Article published online: 2023-08-14

A Neuroenteric Cyst of the Cavernous Sinus: A Case Report

Walid Elshamy[1,2] Burak Ozaydin[1] Sima Sayyahmelli[1] M. Shahriar Salamat[1,3] Mustafa K. Baskaya[1]

[1] Department of Neurological Surgery, University of Wisconsin School of Medicine and Public Health, Madison, Wisconsin, United States
[2] Department of Neurological Surgery, Faculty of Medicine, Ain Shams University, Cairo, Egypt
[3] Department of Pathology and Laboratory Medicine, University of Wisconsin School of Medicine and Public Health, Madison, Wisconsin, United States

Address for correspondence Mustafa K. Baskaya, MD, Department of Neurological Surgery, University of Wisconsin School of Medicine and Public Health, 600 Highland Avenue, CSC, K4/822, Madison, WI 53792, United States (e-mail: baskaya@neurosurgery.wisc.edu).

J Neurol Surg Rep 2023;84:e80–e84.

Abstract

Background Neuroenteric cysts (NECs) are benign lesions mostly found as intradural extramedullary lesions in the cervicothoracic spinal cord. NECs in the cavernous sinus are very rare. To the best of our knowledge, this is only the second reported case and the first in an adult.

Presentation We present a left cavernous sinus NEC in a 75-year-old female with gradually worsening headache and facial pain unresponsive to medical treatment. Imaging revealed a cystic mass lesion in the left cavernous sinus encasing the distal petrosal and cavernous segment of the internal carotid artery. Initial differential diagnoses included more common pathologies located near the cavernous sinus, including cystic schwannoma, craniopharyngioma, and dermoid and epidermoid tumors. The patient underwent a left pterional craniotomy with an extradural transcavernous approach for surgical exploration and possible resection of this mass lesion. Histopathology revealed an NEC lined with benign respiratory-type epithelium. Postoperative imaging revealed gross total tumor resection. The patient remained neurologically intact with complete resolution of facial pain.

Conclusion We present a rare pathology that can easily be misinterpreted as other types of lesions. NECs should be kept in mind for differential diagnosis of cavernous sinus cystic lesions. The surgical aim should be maximal safe excision.

Keywords
- cavernous sinus
- Meckel's cave
- neuroenteric cyst
- trigeminal nerve

Introduction

Neuroenteric cysts (NECs) are rare endodermal-derived lesions of the central nervous system (CNS). Several names have been coined for NECs including enterogenous cyst, endodermal cyst, gastroenterogenous cyst, gastrocytoma, intestinoma, and archenteric cyst.[1] NECs were first described by Puusepp in 1934.[2] These represent 0.01% of CNS tumors along with the other endodermal-derived cystic lesions, including Rathke's cleft cysts and colloid cysts.[3] NECs originating in the CNS are most commonly found in the spinal cord as intradural extramedullary cysts ventral to the cervicothoracic junction,[4,5] representing 0.7 to 1.3% of all intraspinal lesions.[5,6] Most NECs occurring intracranially are

received
February 24, 2020
accepted after revision
June 13, 2023

DOI https://doi.org/10.1055/s-0043-1772157.
ISSN 2193-6358.

© 2023. The Author(s).
This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (https://creativecommons.org/licenses/by-nc-nd/4.0/)

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

located in the craniocervical junction and anterior to the brain stem,[7,8] with some reported cases located supratentorially.[9,10]

Cavernous sinus NECs are very rare. To the best of our knowledge, this report is only the second reported case and the first in an adult. The objective of this case report is to describe this rare pathology, in a rare location, which may be misinterpreted as another more common lesion.

Case Report

A 75-year-old female presented with gradually worsening headache and left-sided episodic facial pain occurring over the last 6 months which was unresponsive to medical treatment. This was initially misinterpreted as a temporomandibular joint problem because of a prior history of periodontal abscess for which she received physical therapy with no improvement. She then sought neurosurgical advice. The neurological examination was normal with no cranial nerve dysfunction except left-sided facial pain in the V2 and V3 distribution of the trigeminal nerve. Contrast-enhanced computerized tomography (CT) of the brain showed a lobulated, slightly hyperdense, septate, and peripherally enhancing mass centered within the left cavernous sinus and encasing the distal petrosal and cavernous segment of the left internal carotid artery (ICA), with widening of the left foramen ovale and superior orbital fissure. In addition, some scattered minute calcifications were observed within the isodose mass (►**Fig. 1**).

