
CASE REPORT

Posterior Cranial Fossa Neurenteric Cyst with Malignant Transformation

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ABSTRACT

Neurenteric cyst is a notably rare epithelium-lined cystic lesion found in the central nervous system. Malignant transformation of the neurenteric cyst is infrequent, with only six cases reported in the English literature. We report the case of a 74-year-old patient with recurrent neurenteric cyst in the posterior cranial fossa that underwent malignant transformation 2 years after initial surgery. Histologically, the initial lesion was compatible with benign neurenteric cyst, whereas the recurrent lesion illustrated features of adenocarcinoma.

Key Words: Central nervous system diseases; Cranial fossa, posterior; Cysts; Neural tube defects

中文摘要

後顱窩神經管原腸性囊腫的惡變

蔣碧茜、羅尚銘、何聯昌、陳智明、潘偉麟、鄧國穎、張毓靈

神經管原腸性囊腫是中樞神經系統一種特別罕見的上皮層囊性病變。神經管原腸性囊腫的惡變很少見，英文文獻中只報導了六個病例。本文報告一名74歲病人繼兩年前因神經管原腸性囊腫進行首次手術後，在後顱窩有惡性復發。組織學上，早期病變與良性神經管原腸性囊腫一致，但復發病灶有腺癌的特徵。

INTRODUCTION

Neurenteric cyst is a congenital, benign, epithelium-lined cyst of the central nervous system formed during fetal notochord development. It may occur in any location of the central nervous system, but is more commonly seen in the spine than in the brain.^{1,2} When located intracranially, it is more commonly seen in the posterior cranial fossa, characteristically in the midline anterior to the brainstem.² Malignant change of a

neurenteric cyst is extremely rare, with only six cases reported in the English literature.³⁻⁸ We report a case of neurenteric cyst located in the posterior cranial fossa with subsequent malignant transformation.

CASE REPORT

A 74-year-old woman with a history of hypertension, diabetes, and ischaemic heart disease was admitted to the Queen Elizabeth Hospital with complaints

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of intermittent dizziness for 2 years in June 2010. She complained of increasingly severe dizziness and unsteady gait for a few days before admission. Computed tomography (CT) on admission showed a cystic lesion of cerebrospinal fluid (CSF) density located in the midline of posterior cranial fossa. It impinged

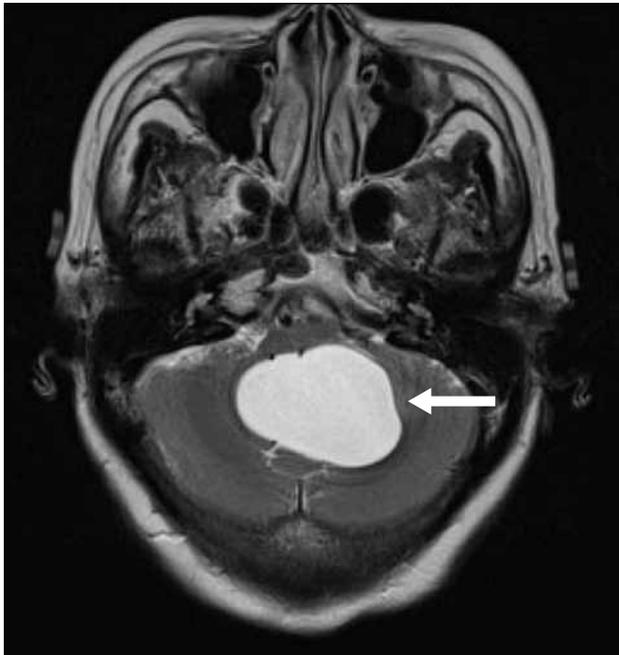


Figure 1. An axial T2-weighted magnetic resonance image shows a well-circumscribed cystic lesion (arrow) in fourth cerebral ventricle.

