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CASE REPORT

Split Cord Malformation Type 2 Complicated by Presence of Tuberculous Arachnoiditis

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ABSTRACT

We present a rare case of split cord malformation (SCM) type II complicated by presence of tuberculous arachnoiditis without any history of systemic tuberculosis or vertebral body lesions. Diagnosis was made based on intraoperative findings and was confirmed by histopathology. Surgical decompression along with a combination of steroid and antitubercular therapy resulted in a good outcome. Clinical features, magnetic resonance imaging (MRI), intraoperative findings, pathology and the relevant literature are herein discussed.

Keywords: Spinal arachnoiditis, Split cord malformation, Tuberculous radiculomyelopathy.

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INTRODUCTION

Split cord malformations (SCMs) are rare congenital anomalies in which the cord is split over a portion of its length to form a double neural tube in a single or separate dural sacs. Majority of these cases present in early childhood, with neurocutaneous stigmata being an early presenting feature.[1] Pang et al proposed a unified theory of embryogenesis of SCM, in which they described the origin of all SCM as an error occurring due incomplete closure of the accessory neuroenteric canal.[2] This canal is invested with mesenchymal tissue splitting the notochord and the neural plate to form a bifid spinal cord separated by bony or fibrous tissue. To the best of our knowledge, this is the first reported case of SCM complicated by tuberculous arachnoiditis.

CASE REPORT

A 40-year-old diabetic female presented with dull aching, progressively increasing back pain, which was radiating to the left lower limb along the posterior-lateral aspect of the buttock up to the foot for 4 years. This was accompanied with tingling and paresthesia of both the lower limbs. A week before coming to us, she developed asymmetric weakness in both the lower limbs, along with bladder disturbances and constipation. Family history of exposure to pulmonary tuberculosis was noted. On clinical examination, she had asymmetric lower motor neuron type paraparesis with bladder and bowel involvement. Sensory loss was present below L1 dermatome, including perianal region. X-ray revealed spina bifida at L5-S1 level. She had normal hemogram and elevated erythrocyte sedimention rate (ESR). Magnetic resonance imaging (MRI) of the lumbosacral region revealed spinal cord extending upto L3/L4 level. Hemicord at L3 level was noted with tethering. Cauda equine fibers were found to be adherent to lateral margin of dura of each side at L4, L5 (Fig. 1).

She underwent L3 to L5 laminectomy and exploration. Peroperatively, spinal cord was noted to be split at L3 level inside a single dural sac (Fig. 2). No intervening bony spur or fibrocartilaginous septum was noted. Arachnoid was found to be partially calcified, rubbery in consistency, completely plastering the split spinal cord and exiting nerve roots to anterolateral wall of dural sac. Histopathology of this thickened milky white membrane revealed it to be fibrous connective tissue focally covered by meningothelial cells and multinucleate Langhan's giant cells with signs of focal chronic inflammation. The patient was started on a combination of antitubercular medication and steroids. Three months later, she is able to walk without support and has regained partial bladder control.

DISCUSSION

Based on this unified theory, SCM is divided into two types: type I consists of two hemicords, each contained within its own dural sheath and separated by a median bony spur, and type II consists of two hemicord housed in a single dural tube separated by a fibrous median septum. In our case, we noted a split cord housed in single dural tube, but lacking the intervening mesenchymal tissue. Katoh et al reported a similar case of SCMII without an intervening fibrous septum.[3] We believe this variation is a subset of type II malformations in which the fibrous septum may have regressed with age. If not, similar

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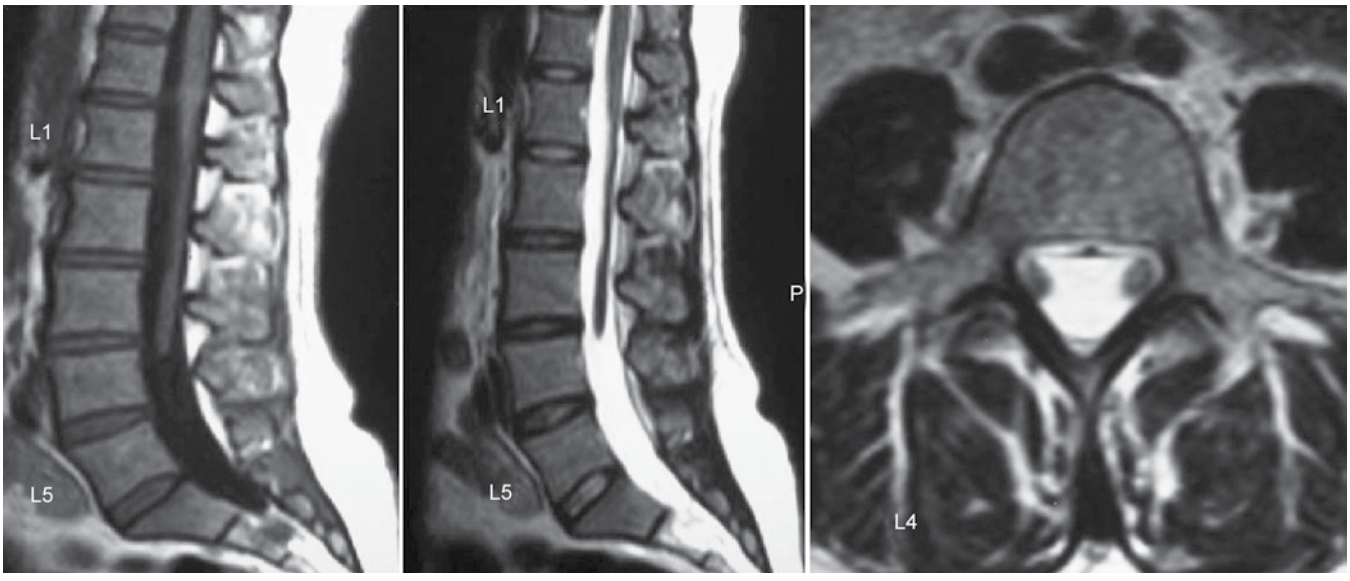

