# An Unusual Case of Chronic Lower Limb Pain in an 11-Year-Old Boy

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Spinal neurenteric cysts are rare congenital abnormalities composed of heterotopic endodermal tissue. These cysts are considered to be a form of occult spinal dysraphism that result from inappropriate partitioning of the embryonic notochordal plate and presumptive endoderm during the third week of embryogenesis. These heterotopic epithelial remnants of gastro-intestinal and respiratory tissue lead to eventual formation of compressive cystic lesion of the spine[1]. The terminology of these cysts are synonymous with enterogenous cyst, split notochord syndrome, endodermal cyst, gastro-enterogenous cyst and teratoid cyst. They account for 0.7-1.3% of all spinal cord tumours[2]. Intradural/extramedullary compartments are the commonest location for the neurenteric cysts, approximately 90%, whereas the remaining 10% are found at an intradural/intramedullary or extradural location[3] (abdomen, mediastinum, pelvis, brain and rarely subcutaneously). Individuals with neurenteric cysts frequently present in the second or third decade of life with an approximate 2:1 male-to-female ratio[4,5]. In the paediatric population, 61.2% patients with these cysts are males with a mean age of 6.4 years at presentation[6]. In this report we are presenting an unusual case of chronic pain and lower limb posturing in a child diagnosed to have a spinal neurenteric cyst.

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## Case Report:

11-year-old boy, a sixth standard student, was referred to the PMR outpatient's department for management of pain and abnormal posturing of the left lower limb. He was apparently well till eight months prior to presentation, when there was a history of trauma to the left foot while playing football in school. Other than mild pain in the foot, child was alright and was able to complete the game and walk back home. As the foot pain did not resolve in the following 3 weeks, he was taken to a local orthopaedic surgeon who diagnosed a metatarsal hairline fracture and conservatively managed this with a below knee plaster cast. This cast was removed after three weeks. However, the pain in the left foot worsened and gradually progressed to involve the entire left lower limb.

He complained of pain on touch but was able to walk with a mild limp for another 2 months from onset, when the severity of pain increased and ambulation became progressively difficult. By the fourth month from the onset of injury, the child stopped walking and found relief in the pain by keeping the left hip and knee in a flexed posture. There is no history of any cognitive decline, behavioural change, bowel-bladder symptoms, sensory or motor impairment elsewhere in the body, at the time. The hip-knee flexed posture was maintained at all times, even in sleep. Any attempt to correct the hip/knee resulted in severe pain. Child found it increasingly difficult to wear trousers as the touch of the cloth aggravated the pain.

He was taken to several hospitals and was evaluated for the painful condition. At most places, he was treated with analgesics. He was diagnosed as chronic regional pain syndrome type 1 at one hospital and was given an epidural block following which manual stretching of the left lower limb was done. The pain was better; however, it recurred a few days later, with the same intensity, as the effect of the block wore off. He was then brought to CMC, for further management.

On examination, the child was of a cheerful disposition, sitting comfortably with the hip at 90 degrees flexion, knee completely flexed and ankle in plantar flexion. Vital signs were within normal limits and other general and systemic examination were unremarkable. Power in the upper limbs and the right lower limb was grade 5 (MRC grading), and sensations were normal except on the left, below L1, he had allodynia and would not permit examination. On standing on the right lower limb, he was able to extend the hip from 90 degrees by 30 degrees and the knee from complete flexion, by 20 degrees. He was also able to minimally move the toes. Muscle wasting could not be commented upon. The tone was mildly increased in the right lower limb and deep tendon reflexes were within normal limits in the upper limbs but mildly exaggerated in the right lower limb.

Historically the child had good relationships with his parents, siblings, other family members and friends/ classmates. He is an above average student and there was no history suggestive of any difficulties in school. There was no suggestion of secondary gains from the situation. Other than the allodynia and left lower limb posturing, there were no other features suggestive of an ongoing illness.

The investigations done elsewhere consisted of magnetic resonance imaging of bilateral hip joints and x-ray of the left ankle and foot, which were all within normal limits. A bone scan showed a mild tracer uptake at the level of T6 but no significant disease process was reported.

