



Case Report

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Intracranial Neuroenteric Cyst in the Cerebellopontine Angle with Calcified Cyst Wall: A Rare Case Report

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ABSTRACT

Neuroenteric cysts (NCs) are rare benign lesions believed to originate from ectopic endodermal tissue. They are lined by gastrointestinal or respiratory epithelium capable of secreting mucin. Although most frequently located within the spinal canal, intracranial NCs are exceedingly rare and typically occur ventral to the brainstem. We report a case of a young female patient with an NC located in the cerebellopontine angle—an atypical site—exhibiting uncommon imaging features. Radiological evaluation revealed cystic contents with signal intensities similar to cerebrospinal fluid, a partially calcified cyst wall, and mild contrast enhancement. Extensive calcification resulted in firm adhesion to adjacent neurovascular structures. Surgical resection was performed, preserving a small calcified plaque that was tightly adherent to critical surrounding tissues. This case underscores the importance of including NCs in the differential diagnosis of intracranial cystic lesions, particularly when cyst wall calcification is present. Calcification can significantly influence surgical planning and increase the risk of adhesion to neurovascular structures, necessitating cautious dissection.

KEYWORDS: Neuroenteric Cyst, Calcification, Cerebellopontine Angle, Neuroimaging

ABBREVIATIONS: CEA: Carcinoembryonic antigen, CK: Cytokeratin, CSF: Cerebrospinal fluid, CT: Computed tomography, DWI: Diffusion-weighted imaging, EMA: Epithelial membrane antigen, GFAP: Glial fibrillary acidic protein, H&E: Hematoxylin and eosin, MUC5AC: Mucin 5AC, MRI: Magnetic resonance imaging, NC(s): Neuroenteric cyst(s), NSE: Neuron-specific enolase, PAS: Periodic acid-Schiff, T1/T1WI: T1-weighted imaging, T2/T2WI: T2-weighted imaging, T2-FLAIR: T2-weighted fluid-attenuated inversion recovery

INTRODUCTION

Since the central nervous system originates from the ectoderm (3), most congenital cysts within it are ectodermal in origin, such as arachnoid and epidermoid cysts. In contrast, cysts of endodermal origin are relatively rare and are histopathologically characterized by the presence of respiratory or gastrointestinal-type epithelium, hence termed neuroenteric cysts (NCs). These cysts most commonly occur in the caudal cervical or rostral thoracic regions of the spinal cord, whereas intracranial occurrences are exceedingly rare (8), accounting for approximately 10%–17.9% of all NCs (10).

However, due to the variability in the magnetic resonance imaging (MRI) signal of NCs—which depends on the protein content of the cyst fluid—and the lack of specificity compared to other intracranial cystic lesions, achieving a definitive preoperative diagnosis is challenging. Microsurgical resection remains the preferred treatment for symptomatic intracranial cystic lesions. Nevertheless, because of their embryological origin, intracranial NCs often form dense adhesions to critical neurovascular structures such as the brainstem, basilar artery, and cranial nerves (7,13). This report presents a case of an NC diagnosed and surgically treated at our institution. The lesion was located in the right cerebellopontine angle. Preop-

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erative computed tomography (CT) revealed partial calcification of the cyst wall—an extremely rare finding in intracranial NCs (11). Moreover, the proximity of the lesion to the lateral aperture of the fourth ventricle initially led to a provisional misdiagnosis as an ependymal cyst.

CASE REPORT

A 30-year-old female presented to our department with a 5-month history of intermittent dizziness and headaches, occasionally progressing to severe vertigo. As her symptoms gradually worsened, she sought medical attention at our hospital.

