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Case Report

Idiopathic spinal cord herniation syndrome: a case report

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ABSTRACT

Idiopathic spinal cord herniation syndrome is a rare entity. Only 17 cases have been reported in literature so far. They typically occur in the dorsal region of the spinal cord, spontaneously, through a defect/rent in the anterior Dura. Majority present in middle age women, with progressive paraparesis/asymmetrical motor weakness in lower limbs.

Keywords: Dural defect, Spinal cord herniation, Thoracic/dorsal region, Arachnoid cyst

INTRODUCTION

Idiopathic spinal cord herniation syndrome is a rare lesion. Predominantly seen in the middle aged, females being most affected. A vent in the Dura results in dorsal compression of the spinal cord. Patients present with paraparesis/asymmetrical weakness lower limbs, with or without sensory symptoms and bladder involvement. MRI forms the mainstay of the diagnosis. Variable results are seen with surgery.

CASE REPORT

Mrs. KD, 38 year old female,[2,5,6,14] from West Godavari district of AP, presented with difficulty in walking since one year. She was treated by local practitioners with no relief. Later she was evaluated by an orthopedic surgeon and then was referred.

There was history of gradually progressing weakness in both lower limbs initially, followed by asymmetrical weakness, which was more in left lower limb. Gradually there was stiffness, gait disturbance, and swaying to sides on walking. However, there was no bladder & bowel involvement. Conspicuously there was no pain. There was no H/O fever / trauma.[7,8]

Examination revealed wasting of thigh muscles & calf muscles on left side, ↑ tone, more on left side, spastic gait with swaying. DTR brisk on left, with plantar up going.

The MRI scan[1] showed focal atrophy of spinal cord at D4 level, with a ventral apposition of spinal cord[4,7,8] at that level, close to the posterior surface of vertebral body with an acute kink. There was no specific Dural defect[4,6] except a focal hyper intense signal[1] close to D4 body.[9,12] The kink[14] resulted in focal widening of posterior subarachnoidspace,[1,3,6,10] with pooling of CSF & pulsation artifacts. Post Gadolinium sequences do not show any enhancing lesion/an arachnoid cyst. The signal pattern of the spinal cord is normal, except it is very thinned out.

A pre-operative diagnosis of ISCH was established based on radiological evaluation. After counseling of the patient & her family, she was taken for surgery under general anesthesia. A dorsal 4, 5 & partial 3 laminectomy was performed. There was no bulge of the thecal sac nor was

it tense. Dura was opened in the midline. A gush of CSF was noted. Controlled suction was applied to the free CSF. The cord was found to be atrophic, thinned out, flat & ribbon like. The same was gently retracted to right side, after cutting the dentate ligaments, to prevent pressure during retraction & facilitate easy retraction. A subtle gentle kink of the cord was released by delicate retraction of the cord. A probed dissector was introduced to search for the anterior defect.[6] No obvious defect could be identified, but a small dent felt anteriorly.[10] A piece of G patch,[14] cut triangularly, negotiated & passed from one side to other, underneath the cord & overlying the Dural dent. The Dura closed in a water tight manner. Postoperatively, there was mild deterioration of bladder function,[13] which was managed conservatively with post-operative steroids& bladder training.

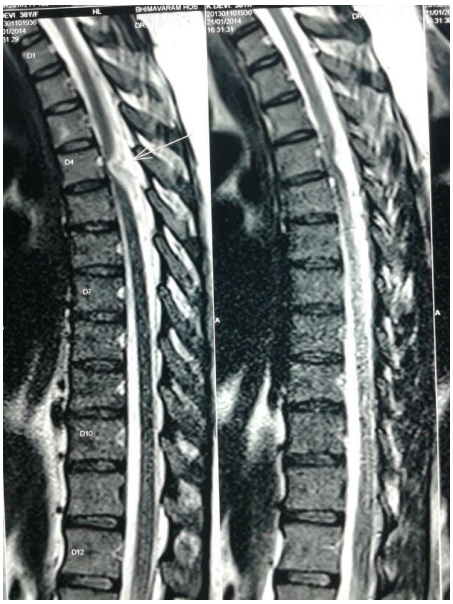

Figure 1: Showing kink cord at D4 level. Anteriorly with widened subarachnoid space posteriorly, filled with CSF in sagittal section T2 WI.

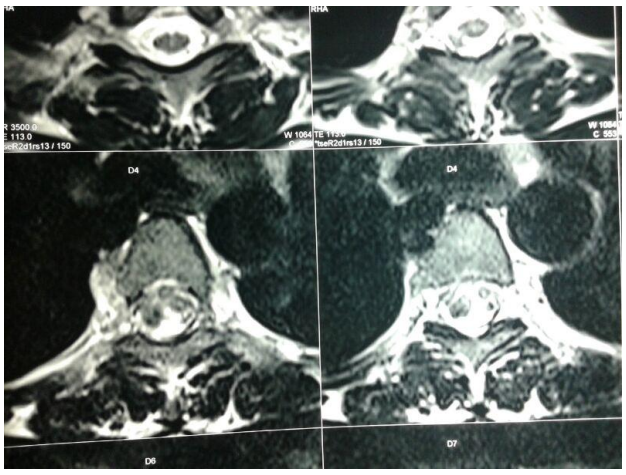

Figure 2: Axial sections of MRI showing herniation/close abutting of cord close to D4.