As history was unremarkable for the cause of pain and the clinical signs were limited, the question of diagnosis and appropriate treatment were discussed with the parents. After having been to several hospitals and not having any relief in the child's symptoms, the parents were on the verge of giving up hope and returning home due to financial and time constraints. A probable need for intervention of child psychiatry unit was also discussed. It was decided to evaluate the condition with

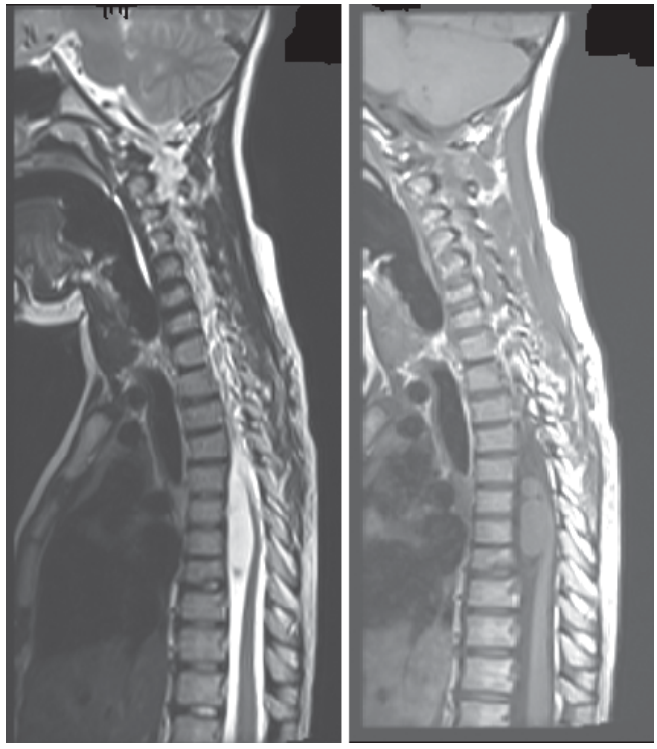

**Fig 1-** *MRI showing sagittal section of the cervico-dorsal spine with the Neurenteric cysts at T5-8 level, T1 (cysts are isointense) and T2 (cysts are hyperintense) weighted images*

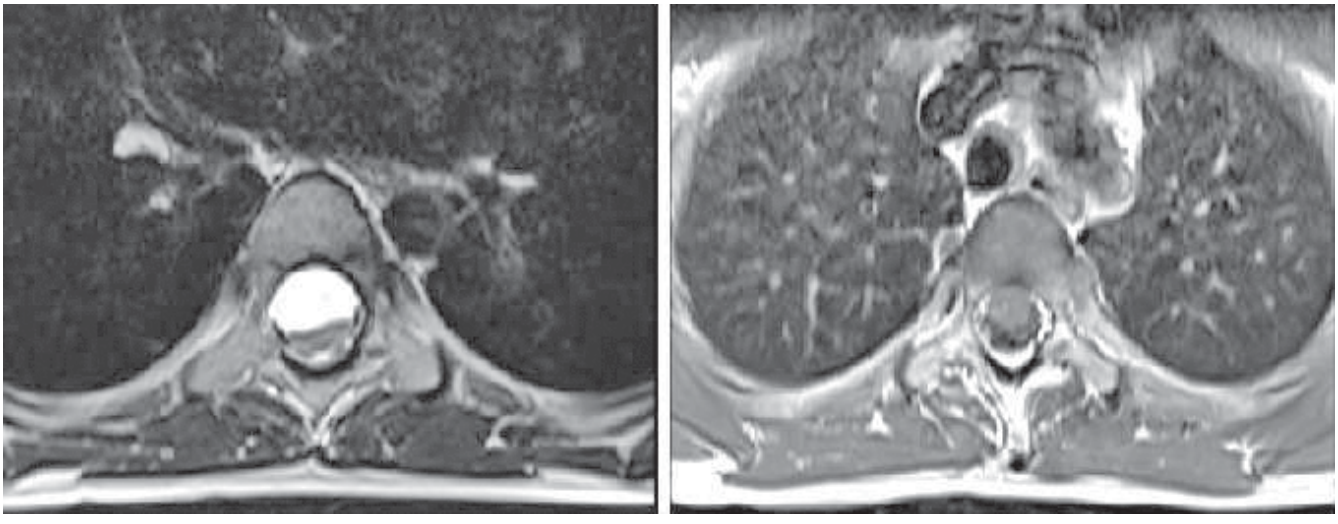

**Fig 2-** *MRI showing transverse section of the cervico-dorsal spine with the Neurenteric cysts at T5-8 level, T1 (cysts are isointense) and T2 (cysts are hyperintense) weighted images.*

an MRI of the spine and electrodiagnostic tests for the lower limbs to look for any spinal pathology causing these symptoms prior to referral to psychiatry.