Upon admission, cranial CT revealed an ovoid cystic lesion in the right cerebellopontine angle containing low-density material (CT attenuation slightly higher than cerebrospinal fluid [CSF]: mean lesion value = 40 HU; ambient cistern = 20 HU) (Figure 1). The cyst had well-defined borders, and partial calcification of the cyst wall was observed (mean CT value of calcification = 300 HU). Subsequent MRI demonstrated that the lesion exhibited low signal intensity on T1-weighted imaging and high signal intensity on T2-weighted imaging. Diffusion-weighted imaging (DWI) revealed a low signal. The lesion measured approximately 49.9 × 21.1 mm and exerted posterolateral compression on the right medulla and cerebellum, likely accounting for the patient's symptoms. Post-contrast MRI demonstrated mild enhancement of the cyst wall without enhancement of the cyst contents (Figure 2).

A retrosigmoid approach was selected for craniotomy. Gentle retraction of the right cerebellar hemisphere exposed a white cystic lesion in the right cerebellopontine angle (Figure 3A). The cyst wall appeared firm and moderately thick. Aspiration of the cyst contents yielded pale yellow fluid (Figure 3B). Significant adhesions were encountered between the cyst wall and adjacent structures during dissection (Figure 3C). Notably, dense adhesions were observed between the calcified plaque located at the root of the lower cranial nerves and the facial and vestibulocochlear nerves, as well as with an inferiorly positioned vessel. To avoid uncontrollable bleeding and minimize the risk of postoperative cranial nerve deficits, a small portion of the calcified plaque was left *in situ*. In contrast, the remaining cyst wall and calcified regions were completely excised (Figure 3D). All critical neurovascular structures were preserved. Following subtotal cyst resection, the choroid plexus of the lateral recess of the fourth ventricle was visualized intraoperatively (Figure 3E).

Microscopic examination revealed the following: (1) cyst fluid containing erythrocytes, sparse leukocytes, and epithelial-like cell clusters with degenerative features; and (2) a cyst wall partially lined by simple to pseudostratified cuboidal epithelium overlying collagenized fibrous tissue with calcifications (Figure 4A,B). Immunohistochemical analysis confirmed endodermal differentiation. Periodic acid–Schiff (PAS) staining and Mucin 5AC (MUC5AC) positivity identified mucin-rich goblet cells (Figure 4C, D). Diffuse immunoreactivity for cytokeratin (CK), vimentin, carcinoembryonic antigen (CEA; luminal membrane), and epithelial membrane antigen (EMA; epithelial) was observed (Figure 4E,F), while glial fibrillary acidic

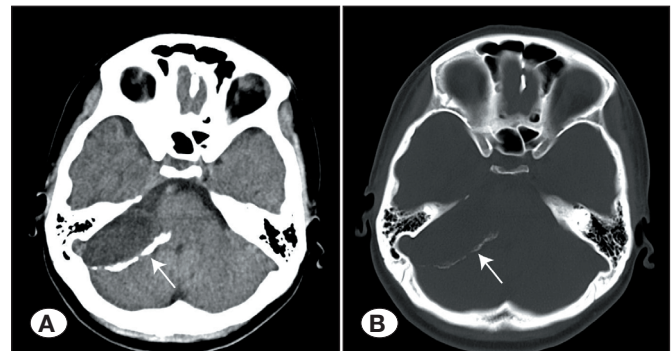


Figure 1: Axial cranial computed tomography at diagnosis. The white arrows indicate the focal calcification within the cyst wall. **A)** Brain window: A right cerebellopontine angle cystic lesion with fluid density slightly higher than cerebrospinal fluid. **B)** Bone window: Focal calcification along the posterolateral cyst wall.