Magnetic resonance imaging (MRI) of the brain with contrast revealed a heterogeneously enhancing, partially cystic mass measuring 3.0 cm in largest dimensions in the left cavernous sinus (►**Fig. 2**).

Differential diagnoses included cystic schwannoma, cystic meningioma, poorly enhancing chondrosarcoma, craniopharyngioma, and dermoid and epidermoid cysts. The patient was offered surgery as there was a failure of

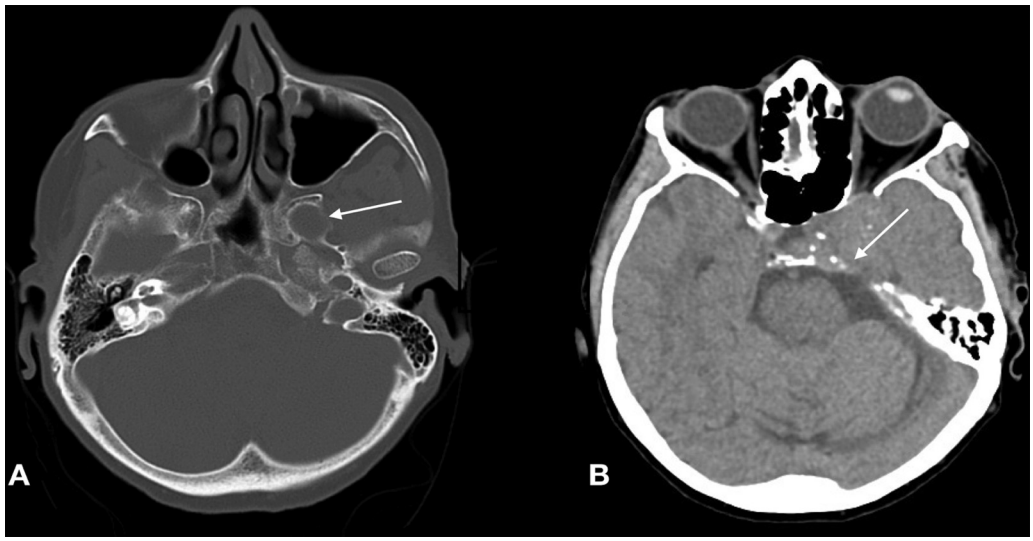

Fig. 1 Preoperative axial CT of the head in bone window (**A**) shows widening of the foramen ovale (*white arrow*), and brain window (**B**) shows an isodose mass lesion in the left cavernous sinus (*white arrow*).

