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Case Report

Radiology Section

Dorsal Cord Herniation with Diastematomyelia- A Rare Case

SAAD SHAIKH[1], SNEHA SIRIGIREDDY[2], VISHAL WALASANGIKAR[3], VIRAJ SHAH[4], RAHUL NAVANI[5]

ABSTRACT

Transdural spinal cord herniation is rare and under reported. Ventral or ventro-lateral cord herniation is relatively more common and a well described entity. Dorsal or posterior spinal cord herniation is very rare, only five cases in the cervical spine have been reported, as per authors' knowledge. It is nonetheless a recognised cause of myelopathy, either acquired postsurgery or post-trauma or idiopathic in cause and congenital. The authors are reporting a rare case of a 47-year-old male patient who underwent Magnetic Resonance Imaging (MRI) spine. The MRI showed cord with diastematomyelia from C3 to C5 levels with kinking of cord at this level attached to the posterior dura. Posterior subarachnoid space at this level is completely effaced with no evidence of Cerebrospinal Fluid (CSF) posterior to the cord and hence, the patient was subsequently diagnosed as postoperative dorsal spinal cord herniation and myelomalacia in the cervical spine associated with diastematomyelia. Patient is being managed conservatively till date.

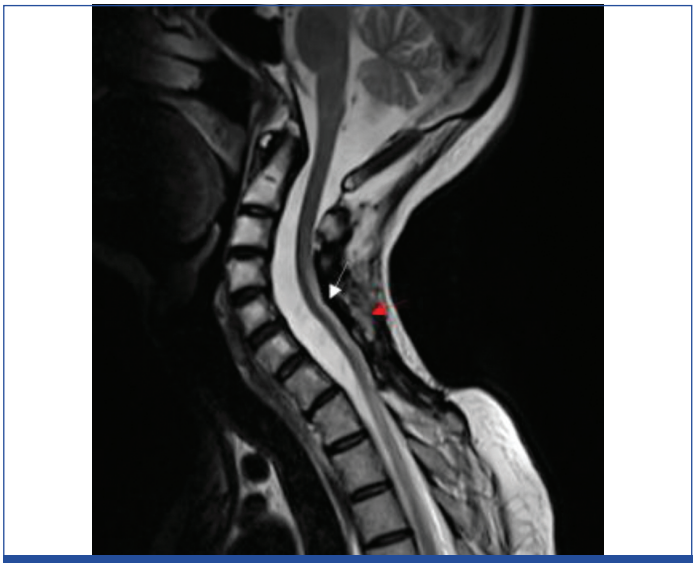
Keywords: Cervical spine, Dural defect, Myelomalacia, Transdural

CASE REPORT

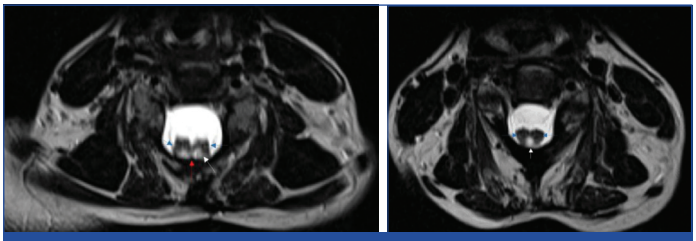
A 47-year-old male patient presented with complaints of low back pain on excessive walking, radiating to right lower limb since one month. He complained of aggravation of pain with activity or excessive standing which was relieved by rest. On examination, patient had features of muscle wasting in bilateral upper and lower limbs and reduction in power of bilateral upper and lower limbs, right upper limb- 3/5, left upper limb- 3/5, right lower limb- 3/5, left lower limb-3/5. Patient had exaggerated reflexes and extensor plantar reflex on examination.

Complete Blood Count (CBC), Liver Function Test (LFT) and Renal Function Test (RFT) of patient were within normal limit. He had been operated 15 years ago, details and records of which were not available for present review. Magnetic Resonance Imaging (MRI) done for the patient revealed postoperative changes in cervical and lumbar spine. The cord showed diastematomyelia from C3 to C5 (cervical vertebrae) with kinking of cord at this level attached to the posterior dura. Posterior subarachnoid space at this level is completely effaced with no evidence of Cerebrospinal Fluid (CSF) posterior to the cord. The Time (T2) hyperintense signal is noted within the posterior cord at this level associated with mild cord atrophy which suggested changes of myelomalacia [Table/Fig-1-3].

Hence, on basis of clinical evaluation and MRI diagnosis of postoperative dorsal spinal cord herniation and myelomalacia in the cervical spine associated with diastematomyelia was made. Presence of myelomalacia and clinical evaluation also suggested that the disease process was chronic. Patient was advised surgical treatment to prevent further neurological deterioration, but patient denied to undergo any surgical treatment, and hence is being managed conservatively till date. On follow-up, there is no further neurological deterioration and symptoms have been reduced, however, no follow-up MRI spine has been done.



[Table/Fig-1]: Time (T2) sagittal image of cervical spine, showing kinking of spinal cord with effaced posterior subarachnoid space Cerebrospinal Fluid (CSF) signal (white arrow). Postoperative changes are seen cervical spine posteriorly (red arrow).