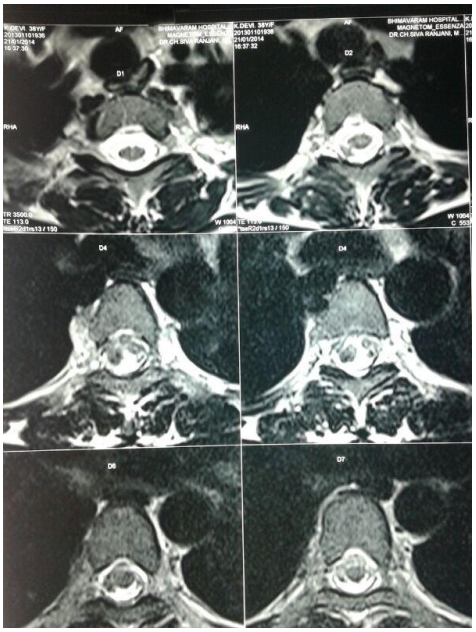

Figure 3: Axial MRI with herniation of cord anteriorly and corresponding dilatation of subarachnoid space.

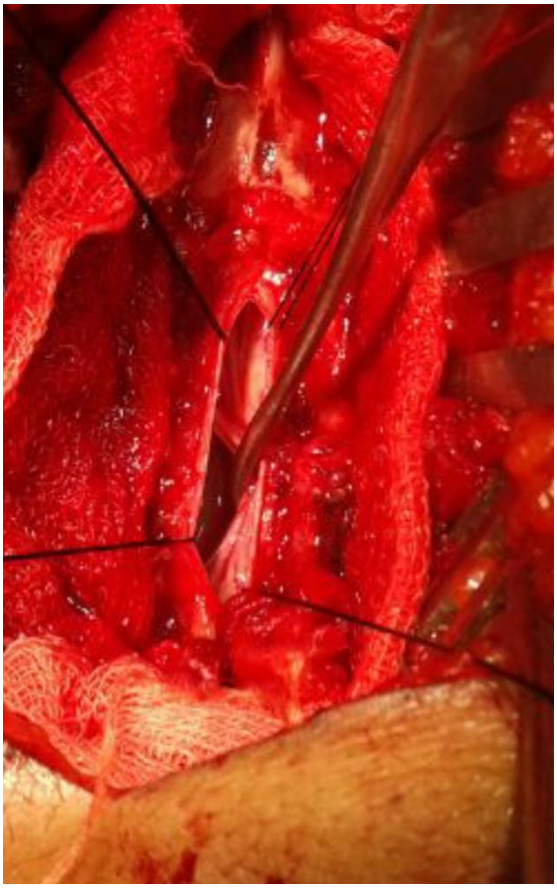

Figure 4: Intra-operative photo showing thinned out dorsal cord & empty space seen anterior to the retracted cord. The dent was felt with the dissector cum retractor, after cutting dentate ligaments and lateral retraction.

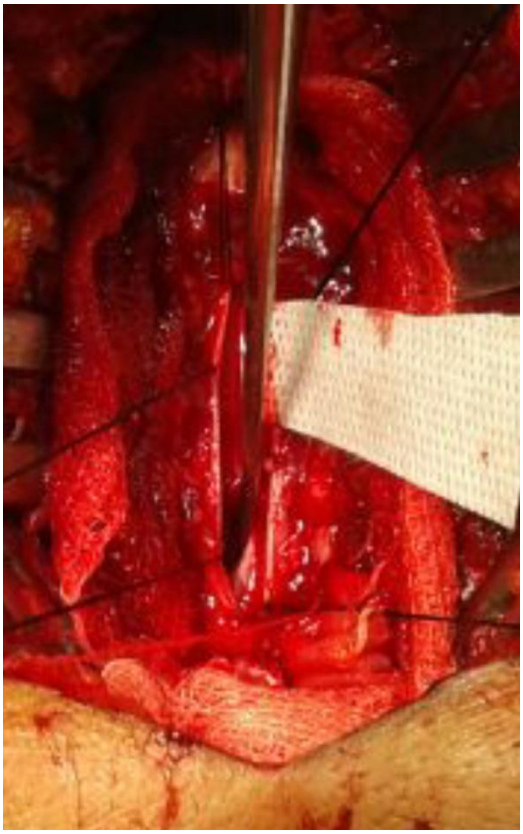

Figure 5: Intra-operative photo showing introduction of the synthetic G patch piece passed anterior to the cord at the level of the defect, as reinforcement after reducing the cord.

DISCUSSION

Idiopathic spinal cord herniation syndrome is an unusual cause of progressive myelopathy. It is a very rare clinical entity[3] which can be confirmed by a MRI/CT myelogram.

The cause of the defect in the anterior duramater is not known, classified as idiopathic, post-traumatic or iatrogenic.

Element of suspicion of this syndrome in middle aged patients presenting with paraparesis/motor weakness is useful in prevention of irreversible damage which is likely to occur if delayed.

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Ethical approval: Not required

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