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CASE REPORT

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# An Unusual Case of Large Posterior Fossa Neurenteric Cyst Involving Bilateral Cerebellopontine Angle Cisterns: Report of a Rare Case and Review of Literature

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## Summary

**Background:** Intracranial neurenteric cysts are rare cystic masses of endodermal origin lined with mucin producing low columnar or cuboidal epithelium. Approximately 141 cases have been reported so far. Most of the posterior fossa neurenteric cysts are typically small, located anteriorly to the brainstem in the midline or in the cerebellopontine angle cistern area.

**Case Report:** We present a rare, histologically proven case of a large lobulated intracranial neurenteric cyst measuring 4.2 centimeters in the maximal transverse dimension and involving bilateral cerebellopontine angle cisterns. We also present a review of the literature on this uncommon finding.

**Conclusions:** Imaging features of neurenteric cyst are non-specific and it should be considered in the differential diagnosis for any intracranial extraaxial cystic lesion.

**MeSH Keywords:** Cerebellopontine Angle • Magnetic Resonance Imaging • Neural Tube Defects

**Abbreviations:** **NC** – neurenteric cyst; **CPA** – cerebellopontine angle; **MRI** – Magnetic Resonance Imaging; **CSF** – cerebrospinal fluid

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## Background

Neurenteric cysts are congenital cystic lesions that develop during or shortly after the third week of gestation from notochordal dysgenesis and are of endodermal origin [1]. Most neurenteric cysts occur in the spine. Intracranial NCs are uncommon small cysts often located in the posterior fossa in midline or occasionally in cerebellopontine angle cistern [2,3]. We described a case of large lobulated NC measuring 4.2 centimetres in maximal dimension and involving bilateral CPA cisterns, We also reviewed the literature. We could not find any similar case of such a large posterior fossa NC involving bilateral cerebellopontine angle cisterns in the literature reported previously.

## Case Report

A 35-year-old man presented with a four-month history of headache and vertigo. Detailed neurological examination, chest X-ray and routine laboratory investigations were normal. Magnetic Resonance Imaging of brain was advised which showed a large lobulated non-enhancing lesion of 4.2×2.7×4.2 cm in size, located in the right cerebellopontine angle cistern extending across the midline, anteriorly to the brainstem to involve the left CPA cistern as well. The lesion showed a high signal intensity on T1-weighted (Figure 1) and FLAIR image (Figure 2) and low signal intensity on T2-weighted (Figure 3) image with no evidence of contrast enhancement (Figure 4A and 4B) and diffusion restriction (Figure 5). The option of NC versus white epidermoid was presented. The patient was subjected to surgery and the lesion was excised. Histopathologically, multiple sections showed a cyst lined with cuboidal to flattened cells and columnar cells sometimes (Figure 6). No goblet cells, appendages or keratin flakes were seen. Immunohistochemistry with Glial Fibrillary Acidic Protein (GFAP) was negative (Figure 7). These features were consistent with neurenteric cyst. Serial imaging follow-ups were suggested.

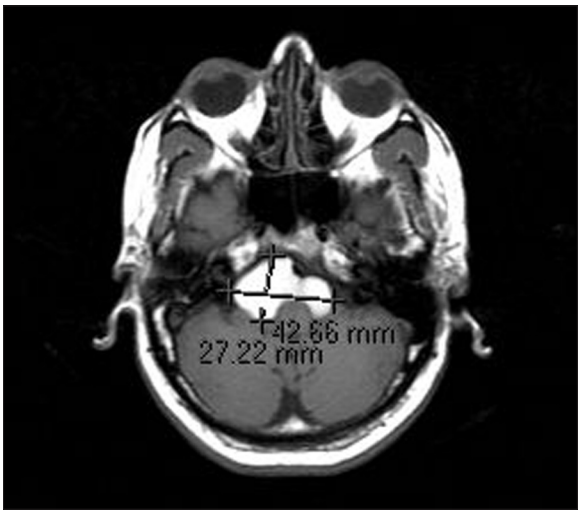


**Figure 1.** Axial plain T1-weighted image of the brain at the level of pons shows a well-defined extraaxial homogenous hyperintense lesion measuring 4.2 by 2.7 centimeters anterior to brain stem and involving bilateral cerebellopontine angle cisterns.

