

Paraplegia with lymphoedema - a rare case report

R Patel,¹ VK Mehta,¹ DP Singh,¹ A Jain,² M Bhatnagar²

¹Department of Medicine, RNT medical College, Udaipur, ²Department of Zoology, University College of Science, MLS University, Udaipur

Corresponding Author:

Ramesh Patel, MD
Department of Medicine, RNT medical
College, Udaipur- 313001, INDIA
Tel.: +91-9530079043
E-mail: rameshpatel61@rediffmail.com

ABSTRACT

Arachnoid cysts are a congenital disorder, and most cases begin during infancy; however, onset may be delayed until adolescence. Here we present an unusual case of atypical Paraplegia with Lymphoedema, with onset during adolescence and rapid progression in a young female patient who showed the characteristic appearance of cyst on magnetic resonance imaging (MRI) of spinal cord. This report intends to highlight paraplegia with lymphoedema, as a rare cause of spinal cord compression in pediatric population along with congenital defect manifestations and an interesting radiology finding of the disease.

KEY WORDS: Arachnoid cysts, Paraplegia, Congenital defect, Spinal cord compression

doi : 10.5214/ans.0972.7531.180412

Introduction

Extradural arachnoid cysts are causative agents of spinal cord compression in the pediatric population that arise from congenital defects in the dura mater.¹⁻³ These cysts are reported to be in communication with the intrathecal subarachnoid space through a small defect in the dura.

Arachnoid cysts, most of which are developmental anomalies, are benign cysts occurring in the cerebrospinal axis in relation to arachnoid membrane and it do not communicate with ventricular system as well. They usually contain clear, fluid which is colourless similar as cerebrospinal fluid; rarely, they contain xanthochromic fluid. In several studies, extradural arachnoid cysts are reported in solitary form as compared to multiple which are very rare in children.⁴

Case report

A young female of 17 years (Fig 1) was admitted in our unit with chief complaints of difficulty in walking for last 5 years which was gradual onset and progressive, it was associated

with frequent falling on ground because she complained of stiff legs and was unable to lift them off the ground. For last one year there was development of swelling over lower limb which initially developed over ankle and gradually progressed up to knee of both lower limbs. There was no history of pain, trauma, fever, backache, bowel or bladder involvement. There was no family history or any significant birth history.

General physical examination showed non-pitting edema of both limbs up to knee. No trophic ulcers were seen. Patient was able to comfortably lie in bed and was able to sit by herself. In order to stand, she required support as was also true for walking. On neurological examination, mental function, cranial nerves, speech and upper limbs were found to be absolutely normal. In lower limbs, tone was found to be increased bilaterally. The power was 3/5 in bilateral toes, 4/5 in bilateral ankles, 4/5 in bilateral knees. Plantar reflex was found to be bilateral extensor. There were brisk reflexes in lower limbs but without clonus. Sensory examination showed abnormal decrease in touch, pain, temperature in both lower limbs up to knee. Severely impaired proprioception in both lower limbs, coupled with abdominal



Fig. (1a): Lymphoedema of lower limbs before surgery.



Fig. (1b): Improvement in Lymphoedema of lower limbs after surgery.

reflex was absent in both lower quadrants while gait was spastic. Spine revealed no abnormality on inspection and palpation. The remaining systems were normal.

Considering history and findings on examination the patient was diagnosed as spastic paraplegia with probable involvement of D10 with lymphoedema. She was advised MRI of dorsal spine (Fig 2) which showed a fairly large well – defined lobulated cystic lesion involving the extradural compartment of the dorsolumbar cord extending from D9 to L2 level compressing the cord which is seen flattened and displaced anteriorly within the bony spinal canal suggestive of an arachnoid cyst. The patient underwent laminotomy and excision of cyst in our neurosurgery OT. Biopsy report of excised material was suggestive of arachnoid cyst. Following surgery there was remarkable improvement in swelling of the legs (Fig 1b) and some neurological improvement as well. She was advised continuous follow up. Informed Consent was obtained from the patient.

Discussion

Congenital extradural spinal cysts are also known as developmental extradural arachnoid cysts become visible during second decade of lifespan. Generally cysts are located in thoracic area of the body. The male to female ratio of occurrence is 4:3.⁵ Congenital extradural spinal cyst is attached to dura mater by narrow pedicle posterolaterally at point of attachment of dural sleeve which covers the dorsal nerve root. The communication between cyst and subarachnoid space may or may not take place. Such cysts may be present in multiple numbers in same patient. Under the microscope cyst may appear as a flattened cell layer resembling arachnoidal, avascular collagenous connective tissue forms the rest of wall. Most cases have reported these cysts as sporadic but not familial. Numerous hypotheses imply a congenital defect of some sort. Regardless of their origin, hydrostatic pressure and cerebro-spinal fluid (CSF) pulsations (perhaps with a ball-valve mechanism) appear to be