[Table/Fig-2]: Time (T2) Axial image of cervical spine showing diastematomyelia. Posteriorly displaced hemi cords (blue arrows), effaced posterior subarachnoid space CSF signal (red arrow) and shows abnormal T2 hyperintense signal within dorsally (white arrow); **[Table/Fig-3]:** T2 axial image of cervical spine (at lower level than [Table/Fig-2]) showing diastematomyelia (blue arrows). There is loss of CSF signal dorsal to cord (white arrow). However, no hyperintense signal was noted in the spine as was observed before.

DISCUSSION

Transdural spinal cord herniation is rare and under reported caused by herniation of spinal cord through a defect in the dura. Ventral or ventro-lateral cord herniation is relatively more common and a well described entity now, with over 100 cases reported so far. Dorsal or posterior spinal cord herniation is very rare. Only six cases in the cervical spine and four in the thoracic or lumbar spine have been reported so far [1]. It is nonetheless a recognised cause of myelopathy with dural defect, either acquired post surgery or post-trauma or idiopathic in cause and congenital [1]. Transdural spinal cord herniation is a rare, but treatable cause of slowly progressive myelopathy. They often present in a delayed fashion. Spinal cord herniation as a result of dural defect is classified into three groups:

- Traumatic
- Postoperative
- Congenital

The mechanism of herniation varies with its cause. In traumatic cases, the herniation occurs due to damage to the dura mater. It is more common in the ventral part of the dural sac. In postoperative cases, it occurs through a defect caused by surgery or incomplete suture. They usually follow posterior approaches with unintended or intended dural opening. These are more common in the dorsal dura mater [2,3]. This results because of inflammatory response to the dural defect which results in adhesion of spinal cord to the dural defect. And the progressive pulsation of cerebrospinal fluid provides a continuous and slow pressure gradient that leads to spinal cord eroding the adjacent dura and then herniated from the thecal sac [4].

In congenital cases, it is reported to be associated with intradural arachnoid cyst causing thinning and tear of the dura [5]. In the present case, the dural defect through which cord herniation occurred could have been created at previous surgery. Once the herniation starts to occur, CSF drainage around this region would be difficult leading to further swelling and incarceration which accounts for gradual and progressive onset of symptoms. CSF pressure difference between the herniated cord and rest of the cord could lead to alteration of blood flow and fluid movement worsening the cord oedema and herniation [6].

Severe post-traumatic spinal cord herniation should be treated with surgery. Operation can help the reduction and decompression of the spinal cord and prevent its recurrence. Physical examination and radiological evidences such as MRI, myelography and Computerised Tomography (CT) myelography are also useful for the diagnosis of the disease. Once the diagnosis of post-traumatic spinal cord herniation is made, surgical treatment should be initiated immediately [7].

Idiopathic Spinal Cord Herniation (ISCH) should be differentiated from various CSF-isointense spaces occupying intramural extra medullary lesions by combined assessment using multi-modality imaging approach and patients clinical symptoms [8]. MRI is the investigation of choice for the diagnosis of ISCH. Phase contrast MRI and CT myelogram can be used to differentiate between a dorsal arachnoid cyst and widened dorsal subarachnoid space and also to confirm the diagnosis of ISCH. In ISCH, CT myelogram demonstrates obliteration of CSF space ventral to the spinal cord with no differential contrast retention dorsal to the cord and phase contrast MRI demonstrates normal dorsal pulsatile CSF flow [9].

There have been previously reported six cases having posterior or dorso-lateral cord herniation in the cervical spine, and all of these cases occurred after surgery [10-15]. Among those studied, were four individuals who had received dural opening during surgery (by posterior laminectomy), as well as two patients who had unintentionally opened dura peri or postsurgery. The most prevalent presentation was myelopathy, with a time lag of up to 18 years between the initial procedure and the first presentation. Improvement was achieved in virtually all cases following surgical correction. However, the authors weren't able to find a case as unique as the present one discussed, having posterior spinal cord herniation involving the cervical spinal cord with diastematomyelia.

CONCLUSION(S)

Posterior spinal cord herniation is a rare occurrence, but can occur in any case with a posterior dural defect of varying causes. If diagnosed, prompt surgical reduction of the hernia is advised as in this instance, to prevent neurological deterioration and potentially reverse any deficit. Patients should be followed closely as long-term recurrences may occur.

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PARTICULARS OF CONTRIBUTORS:
1. Resident, Department of Radiodiagnosis, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pune, Maharashtra, India.
2. Fellow in Cardiac and MR Imaging, Department of Radiodiagnosis, Sri Satya Sai Institute of Higher Medical Sciences, Bangalore, Karnataka, India.
3. Assistant Professor, Department of Radiodiagnosis, B.J. Medical College, Pune, Maharashtra, India.
4. Resident, Department of Radiodiagnosis, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pune, Maharashtra, India.
5. Resident, Department of Radiodiagnosis, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pune, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Vishal Walasangikar,
Assistant Professor, Department of Radiology, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pimpri, Pune, Maharashtra, India.
E-mail: vishalwalasangikar@gmail.com

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