


A Rare Intradural Extramedullary Neurenteric Cyst in the Cervical Spine

Mohammad Ali Abouei Mehrizi¹; Narges Hashemi²; Mehran Sedaghat³;
Neshat Najaf Najafi^{4*} ; Azin Eshaghzade⁴

¹Department of Neurosurgery, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

²Department of Pediatrics, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

³Neurosurgery department, Mir Hoseini private hospital, Hejrat Blvd. Shiraz, Fars province, Iran

⁴Student Research Committee, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

Received: 26 Jul 2025
Accepted: 23 Nov 2025
Published: 1 Apr 2026

ABSTRACT

Neurenteric cysts (NCs), also known as endodermal cysts, are rare congenital lesions of endodermal origin. They were thought to originate from a failure of dissolution of the neurenteric canal between the notochord and endoderm during the third week of embryogenesis. The primary clinical symptoms differ from focal pain to paresthesia and weakness based on the cyst's location. This study presents a case of intradural extramedullary NC of the spine at the cervical level. A 10-year-old boy was referred to the hospital, with neck pain and weakness in his left hand. Imaging studies revealed fusion of cervical vertebrae at the levels of C3-C4 and C5-C6. On MRI, a cystic lesion in the intradural extramedullary region compressing the cord was evident. As the preferred treatment option, surgical total resection was completed through the posterior approach, and the definitive diagnosis was made based on the histochemical analysis. NC is a rare lesion and should be kept in mind as a differential diagnosis. Surgical intervention, particularly total resection, frequently improves the sensory and motor deficits associated with NC and is the treatment of choice.

Keywords:

Congenital cyst
Endodermal cyst
Neurenteric cyst
Spinal cyst

How to cite this article: Abouei Mehrizi MA, Hashemi N, Sedaghat M, Najaf Najafi N, Eshaghzade A. A Rare Intradural Extramedullary Neurenteric Cyst in the Cervical Spine. *Iran J Child Neurol.* 2026;20(2): 61-64. <https://doi.org/10.22037/ijcn.v20i2.49293>

Introduction

Neurenteric (endodermal) cysts (NCs) are rare, benign central nervous system lesions that often occur intraspinally due to abnormal embryological development of the notochord (1-4).

Additionally, NCs usually appear in the second or third decade of life and are prevalent in men (5). Besides, they are commonly discovered in the mediastinum (6).

These lesions usually appear incidentally on neuroimaging or with characteristic symptoms of mass effects, such as headache and dizziness, aseptic meningitis due to irritation from local leaking of cystic

material, and myelopathy in the case of intraspinal lesions (4, 7).

NC typically appears as a lobulated or cystic intradural extramedullary mass on neuroimages, particularly when other kinds of vertebral abnormalities exist (7). Despite the fact that this lesion has traditionally been regarded as benign histologically, malignant change is incredibly unlikely but not impossible (8).

This study presents a case of a 10-year-old boy with endodermal cyst of the anterolateral cord with cord compression and fusion of vertebrae C3-C4 and C5-C6.

Corresponding Author:

Neshat Najaf Najafi
Email: najafnajafin971@mums.ac.ir



This work is licensed under a Creative Commons Attribution-NonCommercial 4.0 International (CC BY-NC 4.0) which allows users to read, copy, distribute and make derivative works for non-commercial purposes from the material, as long as the author of the original work is cited properly.

Case Presentation

A 10-year-old boy with complaints of neck pain and weakness in his left hand was referred to the hospital. In the imaging studies of this patient, vertebral anomaly fusion was observed in the C3-C4 and C5-C6 levels (Figure 1). On magnetic resonance imaging (MRI), a cystic lesion in the intradural extramedullary region of the anterolateral cervical cord was observed, compressing the cord. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images (Figure 2, 3). The patient underwent posterior laminotomy approach and the cyst was resected subtotally and sent for pathology, confirming the presence of a NC. Subsequently, laminoplasty was performed. No complications were found following surgery, and the patient was then released without neurologic abnormalities.