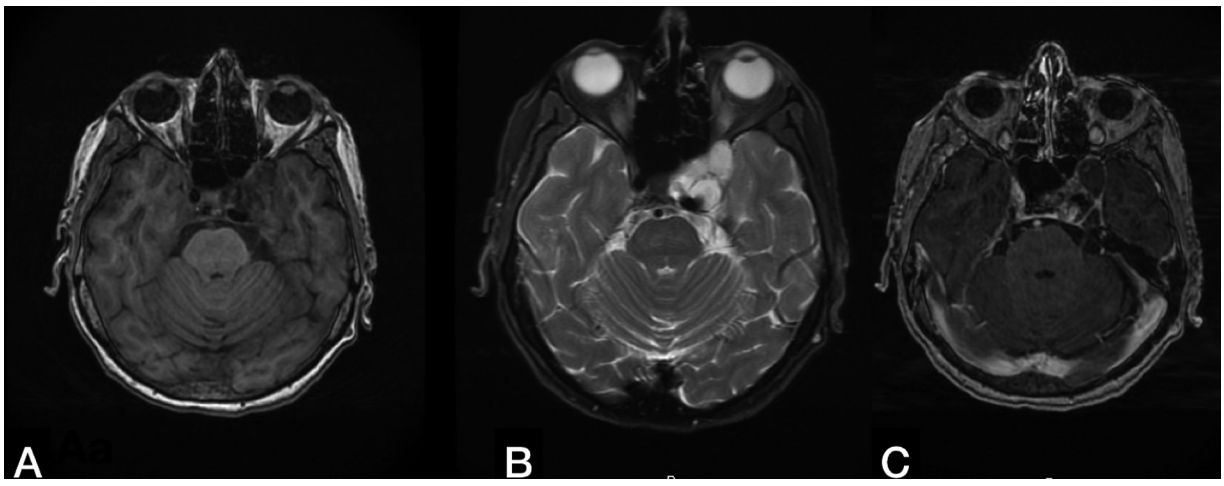

Fig. 2 Preoperative axial T1-weighted (**A**), T2-weighted (**B**), and T1-weighted with contrast enhancement (**C**) MRIs show a cystic mass lesion with septations in the left cavernous sinus. Note the contrast enhancement in the cyst wall and septations.

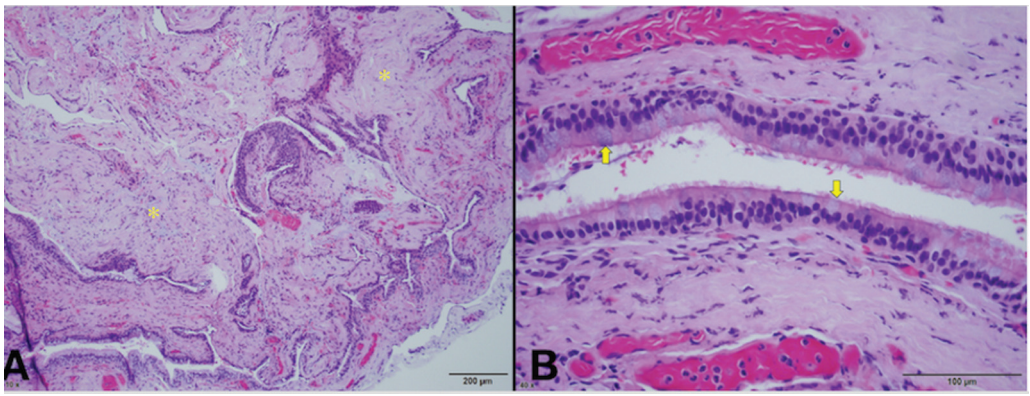

Fig. 3 H&E stain reveals a complex cystic structure with a generous fibrotic wall (asterisks), original magnification 10X (**A**). H&E stained section of the cyst lining at original magnification of 40x reveals a pseudostratified columnar ciliated epithelium that several goblet cells (*arrows*), similar to the lining of the respiratory tract (**B**).

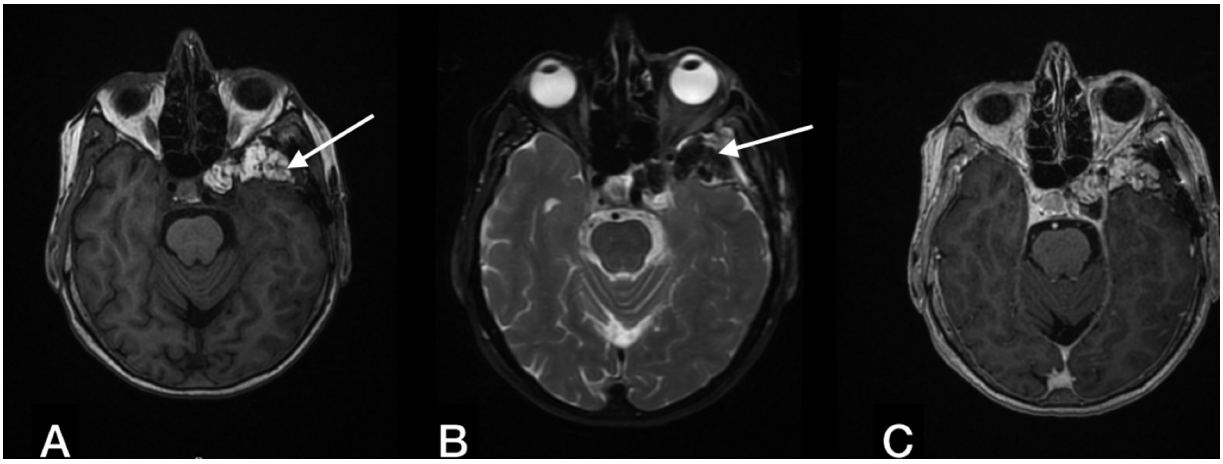

Fig. 4 2nd day postoperative axial T1-weighted (**A**), T2-weighted (**B**), and T1-weighted with contrast enhancement (**C**) MRIs show gross total resection of the tumor. Note the T1 hyperintense and T2 hypointense signal (*arrows*) of the fat graft that was placed at the end of the resection.

conservative management for 6 months, and surgery would enable histopathological diagnosis.

