

Porencephalic cyst as a cause of seizures in adults: a rare case report using MRI and cerebral angiography methods

Muhammad Yunus Amran, David Christian Haurissa, Gita Vita Soraya

Abstract

A porencephalic cyst is a rare intracranial lesion characterised by a cerebrospinal fluid-filled cavity within the cerebral hemisphere, typically surrounded by gliotic or spongiotic white matter. These cysts often present with neurological symptoms such as seizures, developmental delays, or motor impairments, depending on their size and location. We present the case of a 19-year-old male with a three-month history of recurrent seizures. Initial management with valproic acid led to significant improvement, with the patient remaining seizure-free during follow-up. Cerebral MRI revealed a porencephalic cyst with a mural nodule in the left temporo-occipital region; however, angiographic studies demonstrated no vascular abnormalities. Treatment approaches for porencephalic cysts are primarily symptomatic, including the use of antiepileptic medications and physiotherapy to address motor deficits. Surgical intervention may be considered in cases complicated by hydrocephalus or raised intracranial pressure. This case underscores the importance of tailored management strategies in achieving optimal outcomes.

Keywords: MRI of the head, Cerebral angiography, Porencephalic cyst, Seizure.

DOI: <https://doi.org/10.47391/JPMA.25-21308>

Introduction

Porencephalic cysts are rare intracranial cysts that infrequently occur in adults. This condition is characterised by a cavity in the cerebral hemispheres, filled with cerebrospinal fluid, with thin walls lined by gliotic or spongiotic white matter. These cysts are often directly connected to the ventricular system. Their size varies significantly, and they can be cortical or subcortical,

.....
Division of Interventional Neurology and Neuroendovascular Therapy, Department of Neurology, Faculty of Medicine, Hasanuddin University, Brain Centre, Dr. Wahidin Sudirohusodo General Hospital, and Hasanuddin University Teaching Hospital, Makassar, Indonesia.

Correspondence: Muhammad Yunus Amran.

Email: muhyunusamran@med.unhas.ac.id

ORCID ID: 0000-0001-5079-7490

Submission complete: 27-09-2024 **First Revision received:** 27-12-2024

Acceptance: 16-08-2025 **Last Revision received:** 15-08-2025

unilateral or bilateral, commonly occurring in regions supplied by the cerebral arteries. It is believed that porencephalic cysts result from a disruption of the blood supply, leading to cerebral degeneration. Congenital porencephalic cysts develop due to intrauterine vascular injury, causing cerebral ischaemia or intraparenchymal haemorrhage. Intrauterine infections, such as those caused by cytomegalovirus, may also result in the formation of congenital cysts.^{1,2} Clinical symptoms vary depending on the size and location of the cyst. Patients may be asymptomatic or present with epilepsy, focal neurological deficits, or learning difficulties.²

Case Report

A 19-year-old male was admitted to Wahidin Sudirohusodo Hospital, South Sulawesi, Indonesia, on May 24, 2023, with complaints of seizures. He reported a history of recurrent seizures beginning three months earlier. The seizure pattern involved closed eyes, stiffening of the legs and arms, absence of frothing at the mouth, and loss of consciousness, with each episode lasting less than two minutes. Prior to a seizure, the patient experienced dizziness, and following the episode, he reported headaches and drowsiness. There was no history of nausea or vomiting. The patient had experienced his first seizure five years ago and had been on regular Valproic acid, which was discontinued in 2021 after the seizures ceased spontaneously.