Nerve conduction studies could not be done as the child did not permit the test to be done on the left lower limb. However, MRI of the spine revealed two well-defined intradural, extramedullary oval, cystic lesions with the anterior thecal sac, extending from lower T5 up to mid T8 vertebral body (Figs 1&2). The larger cyst measured 3×1.4cm and the smaller lesion measured 9×11mm superior to the lesion mentioned above. Both were isointense on T1 and hyperintense on T2 sequences with no enhancement or diffusion restriction. The lesions displaced the cord posteriorly causing severe thinning and straightening of the mid dorsal column. A thin septum of the cord was seen from T4 extending up to T8 level. The findings were suggestive of probable neurenteric cysts.

The child was referred to neurosurgery for surgical management of the cysts. A T5-8 laminoplasty and total excision of the cystic mass was done. Intra-operatively, the dura was found to be tense but the arachnoid was normal. The cord was displaced dorsally by the two ventrally located cysts which were intradural and extramedullary. The cysts were not found to be communicating with each other. They had thick white walls and contained white, mucoid fluid. While under anaesthesia, the left lower limb was manually stretched and a plaster cast (above knee) was applied. Post-operatively, allodynia in the left lower limb completely resolved, the mild tone and exaggerated reflexes also improved. The mild urinary hesitancy the child had developed prior to surgery also resolved postoperatively. Biopsy of the cystic mass was reported as type A neurenetric cyst.

## Discussion:

In human embryogenesis, during the third week, the neurenteric canal unites the yolk sac and the amniotic cavity as it traverses the primitive notochordal plate. Persistence of the normally transient neurenteric canal prevents appropriate separation of endoderm and notochord. Manifestation of this anomalous union is seen as the congenital abnormalities of the spine defined by the presence of mucus-secreting epithelium reminiscent of the gastro-intestinal and respiratory tract.[1] The diagnosis is by classical histopathological description of the cyst on haematoxylin and eosin staining of the tissue which shows a collection of mucin producing simple columnar or cuboidal ciliated and non-ciliated goblet cells surrounding a central cystic cavity. Wilkins and Odom in 1976, classified these cysts into three types based on the histological features of cyst wall and its contents.[1] The walls of type A cysts mimic gastro-intestinal or respiratory epithelium with a basement membrane supporting single or pseudostratified cuboidal or columnar cells, which may be ciliated or non-ciliated. type B cysts include all the features of type A as well as additional tissue containing glandular organisation, usually producing mucin or serous fluid. Type C cysts are most complex, containing ependymal or glial tissue within the cyst[7]. As mentioned previously, the commonest age of presentation is the second or third decade, however, they can manifest soon after birth or during the neonatal period if they are associated with severe cardiopulmonary abnormalities[8]. In rare cases, they may remain latent until the seventh decade of life[3].

Adult patients with neurenteric cysts may present with focal pain at the level of the spinal pathology, radicular symptoms or fluctuating neurological signs. These symptoms are associated with lesions in the cervical and thoracic spine, whereas radicular symptoms are seen in persons with cysts in the cervical or lumbar spine.

The volumetric flux of the cyst associated with periodic leakage of fluid content secondary to osmotic and haemodynamic factors is responsible for the fluctuating nature of the symptoms[9]. The waxing and waning nature of the signs and symptoms associated with spinal cord compression secondary to the volumetric instability frequently leads to misdiagnosis of central nervous system demyelinating disorders[10] like multiple sclerosis. A variety of clinical manifestations have been seen in the paediatric population in addition to the common signs and symptoms. Case reports of children with presentations of aseptic meningitis, pyogenic meningitis, chronic pyrexia, incontinence and paraplegia , have been described[6,9,11,12].

Magnetic resonance imaging is the investigation of choice for the diagnosis of neurenteric cysts. The most common MRI findings associated with neurenteric cysts are non-contrast enhancing lesions that are isointense on T1-weighted sequences and hyperintense on T2 weighted images[13]. The incidence of neurenteric cysts along with osseous malformation warrants plain radiographs or CT imaging to fully delineate the pathologic spectrum of the process.

