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Case Report

Lumbosacral arachnoid cyst with tethered cord: A rare case report

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Abstract

Arachnoid cysts are cerebrospinal fluid collections in the spine that can present with neurological symptoms or be discovered accidentally. Intradural location of such cysts especially in the lumbosacral region is relatively rare. The association of such cysts with other congenital anomalies such as tethered cord lends evidence to the developmental origin of arachnoid cysts. We report a case of lumbosacral arachnoid cyst with tethered cord in a 6-year-old male child and discuss the etiopathogenesis and management options.

Key words: Arachnoid cyst, intradural, lumbosacral, syrinx

INTRODUCTION

Spinal intradural arachnoid cysts occurring in the lumbosacral region are quite rare. Presence of associated anomalies suggests a developmental origin of the arachnoid cysts. We present a case of a 6-year-old boy who presented with paraparesis and was diagnosed as arachnoid cyst of the lumbar and sacral region. A brief review of the etiology and pathogenesis in relevance to clinical decision making is also presented.

CASE REPORT

A 6-year-old boy presented to us with complaints of recent onset difficulty in walking and dribbling of urine. On examination he had grade 3/5 power in both lower limbs and absent reflexes. An MRI spine was performed which revealed a clearly defined intradural cystic lesion extending from L2 to S2 which was hypointense on T1-weighted images [Figure 1] and hyperintense on T2-weighted images [Figure 2] with tethering of cord. A syrinx was also noted in the cervicodorsal spine [Figure 3]. A tentative diagnosis of lumbosacral arachnoid cyst was made and the patient was taken up for surgery. Multiple level laminectomies were performed and on opening the dura a tense cyst was found [Figure 4] along with short and thickened filum terminale. There were no other associated lesions. No communication was found between the cyst contents and the subarachnoid space. The lesion was incised and clear fluid contents were let out. The walls were marsupialized and the dura was closed primarily. Detethering of cord was done by transection of thickened filum terminale. Postoperatively the child's motor

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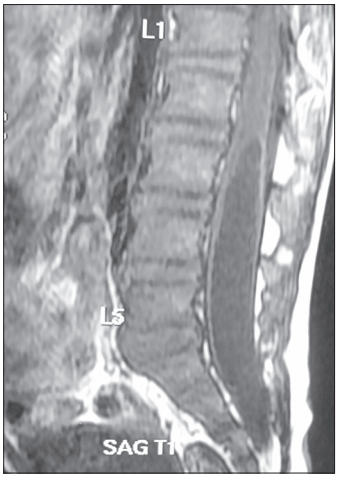

Figure 1: T1-weighted MRI showing hypodense intradural cystic lesion extending from L2 to S2

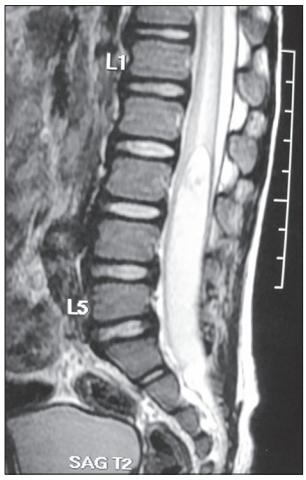

Figure 2: T2-weighted image showing hyperintense lesion

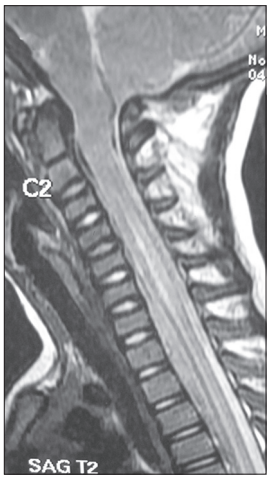

Figure 3: Syrinx in the cervicodorsal spine

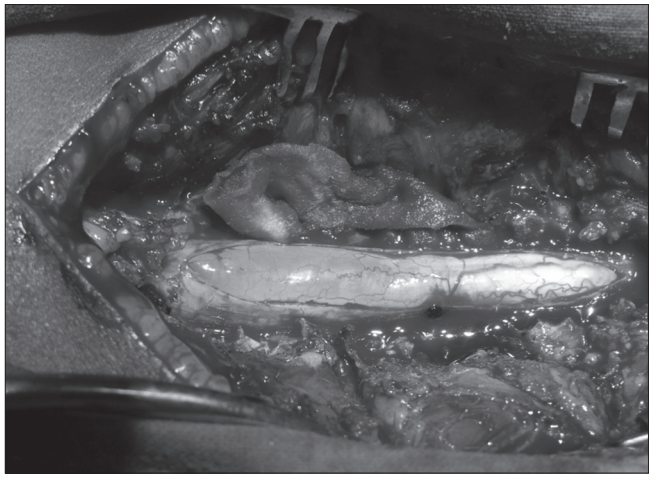

Figure 4: Intra operative photograph showing tense cyst on opening the dura

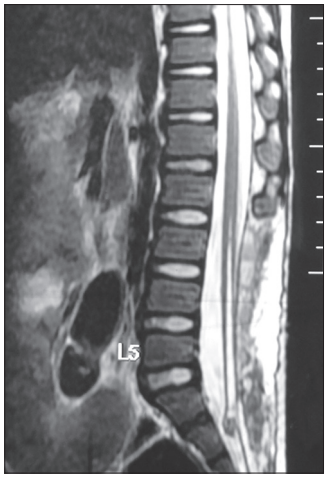

Figure 5: Postoperative MRI showing complete decompression of cyst

power began to improve though his bladder was kept catheterized. Postoperative MRI was done which showed complete resolution of cyst [Figure 5]. At follow-up of 2 months patient could walk independently and had regained bladder control.

