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Case Report *Volume 5 Issue 1*

An Idiopathic Case of Dural Ectasia at the Dorsolumbar Junction -A Case Report and Review of Literature

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Abstract

An eleven-year-old girl followed up in our department presented with swelling over the mid and lower back with intermittent back pain. Neurological examination was normal. MRI Dorsolumbar spine showed dural ectasia at the dorsolumbar junction with lateral meningoceles extending from the D10 to L3 level. Genetic studies for Marfan's syndrome (Fibrillin 1) were negative. She had no features of Neurofibromatosis type 1 or any other connective tissue disorders. The child was treated conservatively with analgesics and kept under follow up. Although reports of dural ectasia in patients with connective tissue disorders exist in literature, idiopathic cases have rarely been reported.

Keywords: Dural ectasia; Lateral meningoceles; Marfan's syndrome

Abbreviations

DE: Dural Ectasia; CSF: Cerebrospinal Fluid; CES: Cauda Equina Syndrome; MRI: Magnetic Resonance Imaging.

Introduction

Dural ectasia(DE) is characterized by circumferential expansion or ballooning of the thecal sac and/or nerve root sleeves along the spinal column [1]. They have been found to be associated with connective tissue disorders like Marfan's syndrome, Ehlers-Danlos syndrome or Neurofibromatosis type 1 [2]. However, rarely they are idiopathic. We report an eleven-year-old girl with idiopathic dural ectasia of the dorsolumbar region with lateral meningoceles whom we chose to follow up closely without intervening surgically.

Case Report

An eleven-year girl was brought to our outpatient department with complaints of low and mid back pain with intermittent radiation to bilateral lower limbs. There was no history of trauma or any prior surgical intervention. On examination she had a diffuse swelling in the lower back, more on the left side. Her neurological examination was normal. MRI of the Dorso-lumbar spine showed enlargement of the dural sac at the lower dorsal and upper lumbar levels with posterior vertebral scalloping (Figure 1). There was evidence of dilated nerve roots sheaths/lateral meningoceles extending bilaterally from D10-L3 levels with intramuscular extension paraspinally on the left side (Figure 2). There was no abnormal medullary signal. Her genetic studies were negative for any connective tissue disorders. We treated her

symptomatically and decided to follow up closely without any immediate surgical intervention.

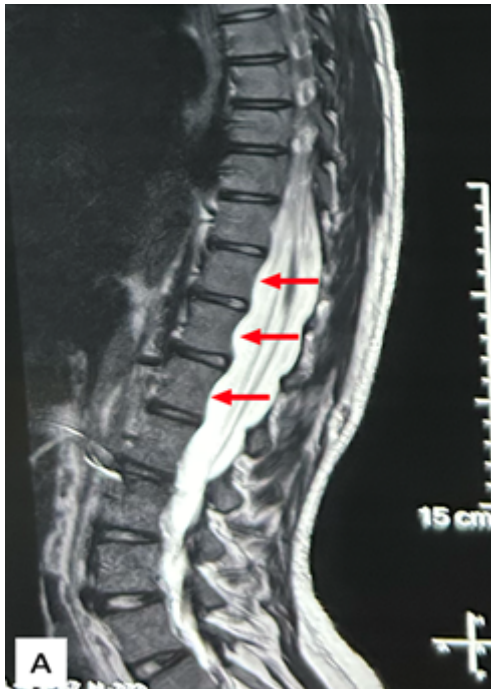

Figure 1: showing expanded thecal sac lower dorsal and upper lumbar region with posterior vertebral scalloping (red arrows).

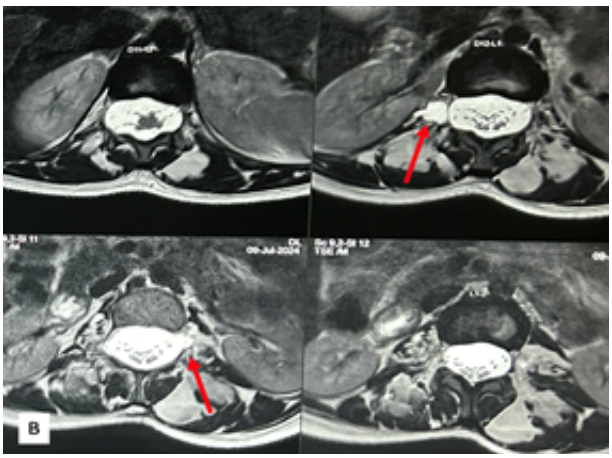

Figure 2: Axial sections at the level of the dural ectasia showing dilated nerve root sleeves (red arrows).

