Thoracolumbar spinal neurenteric cyst with tethered cord syndrome and extreme cervical lordosis in a child

A case report and literature review

Zhi Gang Lan, MD, Seidu A. Richard, MD, Chuanfen Lei, MD, Siqing Huang, MD, PhD

Abstract

Rationale: Neurenteric cysts, are rare benign tumors of the central nervous system that are mostly located in the spinal cord and predominantly seen in male children although adult form of the disorder also occurs. The etiology and treatment of this disorder is still a matter of debate. Our case further throws more light on the pathogenesis and treatment of this disorder.

Patient concerns: A 4-year-old boy presented with 5-month history of cervical lordosis and bilateral lower extremity pain that progressed to his abdomen and upper body. The pain was general, recurrent, non-persistent and progressive in nature with no paralysis. The pain was aggravated by trunk stretching and relieved when he assumed opisthotonos position so he preferred sleeping in this position at night.

Diagnoses: Magnetic resonance imaging (MRI) revealed a cystic lesion at the thoracolumbar spine with tethering of spinal cord and cervical lordosis.

Interventions: He was operated on successfully and the cervical lordosis and pain resolved.

Outcomes: The child recovered well with no tumor recurrence and massive improvement of his life.

Lessons: The gold standard treatment for this disorder is surgery although the precise surgical approach is still a matter of debate. We are of the view that surgical approach should be individualized and aim at total excision of the cyst.

Abbreviations: CNS = central nervous system, CT-scan = Computerized tomography scan, IONM = intraoperative neuromonitoring, MEPs = motor evoked potentials, MRI = magnetic resonance imaging, SSEPs = somatosensory evoked potentials.

Keywords: lordosis, neureteric cyst, opisthotonos, spine, thoracolumbar

1. Introduction

Neurenteric cysts are rare benign tumors of the central nervous system which constitutes about 0.3% to 1.3% of all spinal cord tumors. This cystic lesion is referred to as enterogenous cysts or endodermal cysts. They are mostly seen in children between the ages of 1 and 10 years with a mean age of 6.4 years. These tumors may sporadically occur in adults with male preference rate of about 60.4%.[1-3] Characteristically, the lesions may be located intradural or extramedullary in about 78.6% of cases. Furthermore, about 73.6% of this cyst originate from the cervical, cervicothoracic, and thoracic spine.[1-5] Clinically, most patients present with progressive focal pain at the level of spinal axis of the lesion with myelopathic signs, and radicular symptoms being most common and often depends on the location of the pathology.[3,4,6,10-13] The simultaneous occurrence of neurenteric cysts and tethered cord syndrome has been seen in clinical practice.[1,2,14-18] Magnetic resonance imaging (MRI) now offers clinician the golden opportunity of diagnosing these disorders preoperatively. On T2-weighted imaging, the cysts are hyperintense and exhibit negligible or no enhancement on T1 postcontrast imaging.[1,4,7] Left untreated, intraspinal neureteric cysts can result in very devastating consequence although they are benign lesions.[7,15,16] The gold standard treatment option for this kind of lesions is total surgical resection which ameliorates the presenting neurological symptoms in about 71% of patients.[2,15,20-22] In cases with simultaneous occurrence of the cyst and spinal cord tethering, detethering is the treatment option since detethering prevents progressive neurological deficits and also ameliorates pain as well as enhance neurological function in a sizable number of cases.[1,12-24] We present a case of thoracolumbar spinal neurenteric cyst with extreme cervical lordosis in a child.
lordosis in a child which we operated on and efficiently relieved the cervical lordosis.

2. Case report
A 4-year-old boy presented with 5-month history of cervical lordosis and bilateral lower extremity pain that progressed to his abdomen and upper body. The pain was generalized at the upper trunk, recurrent, nonpersistent, and progressive in nature with no headache, dizziness, vomiting, or paralysis. The pain was aggravated by trunk stretching and relieved when he assumed opisthotonos position so he preferred sleeping in this position at night. His bowel and bladder habit did not change much. His immunization status was completed according to age. His past medical history was unremarkable. He had no history of spinal trauma or association with anybody with chronic cough. General physical examination did not yield much. All systems were grossly normal. Neurological examination revealed normal cranial nerves without obvious skull abnormalities except extreme opisthotonos with cervical lordosis and bilateral low limb hyperreflexia (Fig. 1A). Abdominal reflexes were brisked with abnormal cremasteric reflexes. The patient could not lie in supine position because of the excruciating pain. Digital rectal examination revealed a normal spinster tone. Laboratory and as well as other ancillary investigations (ECG, CXR, etc.) was normal.

