

Spinal cord herniation into an extradural arachnoid cyst in a patient with multiple sclerosis

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Abstract

Idiopathic spinal cord herniation (SCH) is a rare condition, occurring in thoracic spinal cord through an anterior dural defect. We report a case of posterior thoracic SCH into extradural arachnoid cyst in a patient with multiple sclerosis. A 28 year old lady presented with recurrent bilateral visual loss, which improved with steroids. Two weeks after the onset of illness, she developed rapidly evolving spastic paraplegia. MRI of brain was suggestive of demyelination, while MRI spine revealed an extradural arachnoid cyst at D6-9 vertebral level, posterior to dural tube, with herniation of spinal cord into it. Following laminectomy and excision of arachnoid cyst, reduction of SCH and duraplasty, she improved in lower limb function. She later had another episode of demyelination with visual loss and quadriplegia, which improved partially with steroids. This is the first report of posterior spinal cord herniation in a patient with relapsing remitting multiple sclerosis.

INTRODUCTION

Idiopathic spinal cord herniation (SCH) is a rare entity, with only a few reports in literature. Almost all reported cases had anterior herniation of cord, occurring at the thoracic level.¹ The recognition of this condition has been greatly influenced by introduction of MRI, with only two cases reported in the pre-MRI era.² We report a patient with a rare association of relapsing remitting multiple sclerosis and posterior spinal cord herniation into an extradural arachnoid cyst.

CASE REPORT

A 28 year old woman presented with rapidly progressive visual loss in both eyes, which improved partially over two weeks with parenteral methylprednisolone (1 gm daily for 5 days). About 2 weeks after the onset of visual loss, she developed acute onset paraparesis, progressing to involve the truncal muscle weakness and urinary retention in the next three days. She had past history of acute bilateral visual loss one year ago that improved completely in a month with steroid injections. However, she was not investigated then.

Examination revealed that visual acuity was finger counting at 2 feet in right eye and only light perception in left eye. Optic fundi examination revealed pale discs. She had spastic paraparesis with muscle strength of 0/5 and 1/5

(MRC grade) in right and left lower extremities respectively. There was 50% hypoesthesia to all modalities below D7 dermatome. Joint position sense was impaired at both the toes and ankles. A provisional clinical diagnosis of relapsing remitting demyelinating illness possibly multiple sclerosis (MS) was considered affecting the optic nerves and spinal cord.

The routine hematological and serum biochemical tests were normal. The CSF analysis revealed a cell count of 5 lymphocytes/cu.mm and glucose of 101 mg% and protein of 47 mg%. There were oligoclonal bands in the CSF. Serum RA factor, ANA, VDRL and HIV were non-reactive. Visual evoked potential (VEP) study showed absent response on left side and poor waveform on right side with P100 of 106.5 msec (Upper limit of normal: 108 msec). The brainstem auditory evoked response was normal. Somatosensory evoked potential (SSEP) from left posterior tibial and right median nerves were normal. The MRI of brain revealed multiple non-enhancing periventricular subcortical flame shaped hyperintensities in T₂W images, which were isointense on T₁W images and not inverting on FLAIR images (Figure 1). A few lesions involved the calloso-septal interface. The MRI of brain was suggestive of multiple sclerosis. The MRI of spine revealed atrophy of the thoracic cord. A single lesion in the extradural plane was noted extending from D6 to D9 levels, dorsal to the dural tube, uniformly hypointense

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Figure 1. MRI of brain showing multiple periventricular subcortical flame shaped hyperintensities in T₂W images and FLAIR images.

in T₁W and hyperintense in T₂W images, causing significant cord compression. A part of the thoracic spinal cord had herniated posteriorly into the lesion (Figure 2). A possibility of thoracic extradural arachnoid cyst with posterior spinal cord herniation was considered.

She underwent surgical excision of the arachnoid cyst and reduction of the hernia. Intraoperatively, a dural defect was observed in the posterior spinal dura with CSF leak and herniation of a part of the spinal cord through the defect. The arachnoid cyst was excised, the neck