DISCUSSION

Arachnoid cysts of the spine are collections of cerebrospinal fluid (CSF) within protrusions of arachnoid that can occur in a perineural, extradural, intradural, or intra-extradural site.[1] These lesions are most often located posterior to the spinal cord but have also been identified anteriorly. The thoracic spine is the most common location followed by lumbosacral and cervical spine. Spinal arachnoid cysts are equally prevalent in men and women usually presenting in the fourth and fifth decades of life. Rarely they may present in children with other anomalies thus lending support to congenital etiology of spinal arachnoid cysts.[2] Idiopathic arachnoid cysts in children are associated with neural tube defects and in adults with spinal deformities.[3]

Sacral cystic lesions were classified into three types by Nabors *et al*. Type 1 included extradural cysts, type 2 included cysts involving nerve root also known as Tarlov perineural cyst and type 3 consisted of intradural cysts. Intradural cysts are outpouchings of arachnoid that, regardless of size, lie entirely within the dural space. Intradural spinal arachnoid cysts usually consist of multiple lobules. Lumbosacral arachnoid cysts are believed to occur as a disorder of secondary neurulation. While patients with arachnoid cysts/meningoceles are usually neurologically normal, 90% of patients with meningocele have associated occult spinal lesions such as tight filum terminale, split cord malformation, and epidermoids. Subsequent tethering of the spinal cord may lead to progressive neurological deficit.[4]

The etiology of arachnoid cyst formation is still not well established. Alterations in the arachnoid trabeculae that overlie the spinal cord are probably the root cause. These alterations may be a congenital or may be caused by inflammation from previous trauma, surgery, or subarachnoid hemorrhage.[5] In some cases no cause can be established,[6] It has been hypothesized that these lesions develop from the septum posticum of Schwalbe which is an arachnoid membrane dividing the midline posterior cervical and thoracic subarachnoid space. But such a hypothesis does not explain cysts occurring anterior to the cord.[7] According to

the hydrodynamic theory normal pulsations of the CSF dilate weak areas of arachnoid which then progressively enlarge and form a cyst. A ball valve effect at the neck of the diverticulum is probably responsible for the progressive enlargement of the cyst.[5,8] This also helps explain syrinx formation as in our case.[8]

Another theory suggests incarceration of arachnoid granulations producing entrapment of CSF in arachnoid diverticula. These pockets of fluid lead to further disruption of normal pulsatile CSF flow and are capable of expanding and developing into cysts.

Most spinal arachnoid cysts are asymptomatic and are discovered incidentally on magnetic resonance (MRI). MRI is the most sensitive and specific study for detecting a spinal arachnoid cyst and for assessing the extent of the cyst wall. Contrast-enhanced MRI can help distinguish arachnoid cysts from other cystic tumors. The signal intensity of arachnoid cyst fluid is the same as CSF on T1 and T2 images. Sometimes higher signal intensity is seen on T2 which maybe related to increased protein content of sequestered fluid or absence of motion effects. Diffusion-weighted MRI not only helps differentiate the arachnoid cyst from epidermoid cyst, abscess, or tumors but also evaluates spinal cord atrophy and inflammatory changes. Although CT myelography has been used to establish or refute communication between the intraspinal subarachnoid space and the arachnoid cyst, it is more sensitive in determining whether a communication exists rather than locating the communication. 12 Kinematic MRI (cine-MRI) helps locate the communication which appear as pulsating turbulent flow voids. This helps limit the surgical approach.[9,10]

The presentation of sacral arachnoid cysts varies from long standing low back pain to neurological deficits such absent deep tendon reflexes or bowel bladder abnormalities. Lower extremity sensation is usually spared. Perineal pain may be present in approximately 50% of patients. Radicular pain if present is relieved by lying down which pushes fluid out of the cyst.

The treatment decision needs consideration of the extent of the cyst, the point at which the spinal cord is maximally compressed and presence or absence of communication between cyst and subarachnoid space. Aspiration under MRI guidance is advised for small cysts which have no communication with subarachnoid space.[9] For moderate-sized cysts complete excision is advised and multiseptated cysts extending over several segments, cyst fenestration can be done to avoid extended laminectomy. All authors report a good-to-excellent outcome with almost no recurrence. Intradural cysts should be excised by marsupialization, opening to the surrounding intradural fluid. Percutaneous cyst drainage can be tried as a temporary measure to decrease symptoms or as a test of success of operative management. Endoscopic approaches have also been used to treat sacral extradural arachnoid cysts. The success of the treatment modality employed ultimately depends on the degree of correlation of the MRI findings with the patient's symptoms. Associated syrinx have been described to regress after treatment of the arachnoid cyst.[11]

On histopathology, the walls of arachnoid cysts are usually seen as fibrous and lined by meningothelial cells. These cells are usually negative for GFAP, S-100, transthyretin (prealbumin), and CEA staining thus differentiating them from epithelial cysts.[12]

Surgery typically results in excellent outcomes in terms of resolution of symptoms, and is effective across a large range of cyst sizes. Some cysts cannot be fully resected either due to their location or adhesions. These cases may benefit from fenestration, percutaneous drainage, or cystoperitoneal shunt.[8]

CONCLUSIONS

Arachnoid cysts of the spine may occasionally be encountered in the work up of neurological symptoms or incidentally. It is important to be aware of the possibilities of other congenital anomalies of the spine and investigate for the same. For good surgical outcome it is essential to have a correlation of MRI and clinical symptoms. Cine-MRI is useful to guide surgical decision making.

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