The patient underwent a left pterional craniotomy with an extradural transcavernous approach for surgical exploration and possible resection of this mass lesion (►**Video 1**).

Video 1

Two-dimensional video presentation illustrating the surgical removal of a neurenteric cyst in the cavernous sinus using an extradural transcavernous method. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1772157.

Histopathology revealed an NEC lined with benign respiratory-type epithelium (bronchogenic; ►**Fig. 3**). The patient made an uneventful recovery, remained neurologically intact, and had a complete resolution of her facial pain. Postoperative MRI showed a gross total resection of the mass lesion (►**Fig. 4**). At a 15-month follow-up, the patient continued to do well with no headache or facial pain, with MRI showing no recurrence (►**Fig. 5**).

Discussion

Supratentorial NECs are rare with prior reported cases in the frontal lobe,[11] optic nerve,[10] Meckel's cave,[3] and the suprasellar cistern.[12] We are aware of only one known reported case in the cavernous sinus, which was part of a case series of NECs in the CNS.[13] This report described a 2-year-old female who presented with acute third nerve palsy and was operated on with partial resection, and then reoperated following a recurrence after 3-year follow-up. To the best of our knowledge, the case we describe herein is the first NEC case of the cavernous sinus in an adult with documented histopathology, operative data, and a surgical video.

Histologically, NECs are benign cysts and not neoplastic. They have three histological patterns based on the structure of the wall: Group A is covered by cuboidal or columnar epithelium with or without cilia, such as respiratory or

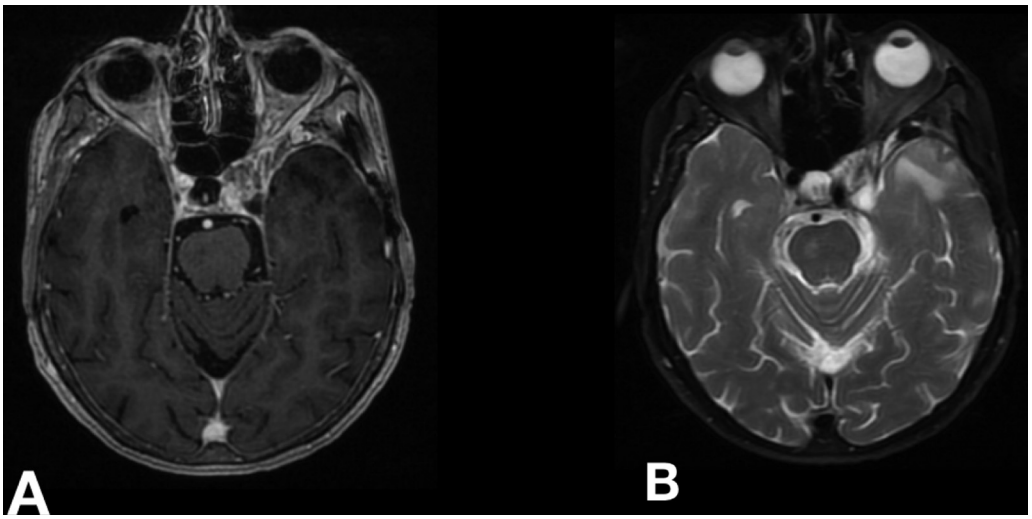

Fig. 5 Postoperative MRI brain after 15 months. Axial T1 weighted with contrast (**A**) and axial T2 weighted images (**B**) show no recurrence.

gastrointestinal epithelium; Group B, in addition to group A features, has smooth muscle in the wall, and Group C has ependymal or glial tissue in addition to the elements of group B.[14] The present case was a Group A with characteristic bronchogenic epithelium type.

