HAYDARPAŞA NUMUNE MEDICAL JOURNAL

DOI: 10.14744/hnhj.2022.14892
Haydarpasa Numune Med J 2023;63(2):223–226


CASE REPORT

# Conflicting Behavior of Glioependymal Cyst: A Report of Three Cases and Review of Literature

**Ersin Hacıyakupoğlu[1], Ali Erhan Kayalar[2], Mustafa Efendioğlu[2], Sebahattin Hacıyakupoğlu[3]**

[1]Department of Neurosurgery, Heinrich-Braun-Klinikum Zwickau
[2]Department of Neurosurgery, University of Health Sciences Türkiye, Haydarpaşa Numune Health Application and Research Center, Istanbul, Türkiye
[3]Department of Neurosurgery, Acıbadem University, Türkiye

## Abstract

Three cases evaluated as glial tumor were operated and diagnosed incidentally as conus medullaris cystic lesion. In the first case, following a traffic accident, paraparesis and urinary incontinence developed. A collapsed fracture at L2 was determined on lumbar Magnetic Resonance Imaging (MRI), so posterior fusion was applied with a transpedicular screw. The urinary incontinence did not improve, and on the follow-up MRI, a cystic mass was determined in the conus medullaris, so gross total surgery was performed. The other two cases had complaints of low back pain. The neurological and systematic examinations were normal. A cystic mass was determined in the conus medullaris on the lumbar MRI taken in each case and as these were thought to be glial tumors, partial excision, and marsupialization was applied. The pathological diagnosis in these three cases was glia ependymal cyst. The cases are presented with a review of the relevant literature.

Keywords: Choroidal cyst; ependymal cyst; glioependymal cyst; neuroepithelial cyst.

Glioependymal cyst, neuroepithelial cyst, and choroidal cyst are used with the same meaning. A glioependymal cyst is a rare lesion constituting 0.01% of all central nervous system (CNS) tumors. Although tumors may be found in the whole CNS, they are 3 times more common in the spine than in the brain[1]. They may be seen in all the areas of the medulla spinalis from the craniocervical junction to the coccyx. The majority of cysts localized in the medulla spinalis is situated anteriorly within the spinal canal and may be related to mediastinal or abdominal cysts through a defect in the vertebral body. Bowel, bladder, or renal abnormalities are seen in patients[2,3]. Glioependymal cysts are composed of a lining of a single layer of epithelium and are classified as neuroepithelial and endodermal types according to the origin. Neuroepithelial cysts have historically been described as ependymal and choroidal. Endodermal cyst refers to colloid, enterogenous, neurenteric, and respiratory epithelial types[1]. Glioependymal cysts are of endodermal origin. They occur during the 3rd week of gestation with an abnormal connection between the primitive gut and the neurectoderm, and this fistula prevents normal notochord formation. The primary pathology is abnormal notochord formation[4].

**Correspondence (İletişim):** Ersin Hacıyakupoğlu, M.D. Department of Neurosurgery, Heinrich-Braun-Klinikum Zwickau
**Phone (Telefon):** +90 533 573 18 80 **E-mail (E-posta):** draekayalar@gmail.com
**Submitted Date (Başvuru Tarihi):** 09.01.2022 **Revised Date (Revize Tarihi):** 02.08.2022 **Accepted Date (Kabul Tarihi):** 08.08.2022

In addition to immunohistochemistry and electron microscopy, computed tomography (CT), and magnetic resonance imaging (MRI) are important in diagnosis[3]. Glioependymal cysts are seen slightly more in males than females and can be seen up to the 5th decade of life. Focal expanding mass lesion findings are obtained according to the localization. Pain is experienced with all spinal lesions and dysfunction develops later[2]. The optimal treatment is total excision, but partial excision, marsupialization, fenestration, cystoperitoneal, and cystoarachnoidal shunt can also be applied. Recurrence is rarely seen[5,6].

## Case Report

**Case 1** – A 60-year-old male presented with findings of L2 compression fracture, paraparesis, and incontinence following a traffic accident on April 19, 2016. Posterior fusion of L5 with the root was applied with a transpedicular screw and L2 vertebroplasty was performed. Despite the recovery of paresis, the incontinence did not improve. A follow-up MRI was taken on May 25, 2016. A cystic lesion was determined almost intramedullary between L2 and inferior of the T12 corpus with a diameter of 8.5 mm in the axial plane and elevation of 34 mm in the sagittal plane, which was isointense on T1-weighted images and mildly hyperintense compared to cerebral spinal fluid (CSF) on T2-weighted images. It was reported as an arachnoid cyst secondary to trauma and surgery. Gross total surgical resection was applied on May 28, 2016 (Fig. 1). Incontinence did not recover. The pathological diagnosis was reported as a glioependymal cyst.