Figure 1. CT scan of the spine in sagittal (a) and coronal (b) view, showing C3-C4 and C5-C6 fusion

Discussion

NCs of the CNS are congenital, rare lesions of endodermal origin that most frequently occur in the spine (5). Kubie et al. in 1928 first described this lesion as a *teratomatous cyst* (9). Later on, Puusepp in 1934 described it as an *intestinoma* (10), and finally, Holcomb et al. in 1954 used the term neurenteric cyst (11). The etiology of this lesion goes back to the third week of embryogenesis, when the separation of the notochord and endoderm does not happen because of the persistence of the normally transient neurenteric canal. NCs are rare, comprising almost 0.7–1.3% of the spinal axis tumors (5). Reportedly, approximately 78–90% of NCs are placed in the intradural extramedullary compartment. The ventral and ventro-lateral aspects of the spinal cord are common locations. They typically involve the cervical, thoracic, and lumbosacral spines, respectively (12). As in the present case, the lesion was located in the intradural extramedullary region of the cervical spine. Besides, although most NCs are solitary lesions, disseminated ones have been described (5, 13).

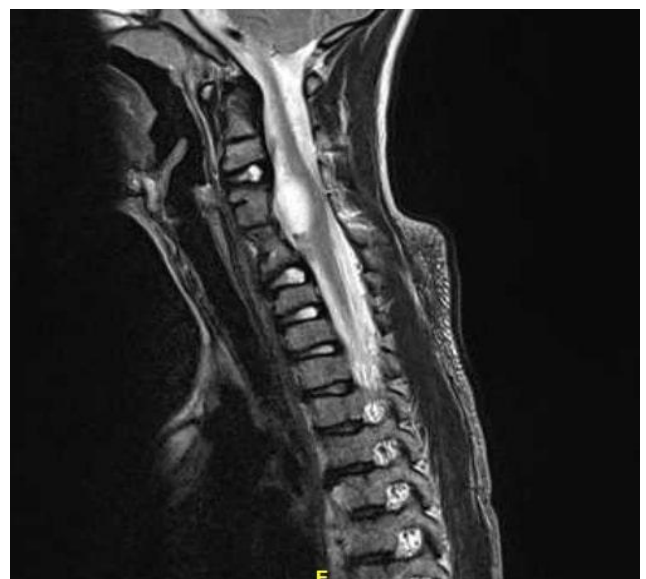


Figure 2. MRI T1W (a) and T2W (b) in sagittal planes, showing a cystic lesion in the intradural extramedullary region, compressing the cord. The lesion was hypointense on T1 and hyperintense on T2.

The primary clinical symptoms differ based on the cyst's location and spinal level. Symptoms consist mostly of focal pain at the lesion level, radicular pain along the dermatome, paresthesia, weakness, urinary incontinence, and signs of myelopathy such as hyperreflexia and spasticity (5). For instance, in the present case, the symptoms were mostly neck pain and weakness in his left hand.

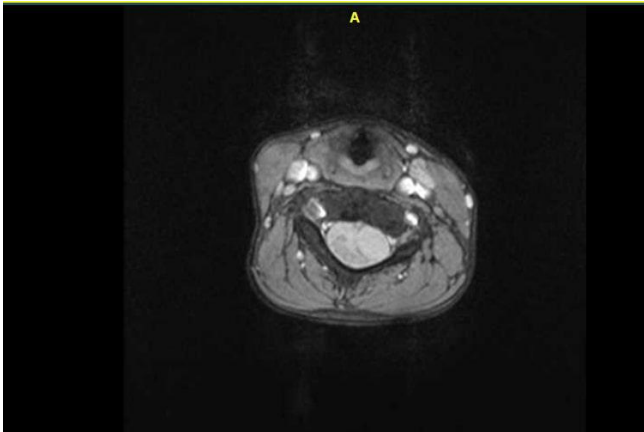


Figure 3. MRI T2W in Axial view plane, shows a ventrally located cyst compressing the spinal cord

The imaging modality of choice in NC is MRI. The only benefit of a computed tomography (CT) scan consists of displaying related spinal canal abnormalities. The imaging intensity pattern on MRIs may be erratic due to variations in cystic content (5). Typical indications of a NC on an MRI include ventral location, isointensity on T1-weighted scans, hyperintensity on T2-weighted scans, and a lack of enhancement with gadolinium (5, 14). Considering the present case, the imaging studies revealed cervical vertebral fusion. The MRI showed a cystic lesion in the intradural extramedullary region, compressing the

cord. The lesion was hypointense on T1 and hyperintense on T2 (Figure 2).