NECs are endodermal in origin, with a few different theories being given regarding their embryologic origin. A commonly accepted theory is that endodermal cells migrated through the neuroenteric canal into the ectoderm to form the NEC. Because they can travel freely, NECs develop anywhere in the neuroaxis including the supratentorial compartment.[15] In the present case, the NEC was located in the cavernous sinus.

Although this is a malformation from birth, symptoms may not develop until late in life, such as with the present 75-year-old patient. The symptoms of NECs depend on the location and size of the cyst compressing surrounding structures. In the present case, neural compression of the maxillary and mandibular division of the trigeminal nerve is likely to have been the cause of the facial pain. Further, although the distal petrosal and cavernous segment of the ICA was encased by the cyst, there was no distal compromise of blood flow or related ischemic changes apart from those related to age. In the previously reported case of a cavernous sinus NEC, the patient developed symptoms at just 2 year of age in the form of acute onset unilateral ptosis.[13]

Preoperative diagnosis can be challenging since there are no specific characteristic findings either on CT scans or MRI sequences. With CT scans, NECs range from hypodense, isodense, to hyperdense, with reported marginal calcification, and may be associated with underlying bony defects.[16] The present case exhibited hyperdensity with minute wall calcifications and widening of the foramen ovale and the superior orbital fissure. MRI is similar, showing no specific characteristics with an appearance that largely depends on the protein content of the cyst, appearing isointense or slightly hyperintense in T1, while appearing isointense to hyperintense or even hypointense in T2-weighted MRI images,[17,18] with either mild or no restricted diffusion[16] and slight marginal enhancement in some cases.[16,18] In the present case, T1 MRI imaging was hypointense, T2 was hyperintense, and there was no restricted diffusion with mild marginal enhancement. These MRI findings share some characteristics with other intracranial cystic lesions, including epidermoid, dermoid, arachnoid, and other endodermal cysts (Rathke's cleft and colloid) and with cystic schwannomas. Arachnoid cysts follow the CSF intensity in all pulse sequences, while epidermoid cysts exhibit a strikingly restricted diffusion pattern.

Since these cysts are benign in nature, surgical excision should be considered the first-line treatment in symptomatic patients,[19] with the surgical approach chosen to provide the best exposure for tumor resection. In the present case, the pterional transcavernous approach provided good exposure allowing gross total resection with no compromise of neurovascular structures.

Despite being benign, due to uncertainty regarding the rate of growth, every effort should be taken for complete excision without jeopardizing critical neurovascular structures, since recurrence or even malignant transformation has been reported in a few cases.[13,20,21] The duration after which malignant transformation has been reported varies, with reports of 3.5 years[22] and 8 years after complete excision after the first operation.[23]

Although gross total resection should be the aim, this may be especially difficult with relatively large lesions, since there is a correlation between the cyst size, wall thickness, and the degree of adhesion to the adjacent neurovascular structures.[24] In the present case, the relatively large (3.0 cm) cyst content was evacuated first. The cyst wall was then peeled away from the surrounding structures including excision from the lateral wall of Meckel's cave since the cyst was adherent at that point, which resulted in egress of CSF. This was then followed by packing the resection cavity with the abdominal fat graft in strips. A postoperative lumbar drain was then maintained for a couple of days.

Conclusion

Intracranial NEC are rare entities that present a preoperative diagnostic challenge. NEC can occur along the neuroaxis from the cranium to the sacrum, so this should be considered as a differential diagnosis for cavernous sinus cystic lesions. The aim of surgery should be safe complete excision.

Previous Presentations

This manuscript has neither been presented nor published before, either as a whole or in part, nor is it under consideration for publication elsewhere.

Conflict of Interest

None declared.

Acknowledgments

The authors would like to thank the contributions of Steven L. Goodman, PhD, for editing this work.