Discussion

Dural ectasia(DE) can be defined as an abnormal enlargement of the thecal sac and/or the nerve root sleeves at any level along the spinal column [1,2]. This may be associated with various connective tissue disorders like Marfan's syndrome, Ehlers Danlos syndrome, Neurofibromatosis type 1, Ankylosing spondylitis or may be secondary to trauma, scoliosis or tumors [3,4]. Although rare, in certain cases, they may have no clear underlying cause [5]. Interestingly, our patient had no history, signs or symptoms to suggest an undiagnosed connective tissue disorder, neither did she have any prior spine trauma or surgery. Literature is relatively sparse when it comes to patients having dural ectasia without any underlying disorder [5].

Although the pathogenesis of dural ectasia is not very clear till date, one of the existing theories is that the same may be a result of cerebrospinal fluid (CSF) pulsations causing distension of an already weakened thecal sac [4]. The most common site for dural ectasia is the lumbosacral region probably because in the upright posture, the CSF pressure is highest in this location [1].

Most patients with dural ectasia are asymptomatic [6]. Common symptoms include back pain, radicular leg pain and/or rectal pain. Certain patients may develop intracranial hypotension secondary to a 'cerebrospinal fluid sink' mechanism leading to postural headaches [7,8]. Rarely, they can cause cauda equina syndrome (CES) when large CSF filled cysts cause significant compression of the cauda eqiuna nerve roots, seen more commonly in patients with Ankylosing spondylitis [4,9,10]. Our patient had back pain with intermittent radiation to bilateral lower limbs which was well controlled with medication. Dural ectasia can also cause vertebral remodelling leading to spinal instability in certain patients [11-13].

Magnetic Resonance Imaging (MRI) of the spine is required for diagnosis. A lateral radiograph of the spine may show posterior vertebral scalloping which provides an indirect hint [14-16]. MRI diagnosis is based on an increase in the anteroposterior diameter of the thecal sac, most commonly seen in the lumbar region [16]. Less commonly, they occur in the cervical or the dorsal spine [17,18]. Other findings include enlargement of the dural sleeves of the nerve roots, formation of meningoceles [5,19]. Bony changes secondary to pressure effects may occur leading to posterior scalloping of the vertebral body, erosion and thinning of the cortex of the pedicles and laminae, expansion of the neural foraminae [1,5]. Bony remodelling may lead to scoliosis, fracture of the posterior elements and therefore spondylolisthesis and spinal instability [5,11,12].

Treatment of dural ectasia is usually based on the symptomatology. In cases associated with spinal instability, surgical stabilization of the spine with instrumented fusion may be required [12,20-22]. In patients with large cysts causing neurological deficits and features of cauda equina syndrome, treatment strategies can be varied like placement

of lumboperitoneal shunts, decompressive laminectomies, marsupialization and/or resection of the cysts, each with variable outcomes [6,9,23]. If associated with postural headaches secondary to intracranial hypotension, conservative measures like proper hydration, bed rest may be adequate with epidural blood patching being required in certain instances [2,19,24]. If the patient does not respond to conservative measures, and the site of CSF leak could be identified on imaging studies, percutaneous injection of fibrin glue and surgical repair have had some success [25]. For patients with only back pain and leg pain, treatment strategies like fibrin glue injection or surgical repair have not shown good results [26]. Our child responded well to symptomatic treatment and hence we decided to follow up closely without any immediate intervention.

Conclusion

Although commonly associated with connective tissue disorders, idiopathic dural ectasia is a rare finding. Most patients are usually asymptomatic. However, pressure symptoms may be noted in certain cases. A number of treatment strategies have been reported depending on the clinical presentation, none proving to be particularly effective. This makes the management of this pathology particularly challenging. We report a young female with idiopathic dural ectasia of the dorsolumbar region whom we treated conservatively and kept under close follow up.

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