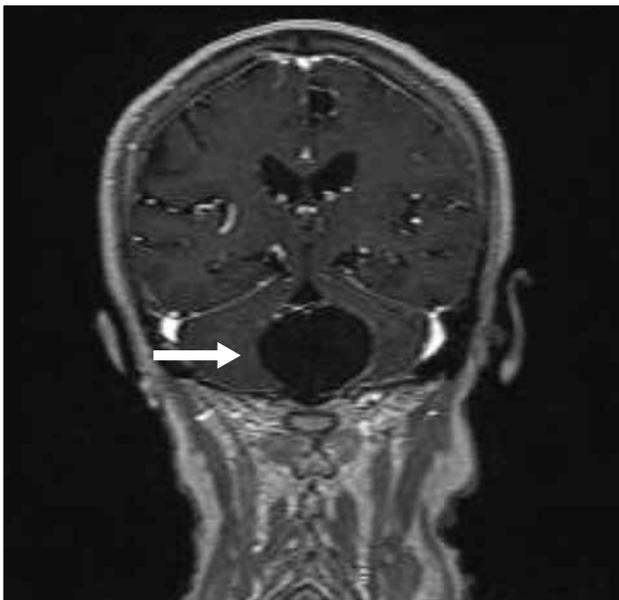


Figure 2. A coronal post-gadolinium-enhanced T1-weighted magnetic resonance image shows no definite wall or soft-tissue enhancement within the cystic lesion (arrow).

on the medulla and displaced the cerebellar vermis and cerebellar hemispheres. Magnetic resonance imaging (MRI) was performed, demonstrating a cystic lesion in the posterior cranial fossa appearing hypointense on T1-weighted images and hyperintense on T2-weighted images. The content of the cystic lesion followed CSF signal intensity. The cyst showed neither internal septa nor focal wall enhancement in post-gadolinium enhanced images (Figures 1 and 2). No restricted diffusion was noted in diffusion-weighted images.

The patient later underwent suboccipital craniotomy and marsupialisation of the posterior fossa cyst. Haematoxylin-and-eosin-stained sections of the surgical specimen showed that the cyst wall was lined by columnar to cuboidal cells, with adjacent fibrous stroma. Histological diagnosis was consistent with intracranial neurenteric cyst (Figure 3).

The patient subsequently complained of persistent dizziness 2 years after surgery. MRI performed at this juncture showed a newly arising, heterogeneous, enhancing solid mass in the posterior cranial fossa, situated at the previous operative site (Figures 4 to 6). Second surgery was performed and histological assessment demonstrated multiple mucin-filled locules lined by atypical epithelial cells (Figure 7). Irregular glandular formation was also seen with features of invasion. Overall histological findings were compatible with adenocarcinoma arising from previously known neurenteric cyst.

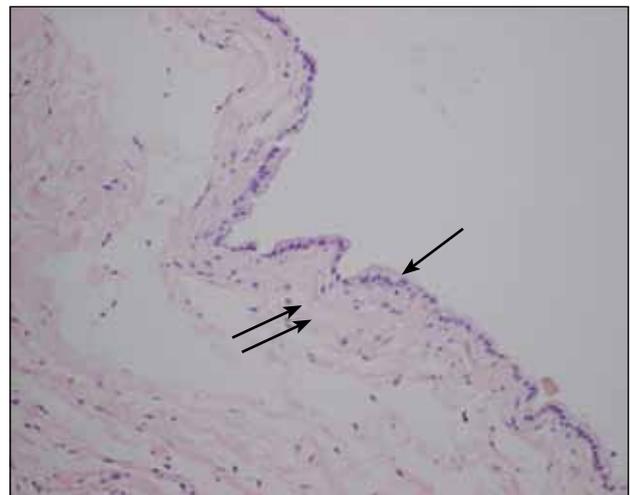


Figure 3. The cyst wall is lined by cuboidal to columnar cells (single arrow), with adjacent fibrous stroma (double arrows) [H&E; original magnification, x 200].



