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Diplomyelia: A Split in the Unsplit

Sushil Kachewar, MD, DNB
Rural Medical College
Loni, India

Smita Sankaye, MD
Rural Medical College
Loni, India

Devidas Kulkarni, MD, DMRD
Rural Medical College
Loni, India

Address for correspondence:
Sushil Kachewar, MD, DNB
Rural Medical College
Loni, India
Email: sushilkachewar@hotmail.com

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Normal human spinal cord is unsplit in nature, meaning that it is single in its entire course. But rarely, this unsplit cord gets split into two and then it is known as Diastematomyelia. When the splitted cord does not reunite distal to the spur then a true duplication of the spinal cord termed diplomyelia is said to occur. This unique congenital malformation occurs due to an abnormal developmental communicating structure between the ectoderm and the endoderm. The splitter of the cord may be in the form of fibrous tissue or purely a calcific bar or purely an ossific bar or even a combination of the earlier mentioned entities.

This clinical entity favors females thrice as often as it does the males. Diastematomyelia may be symptomatic, causing progressive neurological dysfunction including incontinence of the bowel and the bladder, weakness, sensory loss, or spasticity. Many asymptomatic cases are discovered incidentally during spinal imaging especially in patients who have asymmetry of the lower limbs, congenital scoliosis, or cutaneous lesions on the back like tufts of hair or abnormal skin patch.

Proper imaging is important in establishing the diagnosis and enables correct management.

As progressive neurological deterioration is usually a part of the natural history of this entity, many believe that it should usually be treated with resection of the spur. The treatment of the scoliosis seen in these patients is complicated as progression of the congenital scoliotic curve does not appear to be altered by resection of the spur, and the scoliosis needs to be treated independently.

A representative case of Diplomyelia is discussed in this case report with emphasis on role of imaging in the diagnosis.

Key Words: diastematomyelia, diplomyelia, imaging, surgery

When the naturally unsplit spinal cord is split into two hemicords, the condition is called as Diastematomyelia. Females are affected thrice often as the males. Structurally, there is an osseous (bony), cartilaginous or septum in the central portion of the spinal cord which causes a complete or incomplete sagittal division of the spinal cord into two hemi cords. Usually, the split cord unsplits after some distance. But rarely, when the splitted cord does not reunite distally to the spur then a true duplication of the spinal cord termed diplomyelia is said to occur.

It has been noted that the sagittal cleft may extend in the spinal cord, conus medullaris or even filum terminale and cause splaying of the posterior vertebral elements. Diastematomyelia may occur in isolation or may be associated with other segmental anomalies of the vertebral bodies.[1]

Case Report

A 20-year-old male presented with history of progressive dysfunction of the bowel and the bladder, with weakness, sensory loss and decresed sensation in the lower half of body over a period of 6 months. Externally on inspection his back was normal. There were neither

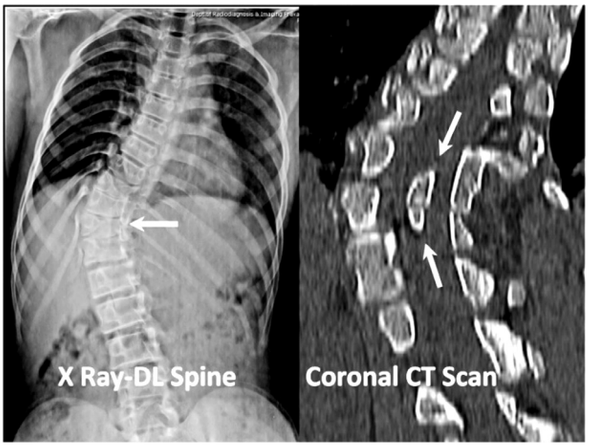

Figure 1: X-ray Spine and Coronal CT image showing calcified bar splitting the spinal canal and a scoliosis having concavity towards left.