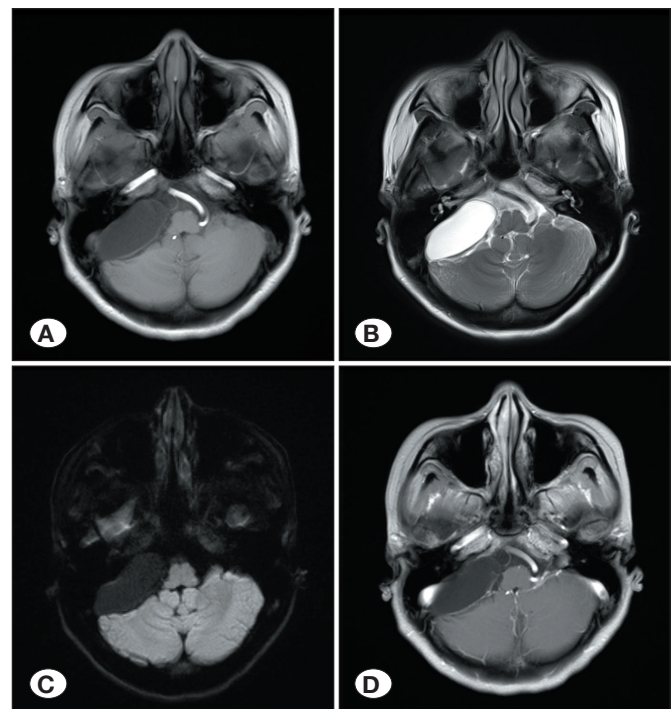


Figure 2: Preoperative brain magnetic resonance imaging. **A)** Axial T1-weighted image: Well-circumscribed cystic lesion with homogeneous fluid signal isointense to cerebrospinal fluid in the right cerebellopontine angle. **B)** Axial T2-weighted image: Hyperintense cyst contents with an intact, thin wall. **C)** Diffusion-weighted imaging: Absence of restricted diffusion. **D)** Axial gadolinium-enhanced T1-weighted image: Mild linear enhancement of the cyst wall.

protein (GFAP) and neuron-specific enolase (NSE) were negative (Figure 4G,H). The Ki-67 proliferation index was <1%. These findings supported a diagnosis of a neuroenteric cyst.

Postoperatively, imaging studies (Figure 5) were conducted. The patient recovered well, with complete resolution of previous symptoms and no emergence of new neurological deficits.