The patient had a history of intermittent headaches since the age of 12. There had been no treatment for the past six months. Physical and neurological examinations revealed no abnormalities, and laboratory investigations showed normal results. Magnetic resonance imaging (MRI) of the head without contrast was performed, revealing a hypointense lesion on the T1-weighted image (T1WI) that appeared hyperintense on the T2-weighted image (T2WI), with intensity suppression on fluid-attenuated inversion recovery (FLAIR), restriction on diffusion-weighted imaging (DWI), well-defined regular edges, and measuring approximately 4.67 x 3.69 x 3.96 cm. The lesion was located in the left temporo-occipital region, connected to the posterior horn of the left lateral ventricle but not to the subarachnoid space. A mural nodule with an intensity measuring approximately 2.15 x

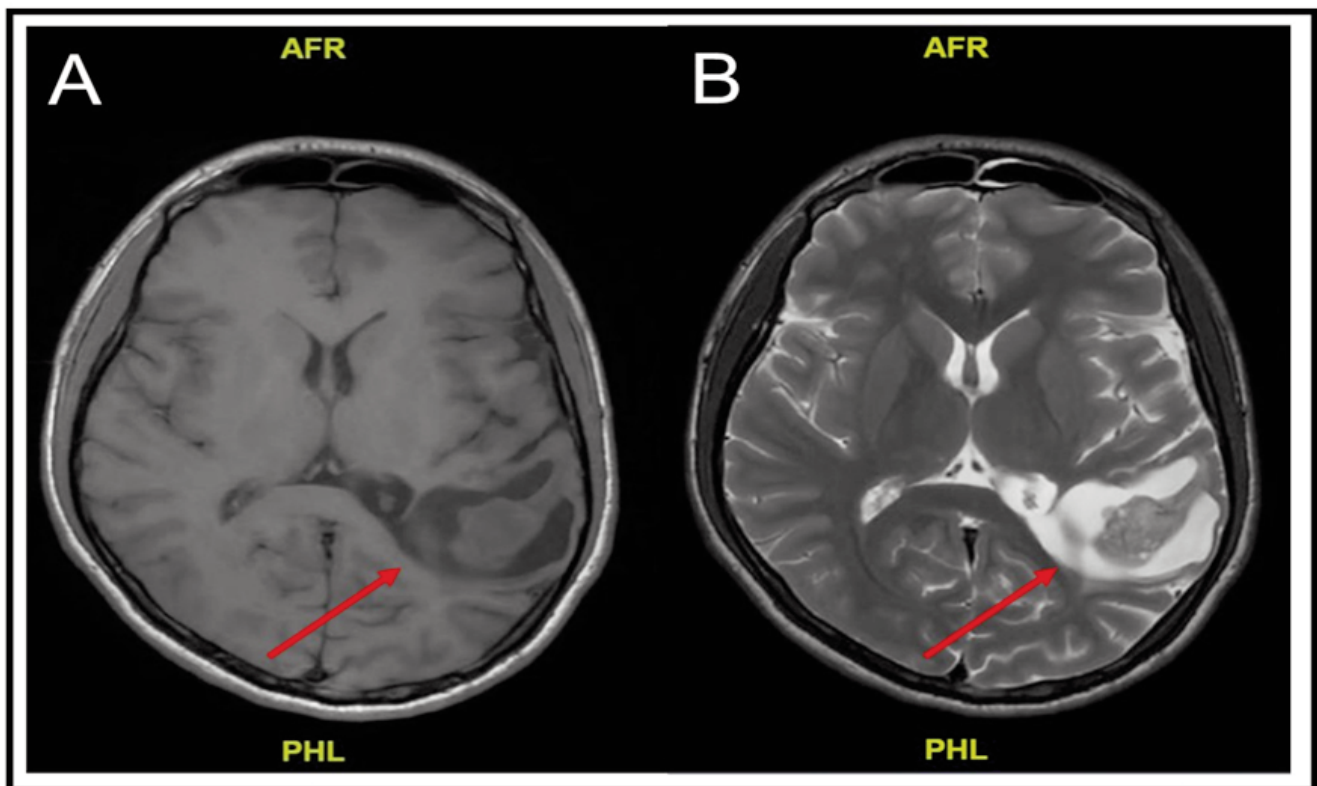


Figure-1: Axial MRI of the head without contrast showing a porencephaly cyst. (A) T1-weighted image (T1WI) and (B) T2-weighted image (T2WI), with the cyst indicated by a red arrow.