Figure 4. An axial post-gadolinium enhanced T1-weighted magnetic resonance image shows a solid-enhancing lesion (arrow) in the left fourth cerebral ventricle.

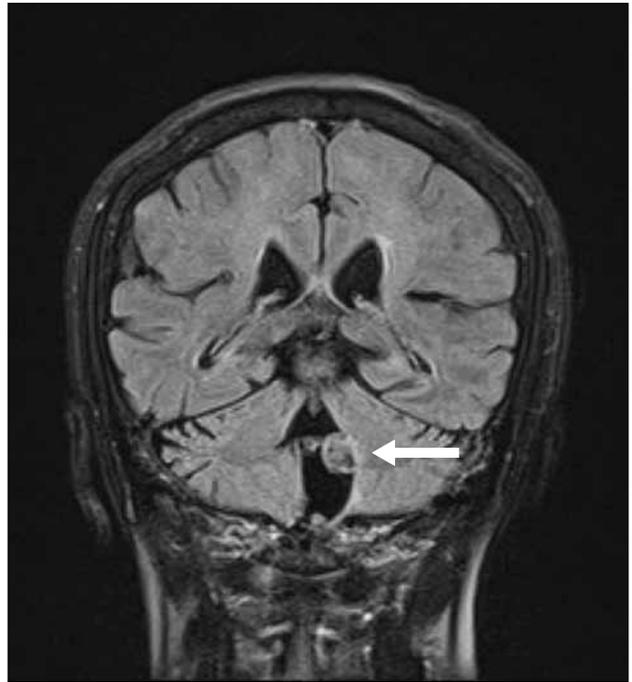


Figure 6. A coronal FLAIR (fluid-attenuated inversion recovery) image shows a solid lesion (arrow) in the location of resected neurenteric cyst.

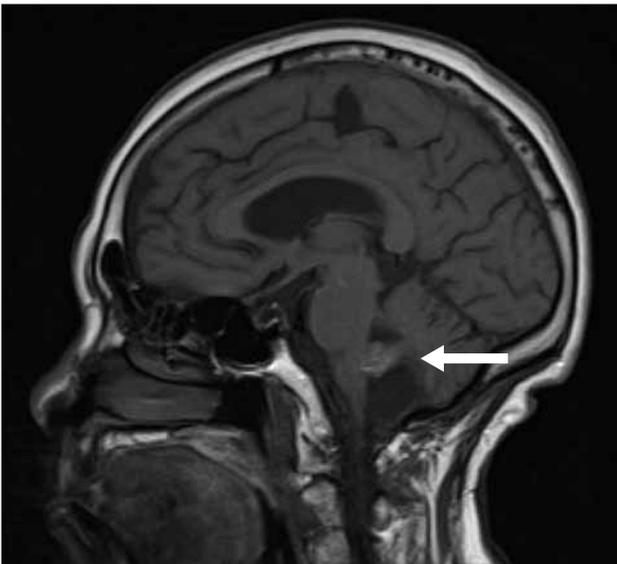


Figure 5. A sagittal post-gadolinium enhanced T1-weighted magnetic resonance image shows the location of the solid mass (arrow) within the fourth cerebral ventricle.

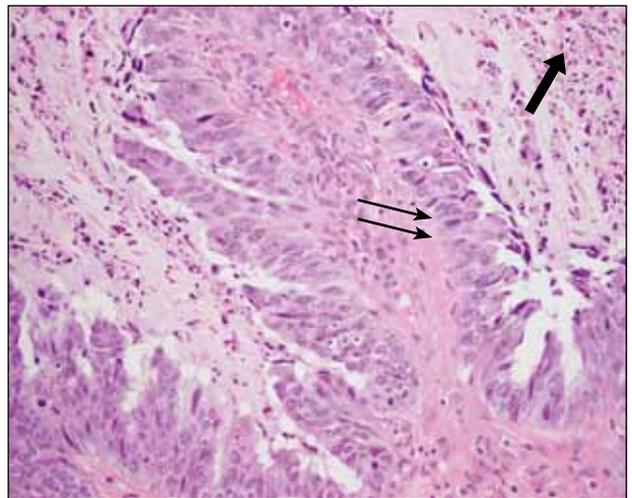


Figure 7. Specimen section after second surgery shows multiple mucin-filled locules (single arrow) lined by atypical epithelial cells with irregular glandular formation (double arrows). Histological findings are compatible with adenocarcinoma arising from neurenteric cyst (H&E; original magnification, x 400).