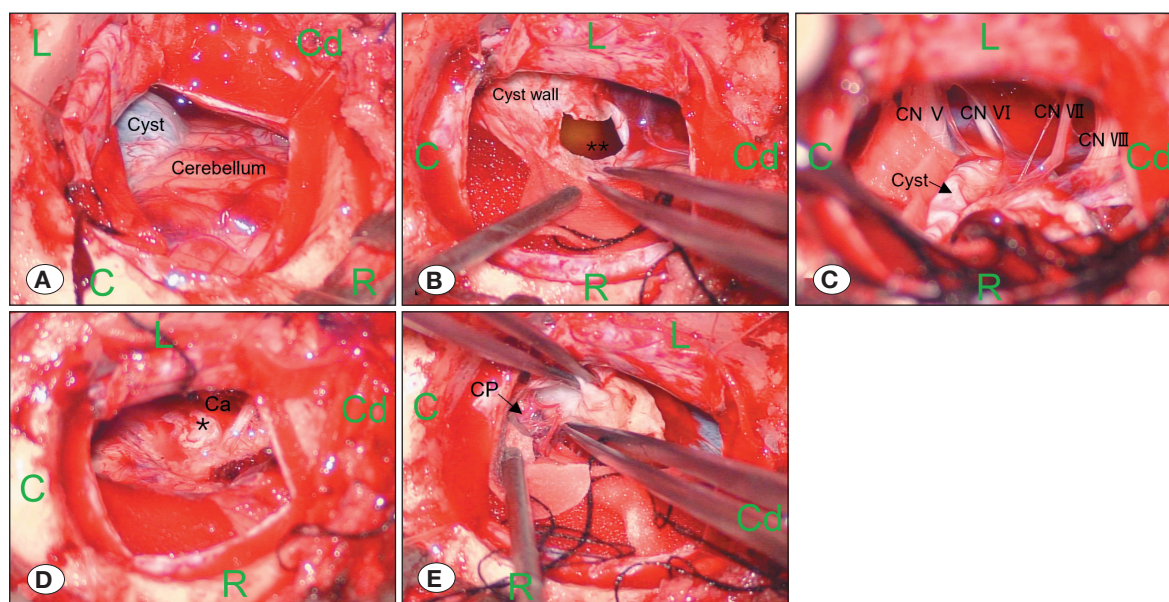


Figure 3: Intraoperative microscopic findings. Green labels (“R” denotes right, “L” left, “C” cranial, and “Cd” caudal) indicate operative orientation. **A)** Retraction of the cerebellum exposes a firm, white cyst wall in the right cerebellopontine angle. **B)** Aspiration of pale yellow cyst fluid (**) following puncture of the cyst wall. **C)** Tight adhesions between the cyst wall and surrounding cranial nerves. **D)** Residual calcified plaque (*) adherent to the facial and vestibulocochlear nerve roots. **E)** Choroid plexus-like tissue (CP) is located laterally near the lateral aperture of the fourth ventricle.

