

A Rare Case of Isolated Conus Medullaris Neurenteric Cyst

Abstract

Spinal neurenteric cysts are very rare congenital anomalies and are commonly associated with other spinal malformations. They are usually located in intradural, extramedullary location. We report a rare case of intramedullary conus medullaris neurenteric cyst without any associated spinal malformation diagnosed preoperatively by magnetic resonance imaging.

Keywords: *Conus medullaris, neurenteric cyst, magnetic resonance imaging*

Introduction

Spinal neurenteric cysts are rare congenital lesions which are common in cervical and cervicothoracic regions of the spine.^[1] They are usually associated with other spinal malformations. Isolated conus medullaris neurenteric cyst without associated spinal malformations is extremely rare with only <10 cases reported in the literature. We hereby report a case of conus medullaris neurenteric cyst without associated spinal malformations which was diagnosed using magnetic resonance imaging (MRI).

Case Report

A 34-year-old male patient presented to neurosurgery outpatient department with complaints of pain over both lower limbs and lower back pain for the past 5 years, progressive weakness of the right lower limb for the past 4 years with decreased sensation, and wasting of the right lower limb muscles. The patient also complained of urinary incontinence for the past 1 year, which was worsening gradually. On examination, there was wasting of the right thigh and calf muscles with the power of 4/5 in the right lower limb. Routine blood investigations were normal. The patient was not a diabetic. MRI of the spine revealed the presence of a well-defined intradural T1 hyperintense lesion within conus medullaris and cauda equina nerve roots at D12 and L1 level on the right side measuring 3 cm × 1.2 cm × 1.1 cm [Figures 1a and 2a]. Lesion showed iso- to hypo-intense signal on T2-weighted

images [Figure 2b] with no suppression of T1 hyperintensity on fat-saturated images [Figure 1b]. The lesion appeared dark on inversion recovery sequences. There was no significant postcontrast enhancement. Rest of the spine was normal. The differential diagnosis would be epidermoid, dermoid, cystic neoplasm, or neurenteric cyst. The possibility of epidermoid was not considered since there was no diffusion restriction, and dermoid was ruled out since there was no fat. The lesion did not show any contrast enhancement and was hyperintense on T1-weighted image; hence, the possibility of cystic neoplasm was not considered. Based on the imaging appearance which is nonenhancing cystic T1 hyperintense lesion, a possibility of neurenteric cyst was given, and the patient was subjected to surgery. D12 and L1 laminectomy with initial incision of the cyst wall was done to decompress the cyst and to reduce trauma to the spinal cord and surrounding nerve roots. Cyst fluid was slightly thick and yellowish in nature [Figure 3]. Due to dense adhesion of the cyst wall with the surrounding nerve roots, only partial excision was done. The histopathology of cyst wall showed mucin-secreting columnar epithelium resting on a basement membrane, consistent with neurenteric cyst. The postoperative period was uneventful. The patient showed improvement in urinary continence and the back pain reduced. The patient was advised follow-up imaging every 6 months for the initial 1st year and annual MRI thereafter to detect recurrence. At 2-year follow-up, there was no evidence of recurrence.

How to cite this article: Aiyappan SK, Thirupathi Rajan VT, Harikrishnan V. A rare case of isolated conus medullaris neurenteric cyst. *Asian J Neurosurg* 2017;12:715-7.

Senthil Kumar Aiyappan, Vivakaran Thanga Thirupathi Rajan¹, Volga Harikrishnan²

Department of Radiodiagnosis and Imaging, SRM Medical College Hospital and Research Centre, Departments of ¹Neurosurgery and ²Pathology, Saveetha Medical College and Hospital, Kancheepuram, Tamil Nadu, India

Address for correspondence:
Dr. Senthil Kumar Aiyappan,
Department of Radiodiagnosis
and Imaging, SRM Medical
College Hospital and Research
Centre, Kattankulathur,
Kancheepuram - 603 203,
Tamil Nadu, India.
E-mail: [senthilkumarpgi@
yahoo.co.in](mailto:senthilkumarpgi@yahoo.co.in)

Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_92_14

Quick Response Code:



This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Discussion

Neurenteric cysts are rare congenital lesions of endodermal origin lined by the columnar epithelium of intestinal type and/or respiratory epithelium.^[1] Spinal neurenteric cysts are rare intradural extramedullary lesions of the spine which are commonly located in the cervical and thoracic regions. Majorities of them are localized ventral to the spinal cord and are associated with other vertebral anomalies. They account for 0.7%–1.3% of all spinal cord tumors and approximately 90% are located in intradural/extramedullary location and the rest 10% are intradural/intramedullary or extradural location.^[1,2] Associated spinal abnormalities are present in approximately 50% cases.^[1] Intramedullary neurenteric cyst of the conus medullaris without associated spinal malformation is a very rare lesion with only <10 cases reported in the English literature [Table 1].

Our case is similar to the case mentioned in the literature. Neurenteric cysts usually present in the second and third decades of life with male to female ratio of approximately 2:1.^[1] The clinical symptoms depend on the patient age, size, and location of the cyst.^[9] The most common symptom is spinal pain which can be local or radicular due to the

spinal cord or nerve root compression. Plain radiograph and MRI are the two important investigations required in the evaluation of spinal neurenteric cysts. Plain radiograph is required to evaluate bony spinal abnormalities such as hemivertebrae, spinal fusion, or spina bifida. On MRI, the signal intensity of the cyst depends on the content of the cyst. The cyst will show hyperintensity on T1-weighted images relative to cerebrospinal fluid (CSF) if it has high proteinaceous contents like in this case. On T2-weighted images, it can show hyper- or iso-intense signal relative to CSF. MRI is also helpful for the assessment of cyst location, extent, degree of the spinal cord, or nerve root compression.^[9,10] In our case, MRI was very helpful for planning the surgery. The treatment of neurenteric cyst is complete surgical removal. However, in most of the cases, complete resection is not possible due to the adhesion of cyst wall with surrounding neural structures. Overaggressive complete resection may sometimes lead to nerve or spinal cord injury, and in those cases, subtotal resection is enough. Subtotal resection has good immediate results, however, has higher recurrence rates.^[9,10] Lifelong follow-up with annual MRI is recommended to detect recurrence.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.



Figure 1: (a) T1 and (b) fat-saturated T1 sagittal magnetic resonance imaging images showing well-defined T1 hyperintense lesion (white arrows) involving conus medullaris and upper cauda equine nerve roots. Lesion is not showing loss of signal on fat-saturated images

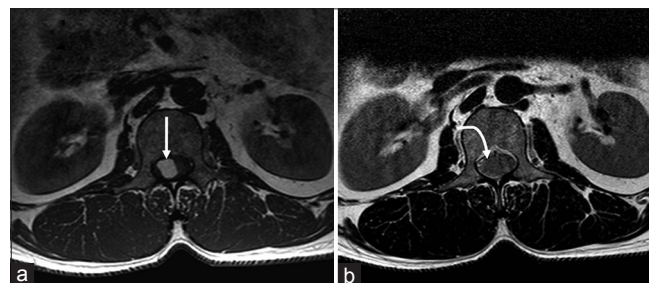


Figure 2: (a) T1 and (b) T2-weighted axial magnetic resonance imaging images showing well-defined T1 hyperintense lesion (straight white arrow) within the right side of conus medullaris which is isointense on T2-weighted images (curved white arrow)