The patient has been well after surgery with no evidence of tumour recurrence or metastasis.

DISCUSSION

Neurenteric cyst is a rare cyst arising from displaced

elements of the alimentary canal. It is most commonly seen in the posterior mediastinal region, but may also occur in any location within the central nervous system.¹ Within the nervous system, it is more commonly seen in the spine (accounting for 0.3% to 0.5% of spinal

tumours).⁹ Intracranial neurenteric cysts account for 10% to 17.9% of all neurenteric cysts,¹⁰ most commonly located in the posterior cranial fossa. The cyst is classically located in the midline anterior to the brainstem,² but has been reported in the cerebellopontine angle or along the clivus. Supratentorial neurenteric cysts are very rarely seen.

The most common clinical presentation is related to the mass effect of the neurenteric cyst. Posterior fossa cyst may, particularly in relation to its location, present with vertigo, imbalance, tinnitus, or hearing loss. It is almost always benign, although malignant change has been previously reported.²⁻⁹

Most of the neurenteric cysts are smooth, thin-walled lesions containing proteinaceous contents. Microscopic examination of the cyst wall demonstrates endothelium-lined structures of columnar to cuboidal cells. Imaging findings of a neurenteric cyst are characteristically hypodense on CT scan. On MRI, it is roundish or slightly lobulated in appearance, and shows no enhancement in contrast images. It typically follows CSF signals in both T1-weighted and T2-weighted images. It is hyperintense on fluid-attenuated inversion recovery image and may show mild or no restricted diffusion on diffusion-weighted images.

Differential diagnoses of neurenteric cyst include arachnoid cyst, epidermoid cyst, colloid cysts, Rathke's cleft cysts, and other cystic tumours. Although many of these lesions have similar imaging findings, they may be differentiated by location. Epidermoid cyst is more commonly situated off-midline in the posterior cranial fossa; Rathke's cleft cyst is typically sellar in location, and colloid cyst is typically anterior to the third ventricle.^{1,2}

In the present case, the initial clinical, radiological, and histological features are consistent with a typical benign neurenteric cyst. Approximately 2 years after initial surgery, however, a recurrent solid-enhancing tumour occurred at the same site. There was no evidence of systemic malignancy, and histology slides showed benign neurenteric cyst wall adjacent to neoplastic cells, compatible with a case of malignant transformation rather than brain metastasis.

As far as the authors know, there have been six reported cases of malignant change in an intracranial neurenteric cyst in the English literature.³⁻⁸ These six cases were

equally distributed in both sexes (three female and three male) and the age ranged from 26 to 58 years. Four of these patients were reported to have an infratentorial³⁻⁶ lesion while two had a supratentorial^{7,8} lesion. Four patients were identified with malignant neurenteric cyst at the first surgery,^{3,6-8} while two patients had undergone resection of benign neurenteric cyst which recurred later with malignant change (after a delay of 3.5 years and 14 years, respectively).^{4,5} Similar to our patient, most of these case reports showed complex cystic / solid lesions on MRI. However, one case demonstrated a multilobulated cystic mass with no enhancement,³ suggesting that careful histological examination of all cystic lesions should be performed.

CONCLUSION

Although rare, neurenteric cysts may occur in the central nervous system. These are typically benign but malignant change has been reported. The majority of those with malignant change demonstrate a complex solid component on imaging. However, malignant change has been reported in a multilobulated cystic lesions, stressing the need for thorough radiological and pathological examination.

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