The first-line treatment for NC is surgical resection. Total excision, fenestration of the cyst, simple aspiration, and subtotal resection are the surgical options (12). If feasible, total excision is the preferred effect because of the link between partial resection and the potential return of the cyst (5). The surgical approach that has been reported most frequently is the posterior technique. The success rate of surgery is extraordinary; therefore, most patients experience significant improvement in symptoms after surgery. Surgical intervention, particularly total resection, commonly improves the sensory and motor deficits associated with NC (15).

In Conclusion

The case of a 10-year-old boy with cervical vertebrae fusion and a NC causing spinal cord compression is a rare. Hence, serious condition requires prompt diagnosis and treatment. Surgical resection is the preferred treatment option, and the prognosis for patients undergoing surgery is generally reliable.

Acknowledgement

Not applicable.

Authors' Contributions

Mohammad Ali Abouei Mehrizi and Narges Hashemi helped with the conception and design of the work. All authors helped for Data collection and drafting the article. Furthermore, all authors read and approved the final manuscript.

Conflict of Interest

The authors declared no conflict of interest.

References

1. Anderson T, Kaufman T, Murtagh R. Intracranial neurenteric cyst: A case report and differential diagnosis of intracranial cystic lesions. *Radiol Case Rep.* 2020;15(12):2649-54.
2. Cheng JS, Cusick JF, Ho KC, Ulmer JL. Lateral supratentorial endodermal cyst: case report and review of literature. *Neurosurgery.* 2002;51(2):493-9; discussion 9.
3. Harris CP, Dias MS, Brockmeyer DL, Townsend JJ, Willis BK, Apfelbaum RI. Neurenteric cysts of the posterior fossa: recognition, management, and embryogenesis. *Neurosurgery.* 1991;29(6):893-7; discussion 7-8.
4. Wang L, Zhang J, Wu Z, Jia G, Zhang L, Hao S, et al. Diagnosis and management of adult intracranial neurenteric cysts. *Neurosurgery.* 2011;68(1):44-52; discussion
5. Savage JJ, Casey JN, McNeill IT, Sherman JH. Neurenteric cysts of the spine. *J Craniovertebr Junction Spine.* 2010;1(1):58-63.
6. Jeung MY, Gasser B, Gangi A, Bogorin A, Charneau D, Wihlm JM, et al. Imaging of cystic masses of the mediastinum. *Radiographics.* 2002;22 Spec No:S79-93.
7. Brooks BS, Duvall ER, el Gammal T, Garcia JH, Gupta KL, Kapila A. Neuroimaging features of neurenteric cysts: analysis of nine cases and review of the literature. *AJNR Am J Neuroradiol.* 1993;14(3):735-46.
8. Ho LC, Olivi A, Cho CH, Burger PC, Simeone F, Tihan T. Well-differentiated papillary adenocarcinoma arising in a supratentorial enterogenous cyst: case report. *Neurosurgery.* 1998;43(6):1474-7.

9. Kubie LS. A clinical and pathological study of two teratomatous cysts of the spinal cord, containing mucus and ciliated cells. *Surg Gyn Obst.* 1928;47:297-311.
10. Puusepp M. Variété rare de téréatome sousdural de la région cervicale (intestinome): quadriplégie, extirpation, guérison complète. *Rev Neurol.* 1934;2:879-86.
11. Holcomb GW, Jr., Matson DD. Thoracic neurenteric cyst. *Surgery.* 1954;35(1):115-21.
12. Baek WK, Lachkar S, Iwanaga J, Oskouian RJ, Loukas M, Oakes WJ, et al. Comprehensive Review of Spinal Neurenteric Cysts with a Focus on Histopathological Findings. *Cureus.* 2018;10(9):e3379.
13. Oyama H, Ikeda A, Inoue S, Nakamura S, Nishimura Y, Shibuya M. Multiple neurenteric cysts in the posterior fossa and cervical spinal canal--case report. *Neurol Med Chir (Tokyo).* 2004;44(3):146-9.
14. Menezes AH, Traynelis VC. Spinal neurenteric cysts in the magnetic resonance imaging era. *Neurosurgery.* 2006;58(1):97-105; discussion 97-.
15. 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 the literature. *J Neurosurg.* 2005;103(6 Suppl):512-23.