Table 1: Summary of literature

First author	Age (years), sex of patient	Location	Surgery
Rewcastle and Francoeur, 1964 ^[3]	34/female	T7–L2	Subtotal resection
Fortuna and Mercuri, 1983 ^[4]	47/female	T11–L1	Subtotal resection
Rivierez <i>et al.</i> , 1997 ^[5]	46/female	L1–L2	Cystosubarachnoid shunt
Rotondo <i>et al.</i> , 2005 ^[6]	52/female	T12–L1	Total resection
Rotondo <i>et al.</i> , 2005 ^[6]	61/female	T12–L1	Total resection
Balasubramaniam <i>et al.</i> ^[2]	21/male	T12–L1	Near total resection
Park <i>et al.</i> , 2007 ^[7]	6/female	L1–L2	Subtotal resection
Jaiswal <i>et al.</i> , 2011 ^[8]	35/male	L1–L2	Total resection
Sadeghi-Hariri <i>et al.</i> , 2012 ^[9]	40/male	L1–L2	Total resection
Present case	34/male	T12–L1	Subtotal resection

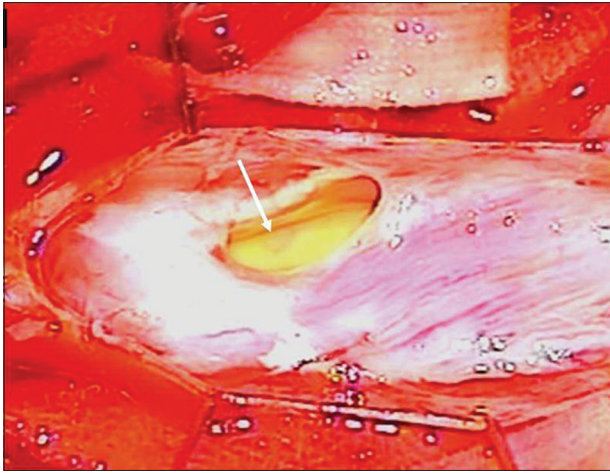


Figure 3: Intraoperative microscopic view showing clear thick yellowish contents of neurenteric cyst (white arrow) within conus medullaris

References

1. Savage JJ, Casey JN, McNeill IT, Sherman JH. Neurenteric cysts of the spine. *J Craniovertebr Junction Spine* 2010;1:58-63.
2. Balasubramaniam S, Tyagi DK, Sawant HV. Intramedullary enterogenous cyst of the conus medullaris presenting as lower limb pain. *J Craniovertebr Junction Spine* 2011;2:46-8.
3. Rewcastle NB, Francoeur J. Teratomatous cysts of the spinal canal; with "sex chromatin" studies. *Arch Neurol* 1964;11:91-9.
4. Fortuna A, Mercuri S. Intradural spinal cysts. *Acta Neurochir (Wien)* 1983;68:289-314.
5. Rivièrez M, Buisson G, Kujas M, Ridarch A, Mignon E, Jouannelle A, *et al.* Intramedullary neurenteric cyst without any associated malformation. One case evaluated by RMI and electron microscopic study. *Acta Neurochir (Wien)* 1997;139:887-90.
6. Rotondo M, D'Avanzo R, Natale M, Pasqualetto L, Bocchetti A, Agozzino L, *et al.* Intramedullary neurenteric cysts of the spine. Report of three cases. *J Neurosurg Spine* 2005;2:372-6.
7. Park YB, Kim SH, Kim SW. An intramedullary neurenteric cyst in the conus medullaris with recurrent meningitis. *J Korean Neurosurg Soc* 2007;41:130-3.
8. Jaiswal S, Jaiswal AK, Paul L, Behari S, Mahapatra AK. Neurenteric cyst in the region of conus medullaris. *Pan Arab J Neurosurg* 2011;15:80.
9. Sadeghi-Hariri B, Khalatbari MR, Hassani H, Taheri B, Abbassioun K. Intramedullary neurenteric cyst of the conus medullaris without associated spinal malformation: A case report and review of the literature. *Turk Neurosurg* 2012;22:478-82.
10. Vachhani JA, Fassett DR. Intramedullary neurenteric cyst associated with a tethered spinal cord: Case report and literature review. *Surg Neurol Int* 2012;3:80.