CASE REPORT

Idiopathic spinal cord herniation of the cervical cord: unusual cause of proximal muscle weakness in upper limbs

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SUMMARY

Idiopathic spinal cord herniation (ISCH) is a recognised rare cause of progressive and potentially curable myelopathy. Around 170 cases have been described in the literature, all to be found between the T2 and T8 vertebrae. We report a case of ISCH in the cervical region. A 23-year-old man with no history of trauma presented with a 6-year history of bilateral mild resting hand tremor and left scapular pain radiating to the left arm for a duration of 8 months. Nerve conduction studies showed some denervation changes of the upper limbs and bulbar regions. MRI of the spine showed anterior midline herniation of the spinal cord at the level of C7 vertebra with an associated collection of cerebrospinal fluid in the extradural space in the cervical region. Owing to the non-progressive nature of symptoms, currently the patient is managed conservatively.

BACKGROUND

Idiopathic spinal cord herniation (ISCH) is a recognised rare cause of progressive and potentially curable myelopathy. This condition was first described in 1974 by Wortzman *et al.*¹ Around 170 cases of ISCH have been described in the literature, and all these lesions were found in the thoracic spine with the majority between the T2 and T8 vertebrae.^{2 3}

ISCH has not previously been reported in the cervical spine. This case report describes an idiopathic anterior herniation of the cervical spinal cord in a 23-year-old man.

CASE PRESENTATION

A 23-year-old man presented with a 6-year history of bilateral mild resting hand tremor and left scapular pain radiating to the left arm for a duration of 8 months. In retrospect, the patient reported difficulty with fine motor skills including impairment of speed of writing 5 years prior to current presentation. In spite of this minimal disability, he was successful at the competitive written examinations and able to enter university. There was no history of trauma, surgery or any medical conditions.

On examination, the patient had symmetrical bilateral proximal muscle wasting in upper limbs with muscle power of 4/5. The distal upper limb muscle and lower limb muscle power was 5/5 with normal reflexes. There was no sensory impairment. The rest of the neurological examination was unremarkable.

INVESTIGATIONS

Initial basic investigations including thyroid function and erythrocyte sedimentation rate were normal. Nerve conduction studies showed some denervation changes of the upper limbs and bulbar regions, suggestive of anterior horn cell disease.

Sagittal MRI of the spine revealed anterior displacement and kinking of the spinal cord at the level of the C7 vertebra with complete focal obliteration of the cerebrospinal fluid (CSF) space anterior and widening of the CSF space posterior to the spinal cord. The dura was displaced posteriorly from C1 to C7 level by a 3.8 mm thick collection of CSF between the dura and posterior longitudinal ligament delineating the dura between the subarachnoid and subdural CSF (figures 1 and figure 2).

Axial image at C5 level demonstrated posterior displacement of the thecal sac and spinal cord by a 3.8 mm thick CSF collection in the subdural space. Normal dura was clearly outlined by CSF in the subarachnoid and subdural spaces (figure 3).

Axial images of the spine at the level of C7 vertebra revealed a 5 mm defect in the dura anteriorly on midline, with herniation of spinal cord anteriorly to the extradural space, through the defect in

Figure 1 T1-weighted sagittal image, focal ventral displacement and angulation of the cervical spinal cord at C7 level.

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Figure 2 T2-weighted sagittal image, focal ventral displacement and angulation of the cervical spinal cord at C7 level. Thick collection of CSF of 3.8 mm between the dura and posterior longitudinal ligament at C1 to C7 level. CSF, cerebrospinal fluid.

the dura (figure 4). There was no signal abnormality in the spinal cord to suggest gliosis.

Cervical and upper dorsal vertebrae and intervertebral discs were normal.

DISCUSSION

ISCH is a recognised cause of progressive myelopathy with reported incidence of <200 cases, almost exclusively involving

Figure 3 T2-weighted images at C5 level demonstrate posterior displacement of the thecal sac and the spinal cord by a 3.8 mm thick CSF collection in the subdural space. Normal dura is clearly outlined by CSF in the subarachnoid and subdural spaces. The spinal cord is normal. CSF, cerebrospinal fluid.

