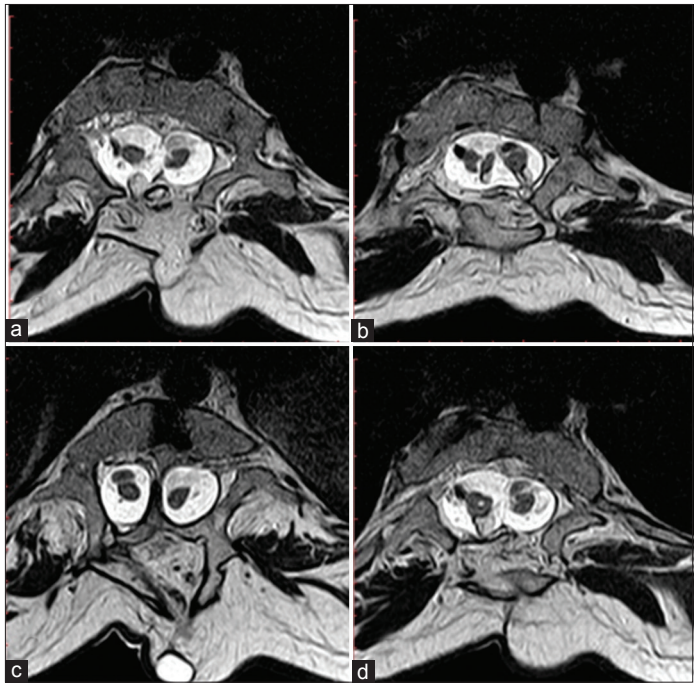

Figure 3: (a-c) Axial T2W images showing two dural sacs with posterior herniation of meninges and spinal nerves which is suggestive of myelomeningocele. (d) The hemicord on the right is showing a dilated central (arrow) which is suggestive of syringohydromyelia

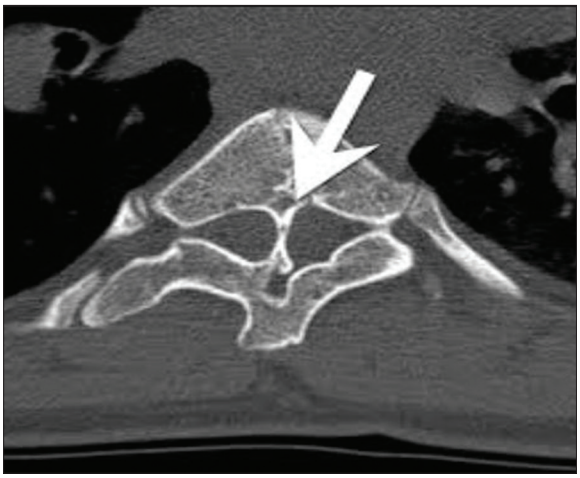

Figure 4: Axial CT image in bone window showing a bony spur (arrow) dissecting the spinal canal

presentation of diastematomyelia in advanced age is very rare and a case was reported in a 72-year-old woman [6]. These lesions are divided into three groups that include open spinal dysraphism with a dorsal protrusion of contents of the spinal canal through the overlying skin defect, closed dysraphism with a protrusion of neural tissue which is covered by the skin, and occult spinal dysraphism without any dorsal protrusions of neural tissue [7].

The clinical-neuroradiological classification of spinal dysraphisms is broadly categorized into open and closed types. In an open spinal dysraphism, there is a defect in the overlying skin, and the neural tissue is exposed to the environment. It includes myelomeningocele, myelocele, hemimyelomeningocele, and hemimyelocele. In a closed spinal dysraphism, the neural tissue is covered by the skin. Closed spinal dysraphisms can be further subcategorized on the basis of the presence or absence of a subcutaneous mass. Closed spinal dysraphisms with subcutaneous mass includes lipomyelomeningocele, lipomyelocele, meningocele, myelocystocele, and skin-covered myelomeningocele (limited dorsal myeloschisis). Closed spinal dysraphisms without subcutaneous mass consist of simple and complex dysraphic states. Simple dysraphic states include intradural lipoma, filar lipoma, tight filum terminale, persistent terminal ventricle, and dermal sinus, whereas, complex dysraphic states include disorders of midline notochordal integration (diastematomyelia, neurenteric cysts, and dorsal enteric fistula) and notochordal formation (caudal agenesis and segmental spinal dysgenesis).