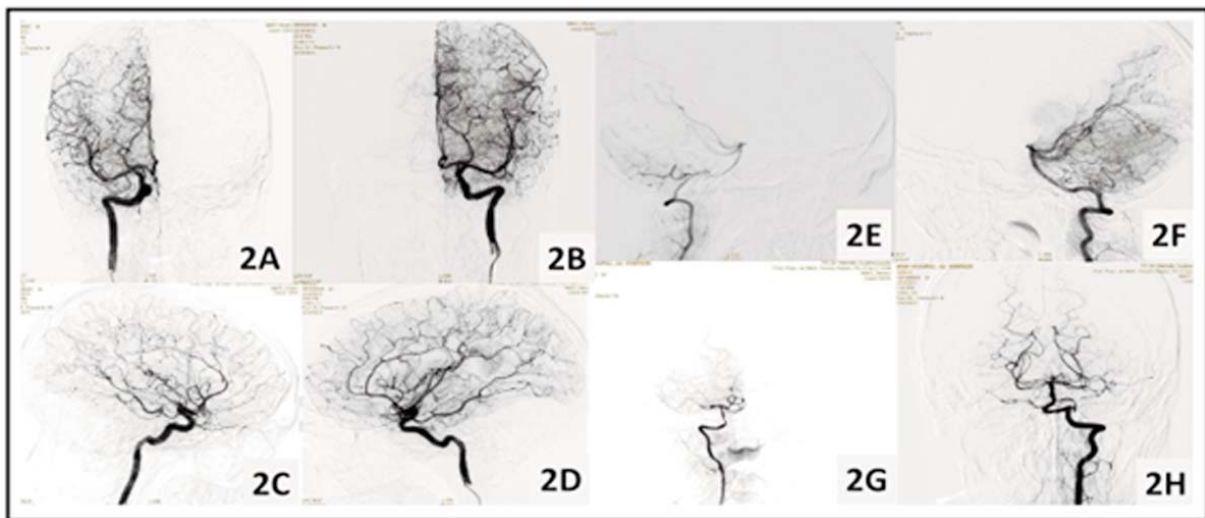


Figure-2: Cerebral angiography examination showing normal vascular patterns. (A, B) Anteroposterior (AP) views of contrast injection from the right and left internal carotid arteries (Rt-ICA, Lt-ICA). (C, D) Lateral views of contrast injection from the Rt-ICA and Lt-ICA. (E, F) Lateral views of contrast injection from the right and left vertebral arteries (Rt-VA, Lt-VA). (G, H) Townes position views of contrast injection from the Rt-VA.

2.35 x 2.19 cm was also identified within the cyst. The MRI findings were consistent with a porencephalic cyst in the left temporo-occipital region, characterised by clear

borders and its connection to the lateral ventricle (Figures 1A and 1B).

The patient also underwent a cerebral angiography, which revealed normal intracranial blood vessels with no abnormalities detected in the region of the cyst, particularly in the left middle and posterior cerebral arteries (Figure 2).

The patient was diagnosed with an acute symptomatic seizure and a porencephalic cyst in the left temporo-occipital region. He was prescribed Valproic acid therapy at a dose of 250mg every 12 hours, taken orally. At the three-month follow-up, it was noted that the patient had experienced no further seizures and was adhering to regular anti-seizure medication.

Discussion

This case highlights a rare instance of acute symptomatic seizure in a 19-year-old, attributed to a porencephalic cyst in the left temporo-occipital region. Congenital heterozygous mutations in the COL4A1 gene, which encodes the type IVa1 collagen chains crucial for maintaining the structural integrity of the vascular basement membrane, have been reported in patients with porencephalic cysts.³ From an epidemiological perspective, porencephalic cysts are primarily observed in new-borns, with an estimated prevalence of 3.5 per 100,000 live births, and are very rarely reported in adults.⁴