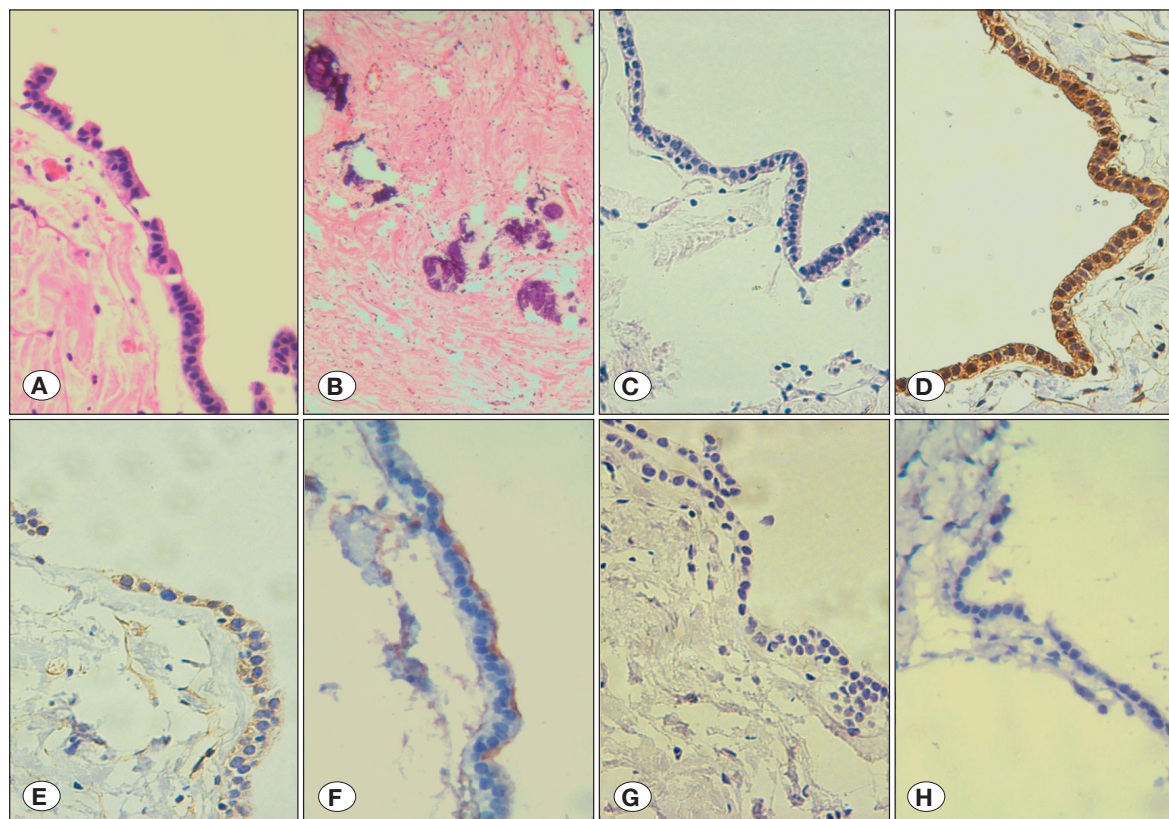


Figure 4: Histopathological and immunohistochemical characterization. **A)** Hematoxylin and eosin (H&E): Simple to pseudostratified cuboidal epithelium overlying collagenized stroma. **B)** H&E: Calcific deposits within the cyst wall stroma. **C)** Periodic acid-Schiff: Goblet cells with magenta-stained mucin vacuoles. **D)** Mucin 5AC: Strong cytoplasmic positivity in goblet cells. **E)** Carcinoembryonic antigen: Luminal membrane immunoreactivity. **F)** Epithelial membrane antigen: Continuous epithelial staining. **G)** Glial fibrillary acidic protein: Immunonegative. **H)** Neuron-specific enolase: Immunonegative. (All 400x original magnification; scale bars = 50 μm).

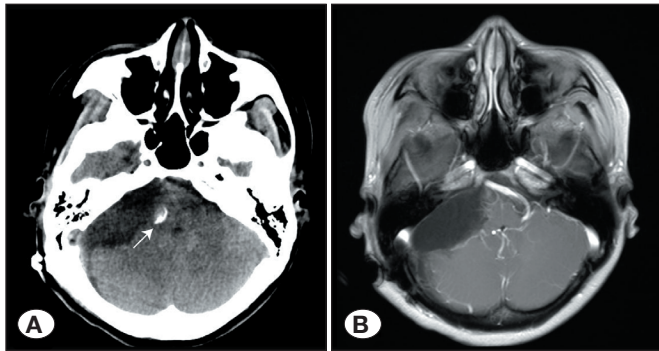


Figure 5: Postoperative imaging. **A)** Computed tomography scan on postoperative day 1 showing residual calcified tissue (white arrow). **B)** Contrast-enhanced magnetic resonance imaging on postoperative day 2 demonstrates subtotal cyst resection with preservation of vessels and nerves in the cerebellopontine angle.

DISCUSSION

Preoperative definitive diagnosis of posterior fossa NCs is exceptionally challenging, as demonstrated in this case. Patients typically present with nonspecific symptoms—such as dizziness, headache, and gait instability—resulting from mass effect on eloquent structures within the confined posterior cranial fossa. CT may reveal hypodense, isodense, or hyperdense lesions, depending on the protein concentration of the cyst fluid. MRI remains the cornerstone diagnostic modality, characteristically demonstrating well-circumscribed, round/oval cystic lesions with intact walls, reflecting their firm consistency and resistance to intraoperative rupture. The signal intensity of the cyst fluid varies considerably. While most cases exhibit isointensity on T1-weighted imaging (T1WI), hyperintensity on T2WI-fluid-attenuated inversion recovery (T2-FLAIR), and mild diffusion restriction on DWI without significant contrast enhancement, these features are critically influenced by protein content (10). Previous studies have shown that cysts with protein concentrations <10,000 mg/dL display T1 hypointensity and T2 hyperintensity; those with concentrations between 10,000 and 17,000 mg/dL show both T1 and T2 hyperintensity; and those >17,000 mg/dL present T1 hyperintensity with T2 hypointensity (5). Based on the preoperative MRI characteristics in this patient, the cyst fluid protein content was likely <10,000 mg/dL.

NCs lack specific clinical presentations and distinct MRI signal characteristics, making it challenging to differentiate them from other intracranial cystic lesions. When the cyst protein content is relatively low, the lesion typically exhibits low signal intensity on DWI, providing valuable preoperative evidence for distinguishing it from epidermoid cysts. Moreover, unlike NCs, which generally expand and displace surrounding nerves and vessels, epidermoid and dermoid cysts tend to grow along anatomical clefts, often encasing nerves and blood vessels. When distinguishing NCs from arachnoid and ependymal cysts, elevated protein content within the NC can assist preoperative differentiation on MRI. Arachnoid and ependymal cysts typically display MRI signals identical to cerebrospi-