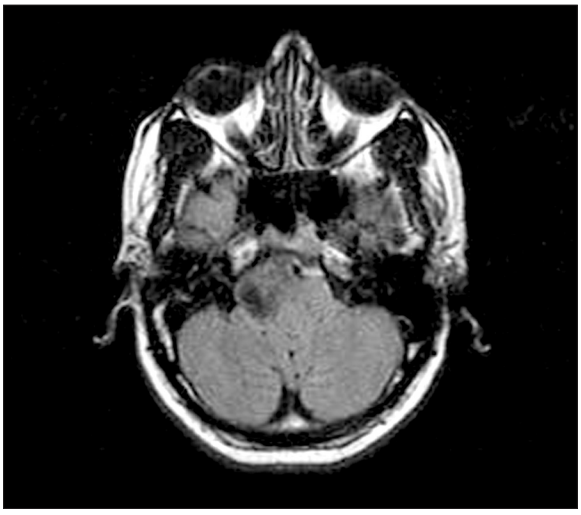

**Figure 2.** Axial FLAIR image shows hyperintense signal.

## Discussion

Neurenteric cysts are congenital lesions that arise at the time of notochordal development possibly due to nonseparation of notochord and foregut during the process of escalation leading to incorporation of primitive endodermal cells into the notochord [4]. They are also known as enterogenous cysts, enteric cysts, endodermal cysts, and archenteric cyst. Though they can occur along the entire neuraxis, they are approximately three times more common in the spine compared to the brain. Although the true incidence is still unknown, the retrospective studies carried out to date suggest that all intracranial NCs comprise approximately 0.15 to 0.35% of all intracranial neoplasms. Almost 141 cases (including our case) of intracranial neurenteric cysts have been reported in the literature [5]. Unlike spinal NCs, where coexisting vertebral anomalies are seen in almost 50 percent of the cases, intracranial cysts are never associated with bony anomalies of the clivus or skull base.

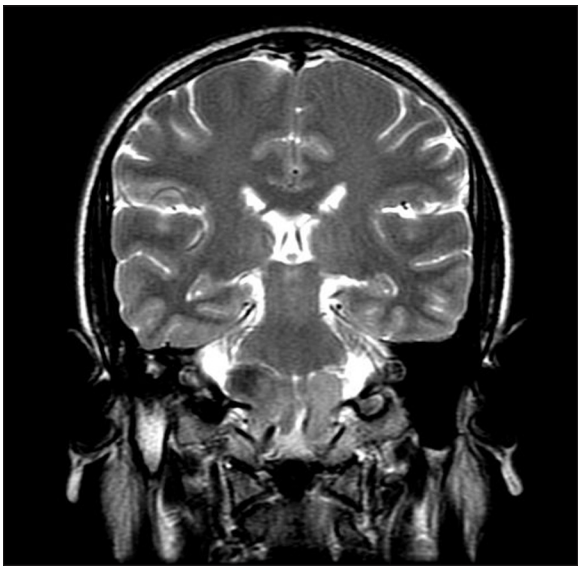

**Figure 3.** Coronal T2-weighted image of the brain shows hypointense signal of the lesion as compared to the cerebrospinal fluid.