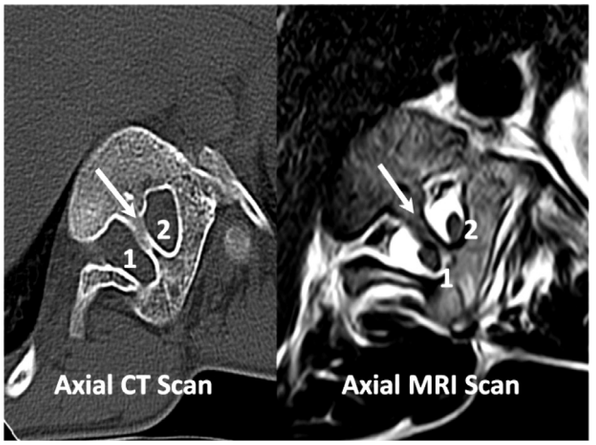

Figure 2: Axial CT & Axial MRI showing calcified bar splitting the spinal cord into two.

cutaneous abnormalities nor any swelling. Clinically he had paraparesis involving both the lower limbs. However, there was no club foot. He was referred for complete imaging evaluation.

As shown in **Figure1**; Antero-posterior X -ray of his dorso-lumbar spine and Coronal CT scan image showed a calcified bar splitting the spinal column into two. As demonstrated in **Figure 2;** Axial CT & Axial MRI images showed calcified bar splitting the spinal cord into two in the thoraco-lumbar region. Coronal MRI images (**Figure 3a, 3b and 3c)** not only showed the bony bar, but in addition demonstrated that the split hemicords diverged distally and did not fuse, thus confirming the diagnosis of diplomyelia. No meningocele was seen at any level. There were vertebral segmentation and fusion abnormalities at the site of the bony intraspinal bar in the lower thoracic-upper lumbar region. The patient was referred for neurosurgery at higher center.

Discussion

Diplomyelia is an uncommon variant of diastematomyelia in which the spinal cord remains divided caudal to the diastematomyelia spur.[8]The formation of Diastematomyelia and therefore of Diplomyelia is believed to be due to an accessory neurenteric canal which initiates this dysraphic state of unknown embryogenesis as is evidenced by the narrowing of a disc space at the same level and interlaminar fusion. In most cases the dividing bar is cartilaginous or fibrous or even bony.[3] Other anomalies of the vertebral column such as spina bifida, kypho-scoliosis, butterfly vertebra and hemivertebra are also known to be associated.[1]

Diastematomyelia usually occurs between D9 and S1 levels.[4] Cervical diastematomyelia is a very rare entity.[6] In about 50% of patients, the hemi-cords are contained in a single dural sac, while in the remaining they lie within separate dural sacs. In the latter, bony or fibrous spurs are usually found between the two sacs. Although tethered cord, inclusion dermoid, lipoma, syringohydromyelia and Chiari malformation have been seen to be associated, rarely intramedullary tumors have also been described.[7] While the dividing septum is believed to be the primary cause of symptoms, associated conditions like myelodysplasia and dysraphia of the spinal cord can also cause progressive neurological lesions.

Prompt diagnosis is possible with MRI of entire spine and screening of brain which now is the technique of choice.[3] Use of metrizamide myelography and computed tomographic metrizamide myelography to the diagnose dysraphic lesions of the spine is now a thing of past.[9] A third-trimester obstetric ultrasound shows an extra posterior echogenic focus between the fetal spinal laminae that splays the posterior elements, thereby enabling intranatal diagnosis and paving way for early surgical intervention and an enhanced quality of life.[2]

The American Association of Neurological Surgeons proposes Surgery and Observation as management approach.[2] They advocate surgical intervention in patients who present with new onset neurological signs and symptoms or have a history of progressive neurological manifestations which can be related to this abnormality. The surgical procedure required for the effective treatment of Diastematomyelia requires the decompression of neural elements and removal of bony spur. This may be accomplished with or without resection and repair of the duplicated dural sacs. Their preference is to resect and repair the duplicated dural sacs since the dural abnormality may partly contribute to the "tethering" process responsible for the symptoms of this condition. Minimally Invasive Microsurgical Techniques are becoming available to accomplish these operative tasks. This most often results in complete relief of symptoms or stops the progression of symptoms.