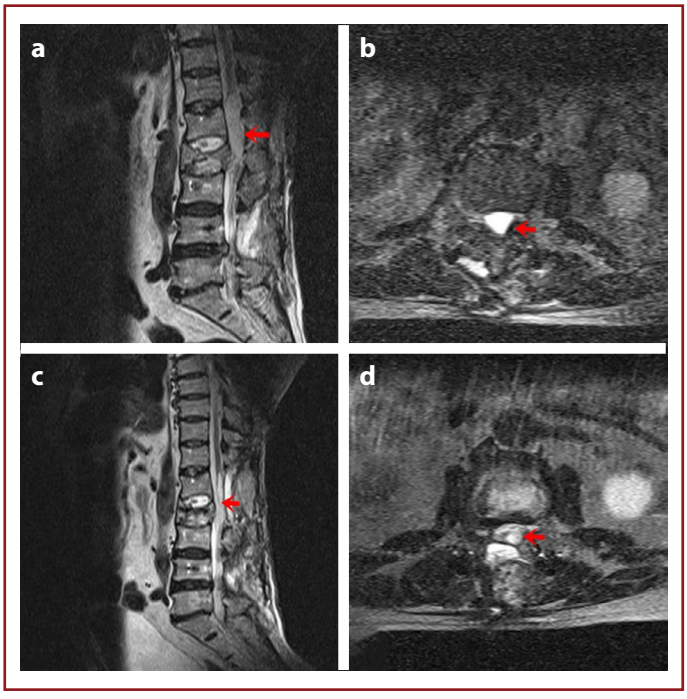


**Figure 1.** Glioependymal cyst total excision (a and b pre-op c and d post-op) L2 compression fracture.

**Case 2** – A 15-year-old female presented with complaints of low back pain. A cystic mass was determined on lumbar MRI, so partial resection and marsupialization were performed on May 20, 2019. There was no neurological deficit, and no additional deficit postoperatively (Fig. 2). The low back pain recovered after the operation. The pathological diagnosis was reported as a glioependymal cyst (Fig. 3).

**Case 3** – A 20-year-old female presented with complaints of low back pain. Despite no neurological deficit, a lumbar MRI was taken, on which a cystic mass was determined in the conus medullaris, so partial resection and marsupialization were surgically applied. The low back pain recovered after the operation. The pathological diagnosis was reported as a glioependymal cyst (Fig. 4).

