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Delayed Onset Brown Sequard Syndrome in Congenital Spinal Cord Hernia: Evaluated with Diffusion and Tensor Imaging

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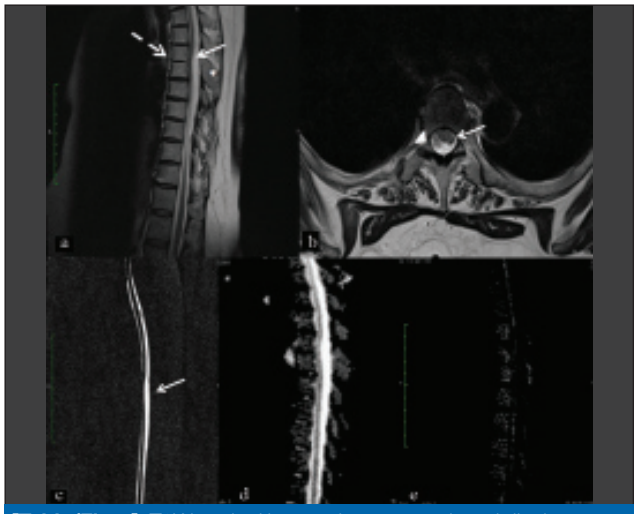
Spinal cord herniation is a commonly undiagnosed entity, which is seen on routine MRI imaging of the spine. We present a case of spinal cord hernia evaluated on routine and advanced MRI sequences using diffusion tensor imaging. There is association of the congenital block vertebra at the same level highlighting the congenital origin of the herniation of the dorsal cord.

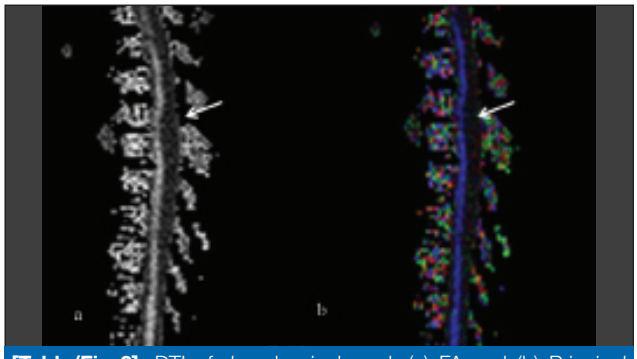
A 40-year-old female presented with back pain and mild weakness in left leg from last two years. The disease course was stable since inception and new symptoms developed over a period of two months, in the form of loss of proprioception in the same limb. There was no antecedent history of trauma or infection. Neurological examination revealed MRC Grade 4 power (Medical Research Council grading) with loss of pin prick sensation in the left leg and hypoaesthesia in right leg correlating with Brown Sequard Syndrome. Bladder and bowel habits were normal. The patient underwent Magnetic Resonance Imaging (MRI) of the brain using 1.5 T MRI scanner (Magnetom Avanto, Erlangen, Germany).

MRI sequences were showing congenital block of D5 and D6 vertebral bodies and their posterior elements with ventral approximation of the spinal cord and kinking. Axial sections showed left anterolateral displacement of the cord with expanded posterior subarachnoid space and CSF flow void ruling out possibility of dorsal arachnoid cyst or mass [Table/Fig-1a-e]. Diffusion MRI revealed normal cord intensity and facilitation in the dorsal CSF space, confirming the inference. Further, imaging was done using Diffusion Tensor Imaging (DTI) to rule out any early signs of cord intensity changes [Table/Fig-2].

Spinal cord herniation is rare and unpredictable phenomenon which is coming under consideration due to progressively increasing role of MRI for spine related problems. Most of the cases remain undiagnosed, probably due to their slowly progressive, non compromising symptoms and sometimes due to lack of imaging facilities and knowledge about the entity.



[Table/Fig-1]: T2W sagittal image shows ventral cord displacement (solid arrow) at D5-D6 level (a) with congenital block vertebra (--) & (*). Axial sections (b) shows anterior approximation of the cord & CSF flow void (arrow head). MR myelography (c) showing anterior shift of the cord with ventral CSF cut-off. Diffusion Weighted Imaging of the cord; ADC map (d) and b0 image (e) showing normal cord intensity without any lesion in posterior CSF space.



[Table/Fig-2]: DTI of dorsal spinal cord. (a) FA and (b) Principal eigenvector map in the sagittal plane. The blue color represents the principal eigenvector aligned along the head-foot direction with anterior displacment of the fibres & maintained color homogeneity.

The pathogenesis of spinal cord herniation is controversial, including congenital deficiency of the dura, antecedent history of trauma, duplication of the ventral dura mater and pressure erosion of the dura [1].

The disease typically manifests in middle aged and elderly patients with female predilection of 2:1 and predominantly occurring from D3 to D7 levels [2]. This may be due to kyphosis of thoracic spine, physiological anterior position of the thoracic spinal cord and pressure changes secondary to cardiac and pulmonary pulsations. Herniation of the cord is noted through a dural defect which can be of congenital, traumatic or iatrogenic origin. This case is being reported, with supportive imaging features of congenital origin.

MRI findings and CT Myelography, have classified cord herniation into three types: Type K (kink), as in our case; Type D (Discontinuous), in which the spinal cord completely disappears at a herniated site; and Type P (Protrusion), in which the subarachnoid space of the anterior spinal cord disappears with no kink [3]. Both later types are relatively uncommon.

Patients with spinal cord herniation show progressive thoracic myelopathy with onset several years prior to the diagnosis [4]. The presenting features are those of Brown Sequard Syndrome in the form of ipsilateral hemiparesis with loss of proprioception, and contralateral loss of pain and temperature sensation [5]. In some cases, there may be spastic paraparesis, bladder bowel dysfunction and intercostal pain. However, the diagnosis relies on MRI confirmation.

There is increasing role of phase contrast MRI to look for CSF flow velocity in the expanded posterior subarachnoid space, which is helpful in ruling out dorsal arachnoid cyst, the most common differential diagnosis of the entity [6]. In our case, the dorsal CSF flow void was present (not seen in dorsal cyst), thus avoiding necessity of using phase contrast MRI. Further imaging with diffusion weighted imaging was done to rule out any subtle intensity changes in the cord and further tensor imaging was also done. However, no disturbance in the diffusion parameters and anisotropy was appreciable. Cortico-spinal and spino-thalamic tractography can be done and correlated with the symptoms.

A diagnosis of congenital spinal cord herniation was made after excluding the space occupying lesion in posterior subarachnoid space. Notably the D5-D6 block vertebra showing rudimentary disc and fusion of posterior elements were indicative of congenital cause due to abnormal embryonic deficiency of the dura. Surgical repositioning of the cord and dural patch procedure was offered but the patient declined and chose to be maintained on physiotherapy and conservative treatment. Two years from then, disease showed stable course with minimal functional improvement on physiotherapy.

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