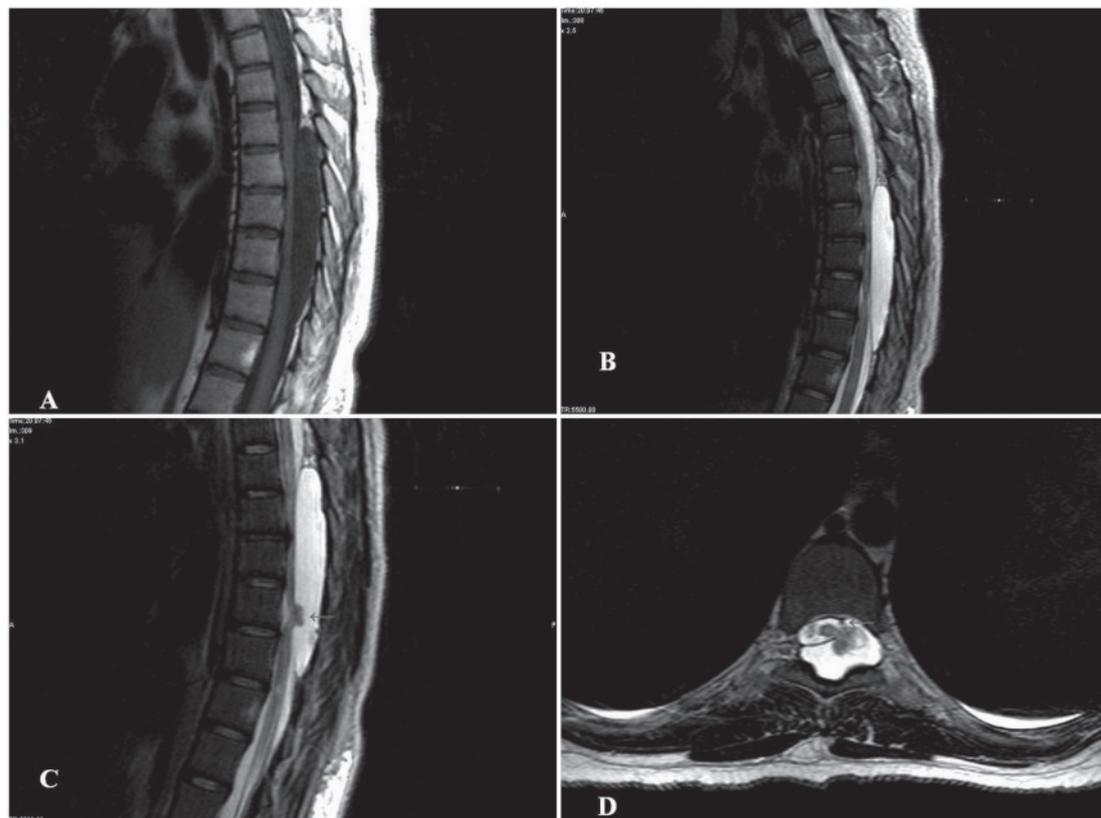


Figure 2. Fig 2 a, b: MRI of spine showing an extradural arachnoid cyst, dorsal to the dural tube, causing significant cord compression; the arrow in Fig 2b indicates the extradural location of the cyst. Fig 2 c, d: MRI of spine, sagittal and axial sections, showing herniation of a part of the thoracic spinal cord posteriorly into the cyst (marked by arrows). Multiple discrete short segment signal changes are also noted in the dorsal cord.

of the dural ring was incised to widen the ring and the herniated spinal cord was released and internalized into dural cavity. The dural defect was repaired with duraplasty. The herniated spinal cord tissue looked globular and congested. Post operatively, the patient's motor power improved. At discharge 2 weeks post surgery, the muscle power was 4/5 in right lower limb and 2/5 in left lower limb.

About 10 weeks later, she again presented with acute onset of impaired vision followed by weakness of all four limbs and dysarthria. She had only perception of light in both the eyes, had spastic quadriplegia and sensory loss up to C5 dermatome. She received pulse methylprednisolone and oral prednisolone for maintenance. At follow up, six months later, she had sustained improvement and was able to ambulate independently. A rare association of relapsing remitting multiple sclerosis, with posterior spinal cord herniation into an extradural arachnoid cyst in the dorsal region was considered to be the diagnosis.

DISCUSSION

This patient has relapsing neurological disease involving the optic nerves and cervical spinal cord, which is consistent with the diagnosis of relapsing remitting multiple sclerosis. She also has thoracic spinal cord herniation shown by MRI and treated surgically. Spinal cord herniation is a rare entity, described initially in 1974 by Wortzman *et al*³ and ever since there have been reports of isolated case or small series. Around 80 cases have been reported till 2006.⁴ Various theories of aetiopathogenesis of SCH have been proposed, based on the limited experience. The condition essentially is a herniation of the cord tissue through a dural defect and based on the etiology of dural defect, it has been classified as idiopathic, post-traumatic and iatrogenic.⁴ The dural defect in idiopathic cases could result from various causes, namely congenital, erosion of the dura due to prolapsed intervertebral disc or due to trivial trauma. In the majority of cases, the dural defect was considered to be congenital. The most common location is in the thoracic region. The natural curvature in the thoracic region results in the anterior displacement of the spinal cord, which becomes adherent to the dural defect. Further progression of the herniation is aided by CSF pulsations.

In almost all cases, the dural defect is noted in the anterior or anterolateral dura. In our patient,

the defect in the dura was found posteriorly. Only two cases have been reported earlier in the literature with posterior dural defect.^{4,5} As in the present case, the posterior spinal cord herniation has been associated with an extradural arachnoid cyst. The pathogenesis of posterior spinal cord herniation is unclear. The dural defect in these cases is most probably congenital; none of these three patients had any prior history of trauma. The continuous pulsations of CSF result in the prolapse of the arachnoid causing an extradural arachnoid cyst. Herniation of the spinal cord and further progression has been attributed to the differential pressure between the spinal subarachnoid space and the arachnoid cyst.^{4,5} Our patient had associated relapsing remitting illness, possibly multiple sclerosis affecting the optic nerves and spinal cord. We thought that the atrophy of spinal cord precipitated the herniation of spinal cord into a pre existing extradural arachnoid cyst. This is the first case report describing intrinsic cord pathology from demyelination associated with spinal cord herniation.

The herniated spinal cord tissue appeared knob like, gliotic and congested in our patient. We preserved the tissue since it might still be functional. Although some authors had performed biopsy of this tissue suspecting neoplasm, it had revealed only gliosis, neural tissue and reactive astrocytes.^{6,7} The management of SCH is essentially surgical release of the herniated cord. A few cases managed conservatively demonstrated minimal deterioration over long duration, but had not improved either, whereas most of the operated patients improved following surgery.² This underscores the importance of recognition of this often undiagnosed entity and early surgical treatment. Although our patient improved in limb function immediately following surgery, she had another episode of demyelination affecting cervical cord and developed quadriparesis, which improved with steroids.

In conclusion, idiopathic spinal cord herniation is an uncommon cause of spastic paraplegia. Further, this is the only report of a rare coincidental association of relapsing remitting multiple sclerosis with posterior spinal cord herniation. An accurate diagnosis and surgical treatment is facilitated by MRI.

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