MRI showed a cystic lesion at T12-L1 with tethering of spinal cord and cervical lordosis (Fig. 1B, C). The lesion was intradural. The cyst is hyperintense on T2-weighted imaging and on T2 it shows negligible or no enhancement on postcontrast imaging. There is swelling of the spinal canal from T12-S1 with compression on the nerve roots of T12 and L1 vertebra. A working diagnosis of neurenteric cyst was made to rule spinal lipoma. Our evaluation put the child into McCormick grade III. After education and counseling the parent’s surgery was scheduled. After general anesthesia, our neurophysiologies applied the intraoperative neuromonitoring which comprise of somatosensory evoked potentials and motor evoked potentials followed by a T12-L2 laminectomy in a prone position. The lesion was intradural. The dura was opened, and the lesion seen at posterior portion of T12-L1 thoracolumbar spinal level. The tumor was cystic, oval-like, with a complete capsule. The capsule was very smooth, and inside this cyst was colorless transparent fluid. The cyst had direct connection with the nerve roots of T12 and L1 as well as tight compressive effect on these nerve roots. The adjacent spinal cord and nerve roots below the tumor appeared swollen, with increased vascularity. The cyst and capsule were totally removed with the aid of the microscope (Fig. 2A). Intraoperative monitoring above was stable throughout the operation.

Pathological examination revealed a benign cystic space occupying lesion with mucinous columnar epithelium lining (Fig. 2C, D) which confirms the diagnosis of neurenteric cyst. Immediate postoperative assessment revealed resolution of the cervical lordosis and pain. The patient recovered very quickly and
was discharged home 10 days after operation. Follow-up outpatient visits were scheduled every 3 months initially and changed to every 6 months after the first 2 visits. Two years’ follow-up revealed no tumor recurrence and massive improvement of the child’s life.

3. Discussion

Neurenteric cysts are benign tumors of the central nervous system, which constitutes about 0.3% to 1.3% of all spinal cord tumors. This cystic lesion is referred to as enterogenous cysts or endodermal cysts.[1–4] They are mostly seen in children between the ages of 1 and 10 years with a mean age of 6.4 years. This tumors may sporadically occur in adults with male preference rate of about 60.4%.[1,3] Majority of the lesions, about 73.6% originate from the cervical, cervicothoracic, and thoracic spine and about 78.6% may be located intradural or extramedullary.[1,5–9] The embryonic origin of this cyst is still a matter of debate but many authors believe that the cyst usually arises from an anomalous gap between the primitive neurenteric canal, notochord, and neural tube to the adjacent endoderm and mesenchyme during the 3rd week of embryogenesis.[1–4]

The clinical presentation of this disorder is often either acute, insidious, prolonged, or fluctuating and associate with persistent local pain, radiculopathy, and myelopathy thus mimicking symptoms of multiple sclerosis.[13,6,10–13] The initial and presenting symptom is mostly the pain as described by most authors. Our case is not exceptional because he presented with sudden back pain that continued for several days and progressed into torticolls. The explanations for this acute onset is the association of acute upsurge in the size of the cyst content.[10] Upper respiratory tract infection may also cause an acute upsurge in mucin secretion of the lesion resulting in acute spinal cord compression.[10] Malcolm et al obverse in their intracranial case that hemorrhage into the cyst can also occur and this may lead to extraordinary acute presentation of this disorder.[10,26] Sporadic fluctuations of the mucinous cyst content, either through osmotic means with intraluminal absorption of the intracystic fluid, or periodic rupture of the cyst wall usually leads to fluctuating symptoms associated with this disorder.[10,11,20]