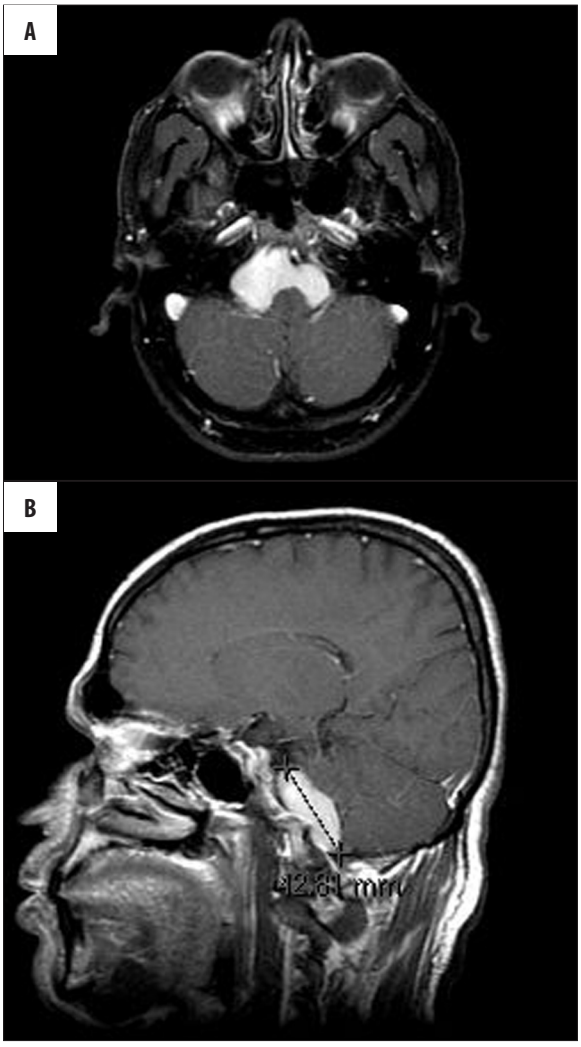


**Figure 4.** Contrast-enhanced axial (**A**), and sagittal (**B**) T1-weighted images of the brain show nonenhancement of the mass and cyst size of 4.2 centimeters in craniocaudal dimension (B).

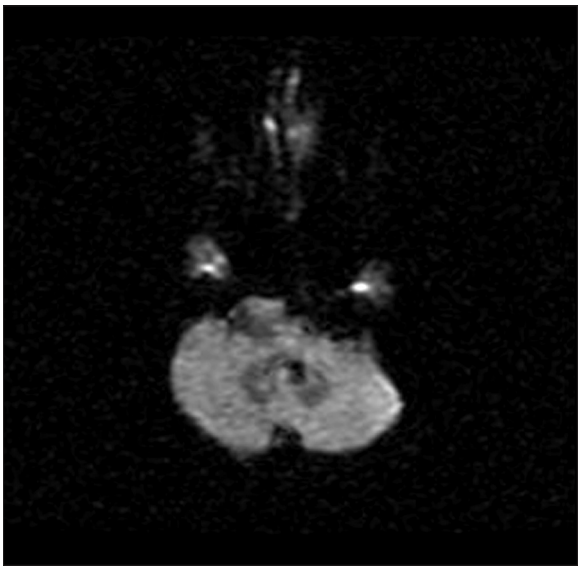

**Figure 5.** Axial diffusion image shows no evidence of intralesional diffusion restriction.

As compared to intraspinal NCs which usually occur in the pediatric age groups, intracranial NCs are more common in the adult population. The possible cause of delayed presentation in intracranial NC could be due to increased susceptibility of the spinal cord to a local mass effect. There is no significant gender preponderance.

Intracranial cysts are mostly found in the posterior fossa but occasionally supratentorial cysts have also been reported. Out of all reported cases, 61 were located in the posterior fossa, 47 in cervicomedullary junction and 19 in supratentorial compartment. The majority of the described posterior fossa cysts (63%) were located in the midline and 29% were lateral, with no details on cyst location in 11 case reports. Bilateral location as in our case has not been reported to date. Supratentorial cysts can be seen in suprasellar & parasellar regions, along the optic nerve or the oculomotor nerve and in the superior orbital fissure [5].

Most NCs in the posterior fossa are small and measure less than 2 centimeters in diameter. Preece et al. described 13 cases of NE cysts in the posterior fossa with size ranging from 1.0 centimeter to 3.4 centimeters [3]. Priamo et al. reported on a case of posterior fossa cyst which had grown to 3.9 centimeters within 1.5 years [6]. No similar case of such a large neurenteric cyst involving bilateral cerebellopontine angle cisterns has been found to our knowledge.