nal fluid (CSF) and appear hypointense on DWI. In contrast, due to their proteinaceous contents, NCs often exhibit signal characteristics distinct from CSF, with hyperintensity on DWI. However, in cases such as the one presented here, where MRI sequences closely resemble CSF across all modalities, a definitive diagnosis may only be established through postoperative histopathological examination. A distinctive feature of the present case is the significant preoperative calcification of the cyst wall, which was confirmed intraoperatively as extensive, gravel-like calcification. The calcified regions increased the degree of adhesion between the cyst wall and surrounding structures, complicating complete resection. A review of the literature indicates that calcified intracranial NCs are exceedingly rare. Excluding cases (1,6,9,12,15), where calcified nodules were present within the cyst—some of which were attributed to xanthogranulomatous reactions involving melanin (14)—only one previously reported case described calcification of the cyst wall, located in the supratentorial region (11). Such cystic growths may also be associated with dystrophic calcification, and some researchers have suggested including NCs in the differential diagnosis of intramedullary calcification. The current case thus provides valuable insights into this rare pathological phenomenon.

For symptomatic NCs, complete surgical excision remains the treatment of choice. However, radical resection should not be attempted at the expense of injuring the brainstem or other critical neurovascular structures (2,13). In the present case, calcification of the cyst wall increased adhesion to adjacent tissues, rendering total excision technically more challenging. Consistent with prevailing neurosurgical practice (7,13), we believe that leaving behind a small portion of the adherent cyst wall represents an acceptable and often necessary compromise to preserve the functional integrity of vital neuroanatomical structures. Complete aspiration of the cyst contents during surgery is also essential, as spillage into the cerebral cisterns may provoke severe aseptic inflammation (13). Recurrence of intracranial NCs has been reported between 4 months and 14 years postoperatively (4). According to Chen et al., subtotal resection during initial surgery and patient age under 30 years constitute risk factors for recurrence (2). Consequently, a long-term follow-up of at least 10 years is warranted in this patient (4). Should recurrence occur, reoperation remains the primary treatment, while cystoperitoneal or ventriculoperitoneal shunting may serve as viable secondary options—both generally associated with favorable clinical outcomes (2).

CONCLUSION

Intracranial NCs are exceptionally rare but generally associated with a favorable prognosis. Although their distinct histopathological characteristics facilitate postoperative diagnosis, the clinical presentation, laboratory findings, and imaging manifestations are typically nonspecific. MRI remains the most valuable preoperative diagnostic tool. To the best of our knowledge, this is the first documented case of a neuroenteric cyst located in the cerebellopontine angle with prominent calcification of the cyst wall. This feature broadens the known imaging spectrum of NCs. Calcified cyst walls often demonstrate dense adhesions to adjacent structures; therefore, sur-

gical management should aim for maximal safe resection of the cyst wall while preserving vital neurovascular structures and functions. Even subtotal resection can result in satisfactory clinical outcomes. NCs should thus be considered in the differential diagnosis of intracranial cystic lesions to optimize surgical strategy and postoperative prognosis.

Declarations

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Informed consent: Written informed consent was obtained from the patient for the publication of any potentially identifiable data or images included in this article.

AUTHORSHIP CONTRIBUTION

Study conception and design: YL, YZ

Data collection: XZ, YM

Analysis and interpretation of results: CQ, YZ

Draft manuscript preparation: XZ, YM, JY

Critical revision of the article: XZ, YZ

Other (study supervision, fundings, materials, etc...): YL, YZ

All authors (XZ, YM, JY, CQ, YL, YZ) reviewed the results and approved the final version of the manuscript.