References

1 Chaynes P, Thorn-Kany M, Sol JC, Arrué P, Lagarrigue J, Manelfe C. Imaging in neurenteric cysts of the posterior cranial fossa. Neuroradiology 1998;40(06):374–376
2 Puusepp M. Variété rare de tératome sousdural de la region cervicale (intestinome): quadriplégie, extirpation, guérison complete. Rev Neurol (Paris) 1934;2:879–886
3 Jhawar SS, Nadkarni T, Goel A. Meckel's cave neurenteric cyst: case report. J Neurol Surg A Cent Eur Neurosurg 2012;73(06): 413–415
4 Dokumcu Z, Uygun O, Turhan T, Yalaz M, Ozcan C, Erdener A. Two-stage approach in the management of thoracic neuroenteric cyst with spinal extension: thoracoscopic excision following dorsal laminectomy. Childs Nerv Syst 2015;31(02):185–189
5 El Ahmadieh TY, Sillero R, Kafka B, Aoun SG, Price AV. Isolated dorsal thoracic neuroenteric cyst with spinal cord compression: case reports in pediatrics. World Neurosurg 2018;118:296–300
6 Can A, Dos Santos Rubio EJ, Jasperse B, Verdijk RM, Harhangi BS. Spinal neurenteric cyst in association with Klippel-Feil syndrome: case report and literature review. World Neurosurg 2015;84(02):592.e9–592.e14
7 Roder C, Ebner FH, Schuhmann MU. Neurenteric cysts of the cerebellopontine angle. J Neurol Surg A Cent Eur Neurosurg 2013; 74(Suppl 1):e36–e40
8 Ranguis SC, Chaganti JR, Winder MJ. A bilateral infratentorial neurenteric cyst. J Clin Neurosci 2013;20(05):735–738
9 Tan GS, Hortobágyi T, Al-Sarraj S, Connor SE. Intracranial laterally based supratentorial neurenteric cyst. Br J Radiol 2004;77(923): 963–965
10 Scaravilli F, Lidov H, Spalton DJ, Symon L. Neuroenteric cyst of the optic nerve: case report with immunohistochemical study. J Neurol Neurosurg Psychiatry 1992;55(12):1197–1199
11 Kachur E, Ang L-C, Megyesi JF. Intraparenchymal supratentorial neurenteric cyst. Can J Neurol Sci 2004;31(03):412–416
12 Kim JH, Wang KC, Phi JH, Park SH, Cheon JE, Kim SK. Intracranial neurenteric cyst arising at the suprasellar cistern with extension to middle cranial fossa. Childs Nerv Syst 2018;34(12):2491–2495
13 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):512–523
14 Lantos P, VandenBerg S, Kleihues P. Tumors of the nervous system. InGraham D, Lantos P, eds., Greenfield's Neuropathology. London, Arnold2002
15 Mittal S, Petrecca K, Sabbagh AJ, et al. Supratentorial neurenteric cysts-A fascinating entity of uncertain embryopathogenesis. Clin Neurol Neurosurg 2010;112(02):89–97
16 Preece MT, Osborn AG, Chin SS, Smirniotopoulos JG. Intracranial neurenteric cysts: imaging and pathology spectrum. AJNR Am J Neuroradiol 2006;27(06):1211–1216
17 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(03): 735–746
18 Shakudo M, Inoue Y, Ohata K, Tanaka S. Neurenteric cyst with alteration of signal intensity on follow-up MR images. AJNR Am J Neuroradiol 2001;22(03):496–498
19 Patel SK, Liu JK. Staged bilateral far-lateral approach for bilateral cervicomedullary junction neurenteric cysts in a 10-year-old girl. J Neurosurg Pediatr 2013;12(03):274–280
20 Yang Y, Fang J, Li DA, Wang L, Ji N, Zhang J. Recurrent intracranial neurenteric cyst with malignant transformation: a case report and literature review. Oncol Lett 2016;11(05):3395–3402
21 Wang W, Piao YS, Gui QP, Zhang XH, Lu DH. Cerebellopontine angle neurenteric cyst with focal malignant features. Neuropathology 2009;29(01):91–95
22 Sahara Y, Nagasaka T, Takayasu M, Takagi T, Hata N, Yoshida J. Recurrence of a neurenteric cyst with malignant transformation in the foramen magnum after total resection. Case report. J Neurosurg 2001;95(02):341–345
23 Rim HT, Song JH, Kim ES, Kwon MJ. Mucinous adenocarcinoma arising from a residual supratentorial neurenteric cyst and expressing mutated KRAS: a case report. Hum Pathol 2016; 58:146–151
24 Wang L, Zhang J, Wu Z, et al. Diagnosis and management of adult intracranial neurenteric cysts. Neurosurgery 2011;68(01):44–52, discussion 52

 © 2023. The Author(s).