The most common presenting features in posterior fossa NCs are vertigo or imbalance, hearing loss, tinnitus, cranial nerve deficits and trigeminal neuralgia. In contrast, supratentorial NCs usually present with symptoms of raised intracranial pressure, like headache, nausea, and vomiting [5].

On Magnetic Resonance Imaging most NCs are isointense or hyperintense compared to CSF in both T1- and T2-weighted images, as the majority of them contain proteinaceous contents. Occasionally, they show homogenous,very bright

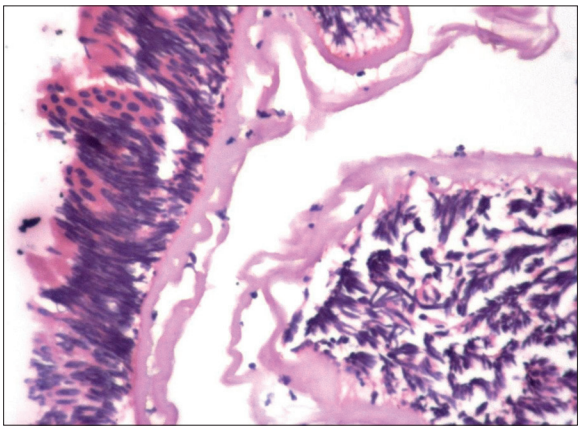

**Figure 6.** Photomicrographs show cyst lining composed of cuboidal cells.

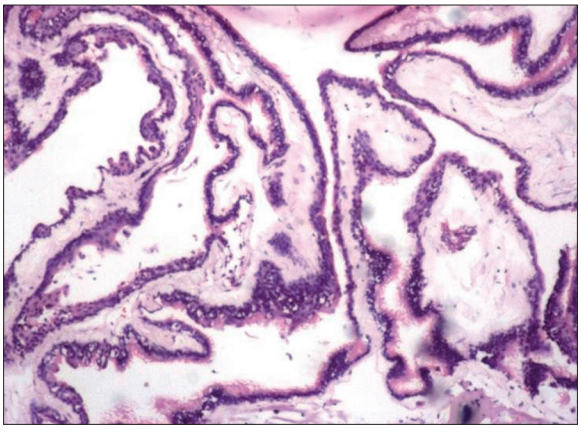

**Figure 7.** Immunohistochemistry shows negative staining with Glial Fibrillary Acidic Protein.

signal intensity on T1-weighted images and low signal intensity on T2-weighted images as in our case, put due to very high protein content in the cyst. Many authors described the relationship between protein concentration of cyst and signal intensity on MR images previously [1]. These cysts are hyperintense on FLAIR and show no or mild diffusion restriction. NCs are nonenhancing but rare cases of rim enhancement and sometimes solid enhancement (when accompanied by marked xanthogranulomatous changes) have been reported on [3,7,8].

The differential diagnoses for intracranial NC include epidermoid cyst, dermoid cyst, arachnoid cyst and other endodermal cysts. Though white epidermoid shows similar features to these cysts on all sequences, unlike NC it shows marked diffusion restriction. However, rare cases of histologically proven NCs have shown diffusion restriction on imaging. Dermoid cysts are quite heterogeneous in contents and most cases have intralesional fat (seen as suppression of signal on fat suppressed sequences). They also show marked diffusion restriction. Arachnoid cysts show cerebrospinal fluid-like signal in all sequences [3,5].

The treatment of choice for intracranial NC is complete surgical removal. Follow-up serial imaging is advisable in these patients because of potential complications like recurrence, dissemination and malignant transformation [6].

## Conclusions

Intracranial NCs are usually small, nonenhancing cystic lesions located in the midline in the posterior fossa with isointense/hyperintense signal in both T1- and T2-weighted images.

However, occasionally they can show unusual features like large size, involvement of both CPA cisterns, hypointense signal on T2-weighted images (as in our case). Therefore, NC should be considered in the differential diagnosis for any intracranial extraaxial cystic lesion.

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