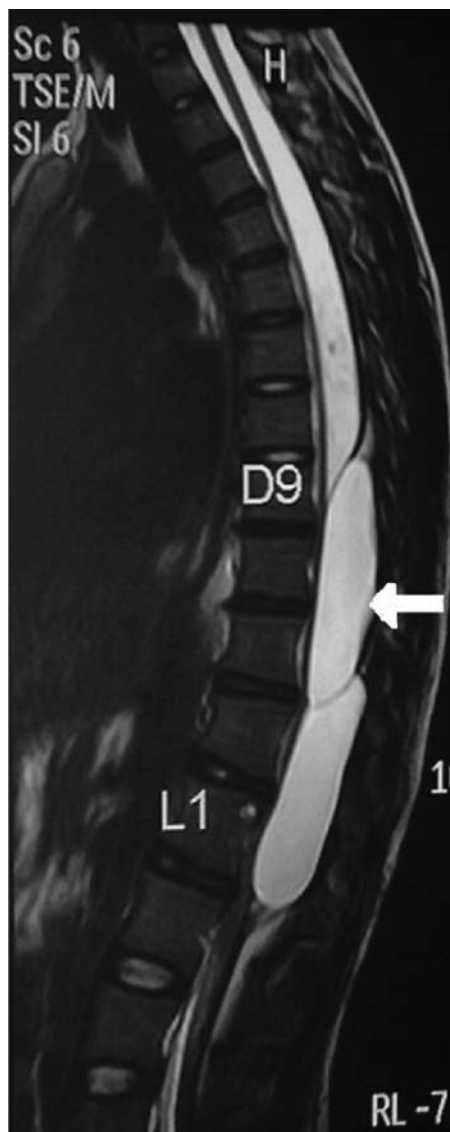


Fig. (2a): MRI image of dorsolumbar spine T1W showing lobulated arachnoid cyst. (Arrow shows the compression in the spinal cord.)

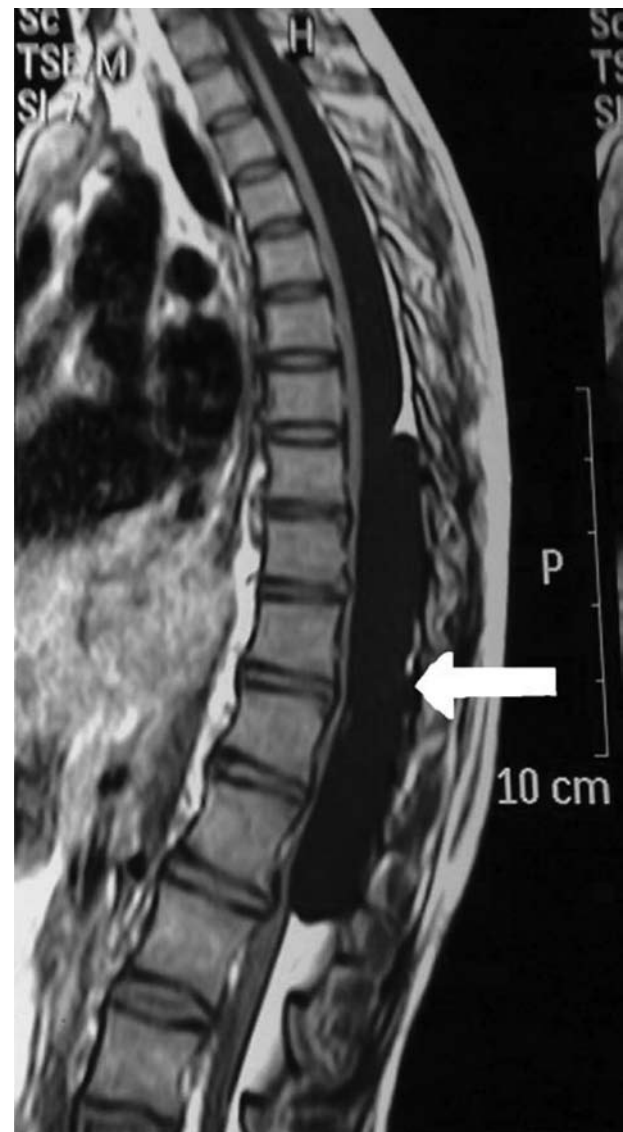


Fig. (2b): MRI image of dorsolumbar spine T2W showing lobulated arachnoid cyst. (Arrow shows the compression in the spinal cord.)

the explanation for cyst enlargement as for some unexplained reason the communication between cyst and spinal subarachnoid space becomes obliterated. Patient with a congenital extradural cyst in the thoracic or cervical region undergoing clinical course is one of spastic paraparesis or quadriparesis of fluctuating and progressive severity. The involvement of motor functions is of higher degree than compared to sensory and autonomic functions. In lumbar area a congenital extradural cyst may present with pain of radicular nature. Cyst enlargement results in enlargement of spinal column, which can be identified with the help of radiography. Myelography usually reveals an extradural defect and occasionally the contrast medium enters the cyst through its pedicle. On other hand MRI of spinal arachnoid cysts reveals an oval, sharply demarcated extramedullary mass which may cause spinal cord compression or local displacement. The cyst is usually hyperintense to CSF on T2-weighted sequences because of the relative lack of CSF pulsation artifacts. The overall prognosis is good in surgically

treated cases. The cyst may be excised or resected and dural defect can be closed. Patient's recovery depends on the severity of the cord compression, but the key is early diagnosis followed by treatment.

References

1. Liu JK, Cole CD, Sherr GT, et al. Noncommunicating spinal extradural arachnoid cyst causing cord compression in a child. *J Neurosurg* 2005; 103(3 Suppl): 266–269.
2. De-Amato MC, Santos MV, Simao GN, et al. Extradural arachnoid cysts in children. *Childs Nerv Syst* 2007; 23(11): 1233–1238.
3. Liu JK, Chad DC, Kan P, et al. Spinal extradural arachnoid Cysts: Clinical, radiological, and surgical feature. *Neurosurg Focus* 2007; 22(2): E6.
4. www.neurosurgery.tv/intraspinalcysts.html, Neurosurgical Videogallery, CNS Clinic, 2005.
5. Bitaraf MA, Zeinalizadeh M, Meybodi AT, et al. Multiple extradural spinal arachnoid cysts: a case report and review of the literature. *Cases J* 2009; 2: 7531.