Initial classification of this disorder was done by McCormick and modified by Aghakhani et al into the current modified McCormick classification which comprises of grade I to IV as follows[4]: grade Ia in which the patient has neurologically function, normal gait, and goes about his or her daily activities. In grade Ib however, the patient is usually tired after walking numerous kilometers, the patient cannot tolerate running and has moderate sensorimotor deficit which does not significantly affect the involved limb, as well as moderate discomfort in his or her normal daily activities. In grade II, patient presents with sensorimotor deficit which affects function of the involved limb, mild-to-moderate gait abnormality, severe pain, or dysesthetic syndrome that limit his or her quality of life but he or she can perform simple daily activities as well as move about. In grade III,
MRI is the gold standard neuroimaging modality use in the diagnosis of this disorder.\textsuperscript{[1,3,4,6,10,13,26,29]} Research has proven that, on MRI, the deficiency of contrast enhancement of the cyst wall and the nonexistence of a mural nodule aid in distinguishing this disorder from other cancerous lesions.\textsuperscript{[10,21]} Furthermore, MRI offers comprehensive evidence concerning the site of the cyst within the neuraxis, the degree of spinal cord compression as well as the co-existence of a posterior mediastinal or abdominal cyst. Studies have further shown that MRI sequences, the cyst content, and CSF may be alike, or maybe proteinaceous with shortening of T1 relaxation time as well as appearing very intensity on T1-weighted images.\textsuperscript{[10,11,20]} Computerized tomography scan however is valuable in ruling out any related bony vertebral malformations like vertebral bony clefts, butterfly vertebrae, and anomalies of segmentation, which may coexist in about 50\% of cases.\textsuperscript{[9,10,21]}

The surgical approaches as well as the extent of resection of this spinal disorder are still a matter of debate since the lesion may arise anywhere in the spinal cord. Therefore, we are of the view that surgical approach should be individualized and aim at total excision of the cyst. Although many surgeons recommend the classical posterior approach via laminectomy,\textsuperscript{[2,4–7,10,11,13,30]} Devkota et al recommended an anterior central corpectomy approach for lesion that arises from the anterior portion of the cervical spine because of the adhesion of the cyst wall to the spinal cord. They argued that the total excision of the cyst via the posterior laminectomy was challenging technically without extra maneuvering of the spinal cord.\textsuperscript{[15,31]} Other surgical approaches that have been used by various surgeons to tackle this lesion include simple aspiration of the cyst content, marsupialization of the cyst wall, partial or total excision of the cyst wall either through an anterior or a posterior approach, and cystosubarachnoid shunt technique.\textsuperscript{[2,4–7,10,13,20,30,32–34]} Total resection should be the target during surgery since incomplete resection has the greater chances of recurrence and may necessitate a 2nd surgery.\textsuperscript{[10]}

Microscopic examination of the excised lesion often reveals cells lined with mucin-producing nonciliated, simple or pseudocuboidal, or columnar epithelium, resembling gastrointestinal tract mucosa.\textsuperscript{[3,10,21]} Based on these histopathological appearances, Wilkins and Odom coined a classification system for disorder.\textsuperscript{[3]} In this classification, type A cysts consist of cases with either columnar or cuboidal cells, with ciliated and nonciliated constituents over a basal membrane made up of type IV collagen. Type B cysts consist of cases with all of the qualities of type A with extra feature like bone, cartilage, lymphatic tissue, fat, or glandular constituents. Type C cysts consist of case with all the qualities of type A in addition to ependymal or glial tissue. Studies have shown that there is no link between this classification and the site, extent, or outcome after resection of the tumor.\textsuperscript{[3]}

4. Conclusion

The embryonic origin of this cyst is still a matter of debate so we propose further studies to understand the precise pathogenesis of this cystic occurrence. The clinical presentation of this disorder can mimic symptoms of multiple sclerosis. MRI is the gold standard neuroimaging modality use in the diagnosis of this disorder although computerized tomography-scan is also beneficial in ruling out bony abnormalities. The gold standard treatment for this disorder is surgery although the precise surgical approach is still a matter of debate. We are of the view that surgical approach should be individualized and aim at total excision of the cyst.
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