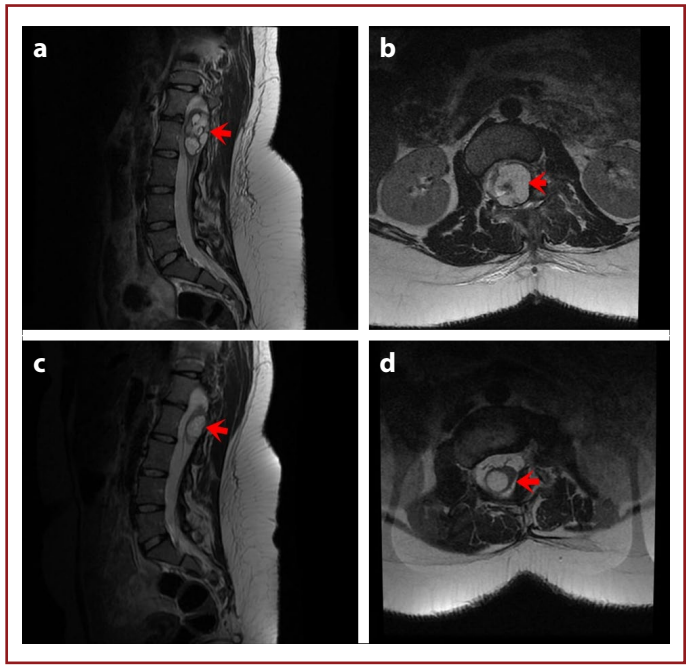


**Figure 2.** Partial excision (a and b pre op, c and d post op).

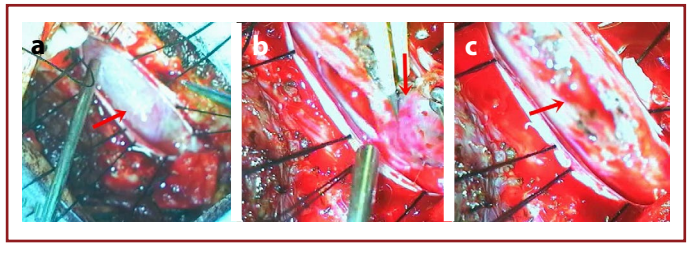


**Figure 3. (a-c)** Intra-operative view.

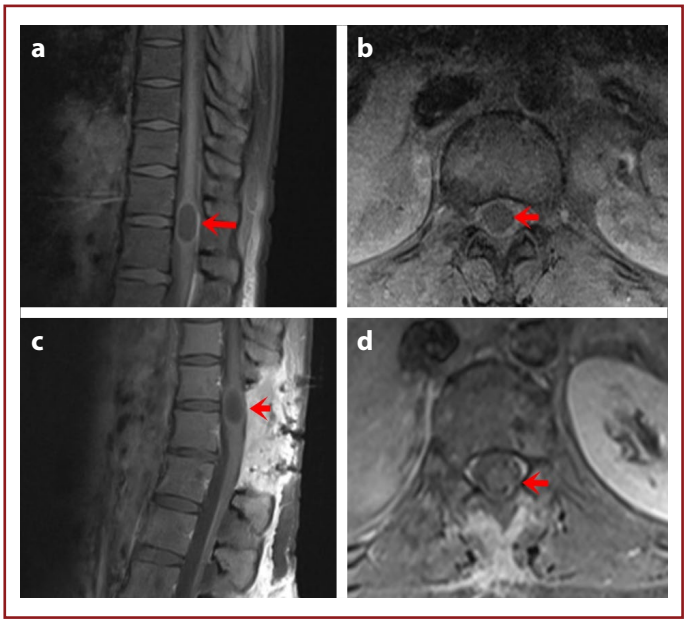


**Figure 4.** Marsupialization (a and b pre op c and d post op).

## Discussion

Glioependymal cysts are also known as neuroepithelial cysts, ependycystscyst, and choroidal cysts. They are thought to arise from ectopic remnants of primitive neuroglial fissure and can therefore occur anywhere in the neuraxis[1,7]. These cysts are three times more common in the spine than in the brain[4,6-8]. Most of these lesions are anterior to the spinal canal and may communicate with a mediastinal or abdominal cyst through a defect in the vertebral body[4]. This contribution is greater in patients of a young age. Bowel, bladder, renal, and cranial anomalies may be seen in patients. A glioependymal cyst is seen more rarely in the brain's posterior cerebellopontine angle and craniovertebral junction[3,9,10]. The current cases did not develop any mental anomaly and there were no expanding mass lesion findings such as headache, seizures, or focal neurological deficits. There were no bowel, bladder, or renal abnormalities. The urinary and fecal incontinence in case no. It was explained as traumatic myelomalacia at L1 and L2 levels. A glioependymal cyst is of endodermal origin. It develops in the 3rd week of gestation when the endoderm fuses with the developing notochord[3,6]. As it grows extremely slowly, neural tissues may adapt to the expansion and neurological findings emerge extremely late. There was no fusion defect in any of the current cases. All three cases were diagnosed incidentally on lumbar MRI. An expansive cystic mass was determined in the conus medullaris, which was hypointense on T1-weighted images and hyperintense on T2-weighted images Glioependymal cysts appear as non-enhancing, CSF-containing, unilocular thin-walled cysts, found both in intra-axial and extra-axial localizations. The current cases were all intra-axial. Differentiation of these lesions from the arachnoid cyst, enterogenous cyst, meningoencephalocele, and congenital dysplasia is difficult[2,3,6]. The cyst wall is composed of an inner glial layer with a luminal ependymal lining and an outer fibrous layer.

Ependymal cells have vacuoles, a bleb-like protrusion, normal and abnormal cilia, and microvilli. These cysts have neither pinocytic vesicles nor a basal membrane. The glial layers contain astrocytes and ependymal cells[2,5]. A definitive diagnosis is made from MRI and CT images and cannot be made from observation in the operation or with light microscopy. Immunohistochemistry and electron microscopy are important in diagnosis. Two different types can be distinguished on an electron microscope[1,8].