As in the reported case, surgical excision of the cystic mass is the first line of management for these cysts. Total excision of the mass is the ideal outcome given the

association between partial resection and cyst recurrence[13]. However, on occasion, vertebral anomalies or extensive adhesions to the neural anatomy makes complete resection hazardous and complicated[9]. Post-operative outcome of total resection is most often reported as curative of the sensory and motor deficits associated with the cysts. In literature, 11% patients have reported worsening of symptoms and 18% have had failure to regain pre-morbid neurological function[6]. Post-surgical recurrence have been reported in the range of 0%[14] and 37% with the later being reported in the longest follow-up at 30 years in eight patients[15].

## Conclusion:

In literature, there are case reports for neurenteric cysts in children and adults. However, this particular case reported serves to illustrate the atypical presentation of the spinal lesion with allodynia with secondary posturing, in an otherwise healthy child with no previous history of any neurological disease. With paucity of neurological signs, a diagnosis of somatisation and psychological disorders are easy to make with the actual disease process being completely missed. Due to the wide range of signs and symptoms, this diagnosis, though rare, should be kept in mind in order to diagnose and promptly treat this lesion before permanent deficits occur.

## References:


1. Savage JJ, Casey JN, McNeil IT, Sherman JH. Neurenteric cysts of the spine. *J Craniovertebr Junction Spine* 2010; **1:** 58-63.
2. Fortuna A, Mercuri S. Intradural spinal cysts. *Acta Neurochir* (Wien) 1983; **68:** 289-314.
3. Lippman CR, Arginteanu M, Purohit D, Naidich TP, Camins MB. Intramedullary neurenteric cysyts of the spine. Case report and review of literature. *J Neurosurg* 2001; **94:** 305-9.
4. Arai Y, Yamauchi Y, Tsuji T, Fukasaku S, Yokota R, Kudo T. Spinal neurenteric cyst. Report od two cases and review of forty-one cases in Japan. *Spine (Phila Pa 1976)* 1992; **17:** 1421-4.
5. Rao MB, Rout D, Misra BK, Radhakrishnan W. Craniospinal and spinal enterogenous cysts-report of three cases. *Clin Neurol Neurosurg* 1996; **98:** 32-6.
6. De Oliveira RS, Cinalli G, Roujeau T, Sainte-Rose C, Pierre-Kahn A, Zerah M. Neurenteric cysts in children: 16 consecutive cases and review of literature. *J Neurosurg* 2005; **103:** 512-23.
7. Rauzzino MJ *et al*. Spinal neurenteric cysts and their relation to more common aspects of occult spinal dysraphism. *Neurosurg Focus.* 2001; **10:** 1-10.
8. Cai C, Shen C, Yang W, Zhang Q, Hu X. Intraspinal neurenteric cysts in children. *Can J Neurol Sci* 2008; **35:** 609-615.
9. Garg N, Sampath S, Yasha TC, Chandramouli BA, Devi BI, Kovoor JM. Is total excision of neurenteric cysts possible? *Br J Neurosurg* 2008; **22:** 41-51
10. Vinters HV, Gilbert JJ. Neurenteric cysts of the spinal cord mimicking multiple sclerosis. *Can J Neurolo Sci* 1981; **8:** 159-61.
11. Shenoy SN, Raja A. Spinal neurenteric cyst. Report of 4 cases and review of literature. *Paediatr Neurosurg.* 2004; **40:** 284-92.
12. Rizk T, Lahoud GA, Maarrawi J, Hourani R, Jabbour P, Koussa S *et al*. Acute paraplegia revealing an intraspinal neurenteric cyst in a child. *Childs Nerv Syst* 2001; **17:** 754-7.
13. Menezes AH, Traynelis VC. Spinal neurenteric cysts in the magnetic resonance imaging era. *Neurosurgery* 2006; **58:** 97-105.
14. Cai C, Shen C, Yang W, Zhang Q, Hu X. Intraspinal neurenteric cysts in children. *Can J Neurol Sci* 2008; **35:** 609-15.
15. Chavda SV, Davies AM, Cassar –Pullicino VN. Enterogenous cysts of the central nervous system. A report of eight cases. *Clin Radiol* 1985; **36:** 245-51.