Figure 4 Axial T2-weighted image through the cervical spinal cord at C7 level demonstrates spinal cord herniation through the dura. The anterior surface of the spinal cord is adjacent to the posterior surface of the C7 vertebral body. The signal intensity of the spinal cord is normal.

the thoracic spine.³ It most commonly presents as Brown-Sequard syndrome.⁴ Other described clinical features are paraparesis, sensory disturbance and bowel and bladder dysfunction.³

Reported incidence is higher in females between the ages 25 and 75 years with a mean of 49 years.³ ISCH has been reported between T2-T8 with T4-T5 being the most common site of herniation.^{2 5} A review of 30 cases of ventral herniation, conducted by Watters *et al*, concluded that 23% of the cases were of post-traumatic origin. In addition, traumatic and iatrogenic spinal cord herniation are reported primarily posterior to the cord as opposed to anterior herniation in ISCH.⁶ Other reported cases included iatrogenic opening of the dura secondary to incomplete closure following spinal surgery.⁷

Presence of idiopathic anterior herniation of the spinal cord in the cervical spine in the current case—a 23-year-old patient who had had symptoms since the age of 16 years in the absence of both trauma and previous history of surgery—favours a congenital aetiology.

Pathogenesis of this condition is not clearly identified at present. Several theories have been proposed in the literature. These include a pre-existing ventral pseudomeningocele, a meningeal diverticulum or an extradural arachnoid cyst² where cord herniation occurs through a pre-existing lesion.⁸

Marwan *et al*, in 2004, proposed that the origin of the formation of the defect in the dura is an inflammatory process involving the spinal cord and meninges leading to the adhesion of the cord to the ventral dura leading to herniation of the cord.⁷

MRI is the investigation of choice for the diagnosis of idiopathic spinal cord herniation. As described in the current literature, sagittal sections demonstrate a C or S-shaped kink of the thoracic spinal cord, with a secondary enlargement of the dorsal subarachnoid space. The axial images may show a defect in the dura anterior to the herniation as well as the herniation itself.⁹ This is consistent with the radiological findings of the current case in the cervical spine.

Since ISCH was first described, the majority of cases of ISCH reported initially were due to incidental surgical findings, but

now, as a result of widespread availability of MRI, more cases are being accurately diagnosed preoperatively.¹⁰

It is fair to hypothesise that, although ISCH has not been previously reported in the cervical region, lack of detection is merely due to lack of awareness of the possibility of this condition occurring in the cervical region.

Two strategies have been described in the management of ISCH. One involves the closure of the defect following repositioning of the spinal cord. The other procedure involves surgical widening of the defect in the dura to reduce the risk of strangulation. Conservative management with watchful follow-up of symptoms has also been described in the literature.^{10 11} Massicotte *et al*¹¹ reviewed eight cases of ISCH, with four stable on conservative management, where watchful observation was carried out.

The present case is managed conservatively, with close clinical observation of symptom progression. Given the non-progressive nature of symptoms and that the patient is currently undergoing

higher education at university, a conservative approach is suitable.

Awareness of this condition, and accurate early diagnosis and early correction of the anatomical defect may cure these patients without any residual neurological deficit.

Contributors This case was supervised by DR and LM, while carrying out their medical elective in radiology in Sri Lanka. The case report was drafted/written by DR and LM, and supervised by SM.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- Idiopathic spinal cord herniation (ISCH) of the cervical region has not been previously described in the literature. This case report shows ISCH occurring in the cervical region.
- MRI is the gold standard in diagnosis of this condition.
- MRI shows a C or S-shaped kink in the spinal cord with enlargement of the dorsal subarachnoid space.
- Early diagnosis is pivotal with possible surgical correction of the anatomical abnormality offering potential curability, or conservative management with watchful observation of symptom progression.

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