Split cord malformations (SCM) are more common in the lower cord but can sometimes occur at multiple levels. The majority of patients with diastematomyelia are symptomatic, presenting with signs and symptoms of tethered cord that include leg weakness, low back pain, scoliosis, and incontinence. The patients with mild form are minimally affected or asymptomatic. Diastematomyelia is a type of occult spinal dysraphism in which there is a vertical split in the spinal cord. The bony septum dividing the spinal canal is confirmatory, and in most of the cases, the split is due to the cartilaginous or fibrous septum. A bony septum was demonstrated in this case. The hemicords are seen in a single dural sac in around 50% of patients, while in others they

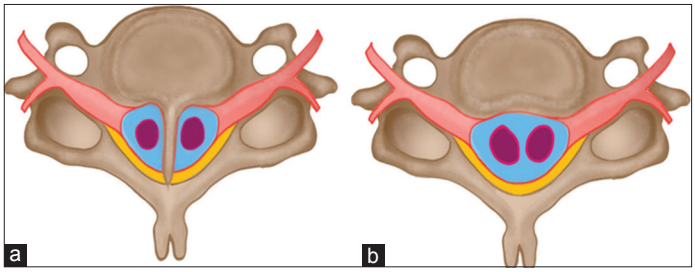

Figure 5: Graphical illustration demonstrating the two types of SCM described by Pang *et al*. (a) Type I with two hemicords one in each dural sac and (b) Type II characterized by a single dural sac containing both hemicords

lie within separate dural sacs as seen with this case. Numerous associated anomalies that usually present with diastematomyelia are myelomeningocele, meningocele, inclusion dermoid, spinal lipoma, tethered cord, syringohydromyelia, neuroenteric cysts or dermal sinus, and Chiari II malformation. Occult dysraphisms may be seen with a normal skin covering. In about 50–75% of cases, multiple skin lesions were seen. In about 56% of cases, hypertrichosis is the skin marker that is usually observed at the level of the malformation [8,9].

According to the Pang *et al*., the classification of SCM is of two types [2,3]. Pang type I SCM is characterized by a dual dural sac with two hemicords one in each dural sac (Fig. 5a), whereas, Pang type II SCM is characterized by a single dural sac containing both hemicords (Fig. 5b). The two hemicords in Pang type I are separated by an osseocartilaginous spur, while in most cases of Pang type II, no osseous spur is seen. Pang type I split cords are usually associated with surgical morbidity than Pang type II, especially in cases with asymmetrical oblique septum dividing the cord.

Differential diagnosis includes dimyelia (complete spinal cord duplication) and diplomyelia (the presence of an accessory spinal cord) [10]. Bony dysplasias and malformations are usually diagnosed on plain radiographs and CT. However, MRI provides adequate information of the spinal deformities and is the diagnostic modality of choice for spinal dysraphisms which helps in arriving at a final diagnosis and planning for surgery.

CONCLUSION

The reported case of diastematomyelia was diagnosed on MRI images of a 23-year-old female patient with chronic back pain. Diastematomyelia is a rare spinal dysraphism that is uncommon in adults. It is usually diagnosed on plain radiographs and CT. However, the MRI will provide adequate information. This disorder has to be picked up in patients with plain radiographs revealing vertebral segmental anomalies such as spina bifida, kyphoscoliosis, butterfly vertebrae, or vertebral fusion, and hemivertebrae, especially when associated with additional skin lesions at the level of the detected osseous anomalies, dorsally.

REFERENCES

1. Karasahin KE, Gezginc K, Alanbay I, Ulubay M, Başer I. Ultrasonographic diagnosis of diastematomyelia during the 14th week of gestation. Taiwan J Obstet Gynecol 2009;48:163-6.
2. Pang D, Dias MS, Ahab-Barmada M. Split cord malformation: Part I: A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery 1992;31:451-80.
3. Pang D, Hoffman HJ, Rekate HL. Split cord malformation part II: Clinical syndrome. Neurosurgery 1992;31:481-500.
4. Kulkarni M, Ruparel M, Redkar R. Fetal diastematomyelia: MR imaging: A case report. Indian J Radiol Imging 2009;19:78-80.
5. Netto JM, Bastos AN, Figureueiredo AA, Pérez LM. Spinal dysraphism: A neurosurgical review for the urologist. Rev Urol 2009;11:71-81.
6. Maebe H, Viaene A, De Muynck M. Diastematomyelia and late onset presentation: A case report of a 72-year-old woman. Eur J Phys Rehabil Med 2017;54:618-21.
7. Biri AA, Turp AB, Kurdoglu M, Himmetoğlu O, TokgözErcan N, Balci S. Prenatal diagnosis of diastematomyelia in a 15-week-old fetus. Fetal Diagn Ther 2005;20:258-61.
8. Sonigo-Cohen P, Schmit P, Zerah M, Chat L, Simon I, Aubry MC, *et al*. Prenatal diagnosis of diastematomyelia. Childs Nerv Syst 2003;19:555-60.
9. Lewandrowski KU, Rachlin JR, Glazer PA. Diastematomyelia presenting as progressive weakness in an adult after spinal fusion for adolescent idiopathic scoliosis. Spine J 2004;4:116-19.
10. Nour TA, Alexander JT, Richard BT. Pediatric radiological case: Diastematomyelia. Appl Radiol 2014;43:1-4.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Narra R, Kadiyala C, Gadde P, Pasupuleti BP. Imaging Findings in Diastematomyelia: A Rare Type of Spinal Dysraphism. Indian J Case Reports. 2020;6(12):715-717.