REFERENCES

1. Agrawal M, Dharanipathy S, Nakra T, Garg K, Gurjar H, Mishra S, Singh M, Chandra PS: Supratentorial neurenteric cyst: a rare differential for a frontal cyst. *World Neurosurgery* 129: 140-142, 2019. <https://doi.org/10.1016/j.wneu.2019.05.246>
2. Chen CT, Lee CY, Lee ST, Chang CN, Wei KC, Wu CT: Neurenteric cysts: risk factors and management of recurrence. *Acta Neurochir (Wien)* 158: 1325-1331, 2016. <https://doi.org/10.1007/s00701-016-2828-y>
3. Clevers H: Modeling Development and disease with organoids. *Cell* 165: 1586-1597, 2016. <https://doi.org/10.1016/j.cell.2016.05.082>
4. Gauden AJ, Khurana VG, Tsui AE, Kaye AH: Intracranial neuroenteric cysts: A concise review including an illustrative patient. *J Clin Neurosci* 19: 352-359, 2012. <https://doi.org/10.1016/j.jocn.2011.10.005>
5. Hayashi Y, Tachibana O, Muramatsu N, Tsuchiya H, Tada M, Arakawa Y, Suzuki M, Yamashita J: Rathke cleft cyst: MR and biomedical analysis of cyst content. *J Comput Assist Tomogr* 23: 34-38, 1999. <https://doi.org/10.1097/00004728-199901000-00008>
6. Jeon SJ, Sohn CH, Kim EH, Son KR, Park SH, Chang KH: Craniospinal neurenteric cysts: Various MR imaging features. *Investigative Magnetic Resonance Imaging* 13: 54-62, 2009. <https://doi.org/10.1007/s10143-018-01070-4>
7. Lemos SS, Sá MI, Casimiro CD, Guerreiro C, Faria C: A ventral brainstem neurenteric cyst – A case report and review of the pre-brainstem location. *Surg Neurol Int* 15: 195, 2024. https://doi.org/10.25259/SNI_111_2024
8. Pettersson SD, Ali S, Burmaka P, Fercho J, Szmuda T, Abuhaimed A, Alotaibi Y, Słoniewski P, Krakowiak M: Predictors for complete surgical resection of posterior fossa neurenteric cysts: A case report and meta-analysis. *Surg Neurol Int* 12: 530, 2021. https://doi.org/10.25259/SNI_723_2021
9. Preece MT, Osborn AG, Chin SS, Smirniotopoulos JG: Intracranial neurenteric cysts: imaging and pathology spectrum. *AJNR. AJNR Am J Neuroradiol* 27: 1211-1216, 2006. <http://www.ajnr.org/content/27/6/1211>
10. Sangrador-Deitos MV, Sánchez-Cantú TE, Navarro-García de Llano JP, Rodríguez-Hernández LA, Alcocer-Barradas V: Neurenteric cyst of the posterior cranial fossa: a case report and literature review. *Cureus* 14: e22628, 2022. <https://doi.org/10.7759/cureus.22628>
11. Tan GS, Hortobágyi T, Al-Sarraj S, Connor SEJ: Intracranial laterally based supratentorial neurenteric cyst. *Br J Radiol* 77: 963-965, 2004. <https://doi.org/10.1259/bjr/77185587>
12. Tripathi M, Savardekar A, Chhabra R, Radotra BD, Gupta SK: Unusual imaging finding of a “fluid–fluid” level in a neurenteric cyst at anterior margin of the cervico–medullary junction. *Br J Neurosurg* 29: 432-434, 2015. <https://doi.org/10.3109/02688697.2014.993594>
13. Wang L, Zhang J, Wu Z, Jia G, Zhang L, Hao S, Geng S: Diagnosis and management of adult intracranial neurenteric cysts. *Neurosurgery* 68: 44-52, 2011. <https://doi.org/10.1227/NEU.0b013e3181fc5ee0>
14. Yamamoto J, Shimajiri S, Akiba D, Nakano Y, Nishizawa S: Intracranial neurenteric cyst with an enhanced mural nodule and melanin pigmentation: radiologic-pathologic correlation. *World Neurosurg* 97: 758.e11-758.e19, 2017. <https://doi.org/10.1016/j.wneu.2016.09.126>
15. Ziu M, Vibhute P, Vecil G, Henry J: Isolated spinal neurenteric cyst presenting as intramedullary calcified cystic mass on imaging studies: case report and review of literature. *Neuroradiology* 52: 119-123, 2010. <https://doi.org/10.1007/s00234-009-0624-6>