Fig. 1: T1W and T2W sagittal images showing spinal cord extending up to L3 vertebral body and T2W axial image demonstrating empty sac appearance

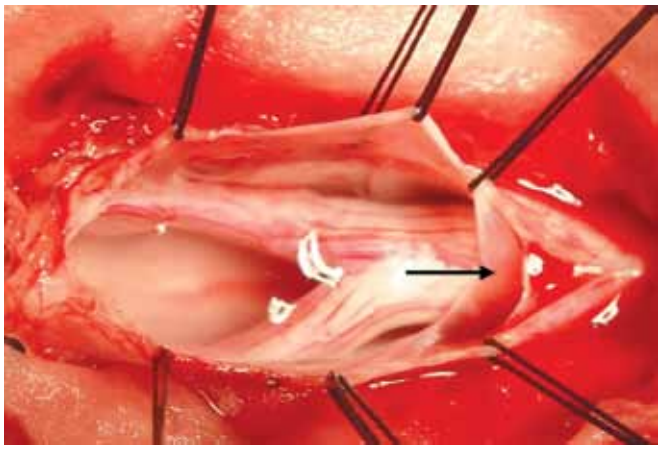

Fig. 2: Intraoperative picture of the split cord with black arrow pointing to thickened arachnoid membrane

cases may raise concern regarding the validity of Pang's unified theory.

Although split cord malformations have been well documented in children, there is no consensus about their surgical indications and clinical course in adults because of their rarity. Adult split cord malformation patients are generally not associated with scoliosis or foot deformities.[4] Most of these patients may be asymptomatic and so are undetected till adulthood. We believe surgical intervention should be offered to all patients with the SCM, as their susceptibility to develop neurological deficits is very high compared normal individuals. Also the risk of having developing a neurological deficit may increase with age.[5]

Tubercular infection of spine occurs in the form of tuberculous spondylitis, intradural tuberculosis or tubercular myelitis. Intradural tuberculosis mostly develops from a downward extension from the intracranial tuberculous meningitis or a secondary spread from adjacent vertebral disease. The origin of primary solitary tuberculosis of spinal meninges without systemic pathology is debatable. In our case, there was extensive lumbar arachnoiditis from L3 to L5 and there was no primary focus either in the brain or in adjacent vertebral bodies.

The clinical signs and symptoms are mostly limited to monoradicular or polyradicular pain syndromes that may be accompanied by motor and sensory deficits, usually evolving over several years.[6] It would be right to attribute the symptoms of our patient to tuberculous arachnoiditis rather than the split cord malformation, as the patient was asymptomatic in her early adulthood. However, the rapid and early development of paraparesis in our patient with lumbar tuberculous arachnoiditis, may be due to presence of underlying split cord malformation aggravating the pathological process in an already tethered cord.

Magnetic resonance imaging is a useful imaging tool for lumbar arachnoiditis.[7] The appearance may be varied from clumping of the *cauda equina* roots in the center to clumping in periphery to give an appearance of soft tissue mass. The most common appearance on axial T2-weighted images show roots are adherent to the margins of the dural sac giving an appearance of empty sac, as was noted in our case (Fig. 1).

A combination of surgery and chemotherapy for all cases of tubercular arachnoiditis with neurological deficits is recommended. Conservative treatment is unwise because it is not always possible to distinguish between tubercular and neoplastic lesions. Also, the presence of split cord malformation demands exploration and detethering of the spinal cord. Surgery aids in histopathological confirmation and relieves the radicular symptoms by arachnoidolysis. Antitubercular therapy remains the mainstay of the treatment and should be started once

the diagnosis is established. High dose corticosteroid is another efficient but unproven adjuvant treatment. The entire course of therapy should continue for at least 9 to 12 months.

The outcome of treatment has been unpredictable, with some reports observing good recovery and some reporting unfavorable outcomes after surgical decompression. Our patient showed significant improvement with a combined surgical, antitubercular and steroid therapy.

CONCLUSION

A possibility of tuberculous arachnoiditis should be suspected despite the absence of primary focus anywhere in the body, particularly in TB endemic regions in patients with chronic back pain. Lack of dividing fibrous septum in SCM of our patient raises concerns over validity of Pang's unified theory. Patients with congenital anomalies of spinal cord may be more susceptible to develop early onset neurological deficits from infective etiologies.

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