In the first type, there is the pseudo-stratified columnar epithelium, greater cuboidal epithelial areas, and fewer goblet cells producing mucinous fluid. In the second type, non-ciliated cells and microvilli are resembling respiratory simple columnar epithelium, and there is much more secretion[1,11]. Cytokeratin is an extremely specific marker in the diagnosis of epithelial membrane antigen and especially carcinoembryonic membrane antigen. In addition to these, GFAP and NSE were positive in the current cases[12]. If a preoperative definite diagnosis can be made of the glioependymal cyst, expansion can be allowed until it produces symptoms[13]. As urinary incontinence was present because of trauma in one of the current cases, the operation was performed to determine whether or not the symptom was related to the mass in the conus medullaris. The other two cases had low back pain complaints and no neurological findings.

## Conclusion

Both patients were operated on with the thought that it could be a glial tumor. The pain recovered and no additional neurological deficit occurred. Total excision is important in treatment. Gross total excision was applied to the first case and as there were no neurological deficits in the other two cases, partial excision marsupialization station to the subarachnoid space were applied. No recurrence was seen in any of the cases.

**Informed Consent:** Written informed consent was obtained from the patient's for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed.

**Conflict of Interest:** None declared.

**Authorship Contributions:** Concept: E.H.; Design: A.E.K.; Supervision: S.H.; Fundings: M.E.; Materials: E.H.; Data Collection or Processing: A.E.K.; Analysis or Interpretation: E.H.; Literature Search: M.E.; Writing: A.E.K.; Critical Review: S.H.

**Financial Disclosure:** The authors declared that this study received no financial support.

## References

1. Christov C, Chrétien F, Brugieres P, Djindjian M. Giant supratentorial enterogenous cyst: Report of a case, literature review, and discussion of pathogenesis. Neurosurgery 2004;54:759–63. [CrossRef]
2. Umredkar A, Mohindra S, Gupta R, Bal AK. Contrasting behavior of glio-ependymal cysts: A report of two cases and literature review. Neurol India 2010;58:659–61. [CrossRef]
3. Kavar B, Kaye HA. Brain Tumors. In: Kaye HA, Laws RE, editors. Dermoid, epidermoid and neurenteric cysts. 3rd ed. United States: Saunders, Elsevier Press; 2012. p.831–48. [CrossRef]
4. Osborn AG. Diagnostic Imaging Brain. In: Osborn AG, Salzman KL, Jhaveri MD, editors. Neurenteric Cyst. 2nd ed. Salt Lake City: Amirsys press; 2015. p.650–1.
5. Boockvar JA, Shafa R, Forman MS, O'Rourke DM. Symptomatic lateral ventricular ependymal cysts: Criteria for distinguishing these rare cysts from other symptomatic cysts of the ventricles: Case report. Neurosurgery 2000;46:1229–33. [CrossRef]
6. Obaldo RE, Shao L, Lowe LH. Congenital glioependymal cyst presenting with severe proptosis. AJNR Am J Neuroradiol 2007;28:999–1000. [CrossRef]
7. Balasubramaniam C, Balasubramaniam V, Santosh V. Intramedullary glioependymal cyst and tethered cord in an infant. Childs Nerv Syst 2004;20:496–8. [CrossRef]
8. Shuangshoti S, Pitakdamrongwong N, Poneprasert B, Bhavilai D, Kasantikul V. Symptomatic neuroepithelial cysts in the posterior cranial fossa. Immunohistochemical and electron-microscopic studies. Surg Neurol 1988;30:298–304. [CrossRef]
9. Schelper RL, Kagan-Hallet KS, Huntington HW. Brainstem subarachnoid respiratory epithelial cysts: Report of two cases and review of the literature. Hum Pathol 1986;17:417–22. [CrossRef]
10. Takumi I, Mori O, Mizutani N, Akimoto M, Kobayashi S, Teramoto A. Expansile neurenteric cyst arising in the frontal lobe associated with status epilepticus: Report of a case and discussion of epileptogenesis. Brain Tumor Pathol 2008;25:97–101. [CrossRef]
11. Kachur E, Ang LC, Megyesi JF. Intraparenchymal supratentorial neurenteric cyst. Can J Neurol Sci 2004;31:412–6. [CrossRef]
12. Zalatnai A. Neurenteric cyst of medulla oblongata--a curiosity. Neuropediatrics 1987;18:40–1. [CrossRef]
13. Walls TJ, Purohit DP, Aji WS, Schofield IS, Barwick DD. Multiple intracranial enterogenous cysts. J Neurol Neurosurg Psychiatry 1986;49:438–41. [CrossRef]