Observation is reserved for patients who are asymptomatic and have been identified with this anomaly while being investigated for other unrelated issues and hence do not require surgical treatment. These patients should undergo periodic neurological examinations since it is known that the condition can be "progressive". In the event that progressive neurological manifestations are identified, a resection should then be performed.

In pediatric patients however, prophylactic operations were associated with the best clinical outcome.[5] Despite

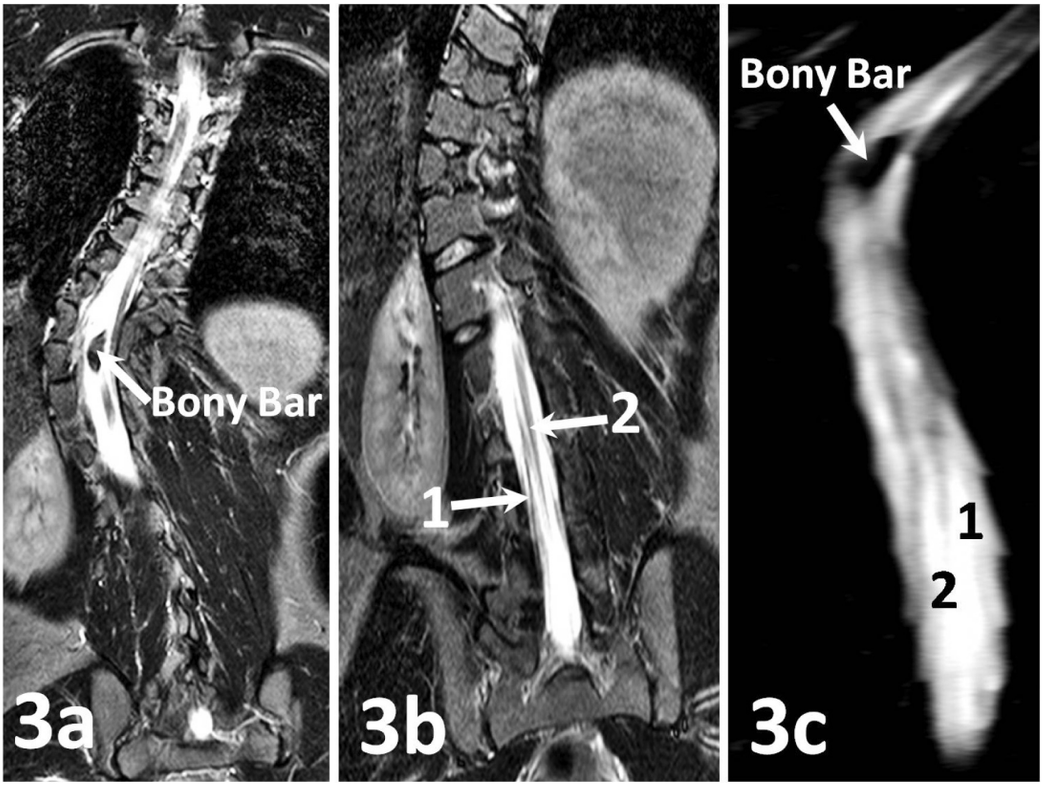

Figure 3: Coronal MRI images (Figure 3a, 3b and 3c) not only showed the bony bar, but in addition demonstrated that the split hemicords diverged distally and did not fuse, thus confirming the diagnosis of diplomyelia. No meningocele was seen. Lower thoracic and upper lumbar spinal segmentation and fusion abnormalities are seen along with a scoliosis having concavity towards left.

improvement, all patients with established preoperative deficit still had residual neurological deficits at their last follow-up. The associated syringomyelia remained unchanged after surgery, indicating that it does not contribute to the neurological syndrome.

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