Clinical symptoms of porencephalic cysts vary depending on the size and location of the cyst. Patients may be asymptomatic or may present with epilepsy, focal neurological deficits, or intellectual disabilities.⁵ Seizures can be partial or generalised, while motor deficits may range from hemiparesis to severe atonic diplegia.^{6,7} Cognitive impairments also vary, from normal or mild learning disabilities to severe intellectual disabilities. Microcephaly is commonly associated with this condition.⁷ In this case, the patient experienced symptomatic epilepsy from the age of 19, without any neurological deficits. Mental status examination revealed normal results. Electroencephalography (EEG) can assist in diagnosis; however, the findings are typically non-specific.⁸ A computed tomography (CT) scan of the head often demonstrates a hypodense intracranial cyst with well-defined borders and central attenuation consistent with cerebrospinal fluid.^{9,10} Generally, there is no mass effect on the surrounding parenchyma, although large cysts in some cases can cause a localised mass effect without showing contrast enhancement.^{1,2,11}

On MRI, a porencephalic cyst appears well-defined, bordered by white matter with or without gliosis. The imaging characteristics include cerebrospinal fluid intensity, with low intensity on T1WI and high intensity on T2WI. FLAIR sequences show suppression of the fluid

signal, and DWI typically reveals no restricted diffusion.¹¹

In this case, an MRI of the head revealed a porencephalic cyst accompanied by a mural nodule in the left temporo-occipital region.

In one study, cerebral angiography of porencephalic cases demonstrated features of space-occupying lesions, including displacement of blood vessels to one side and the presence of avascular areas. Anomalies such as aneurysms may be observed, particularly in cases involving haemorrhage, intracranial sinus thrombosis, or other vascular complications. However, cerebral angiography does not show any abnormalities in certain cases.^{12,13}

In this patient, a cerebral angiography procedure was performed, which did not reveal any abnormalities in the structure of the intracranial blood vessels.

The differential diagnosis of porencephalic cysts includes arachnoid cysts, schizencephaly, and ependymal cysts. Arachnoid cysts are extra-axial lesions that displace the cerebral cortex away from the adjacent cranium. Schizencephaly presents as a cavity filled with cerebrospinal fluid, lined by heterotopic grey matter, and extending from the ventricles to the surface of the brain. Ependymal cysts, on the other hand, are typically intraventricular and surrounded by normal brain tissue.^{1,3}

Treatment for porencephalic cysts includes physical therapy and rehabilitation, antiepileptic drugs to manage seizure symptoms, and bypass surgery in cases of hydrocephalus.⁶ Surgery is also recommended for patients with drug-resistant epilepsy, with procedures such as hemispherectomy or hemicraniectomy being the options. However, these are generally reserved for children or cases involving large porencephalic cysts associated with ischaemia or trauma.¹⁴ In this case, the patient was treated with the anti-seizure medication—Valproic acid. Surgery was not performed, as the seizures were effectively controlled with Valproic acid, and there were no signs of hydrocephalus or raised intracranial pressure.

Conclusion

This case highlights the rare presentation of a porencephalic cyst in the left temporo-occipital region in a 19-year-old male, presenting with recurrent seizures without focal neurological deficits. MRI findings revealed a well-defined cyst connected to the posterior horn of the left lateral ventricle and containing wall nodules, while cerebral angiography did not show any vascular abnormalities. The patient responded well to valproic acid therapy, remained seizure-free during follow-up, and

did not require surgical intervention. This report emphasizes the importance of comprehensive neuroimaging in diagnosing rare intracranial cystic lesions and supports conservative management tailored to the patient's condition when seizures are well controlled, accompanied by close monitoring to detect potential complications.

Abbreviations

MRI: Magnetic Resonance Imaging

T1WI: T1-weighted image.

T2WI: T2-weighted image.

EEG: Electroencephalography.

CT: Computerised tomography.

FLAIR: Fluid-attenuated inversion recovery.

DWI: Diffusion-Weighted Imaging.

Availability of data and materials: All data generated or analysed during this study are included in this published article.

Ethics approval and consent to participate: This study was approved by the ethical committee at the Health Research Ethics Committee, Faculty of Medicine, Hasanuddin University, Makassar, registration number: 2521UN4.6.4.5.3L/ PP36/ 2024. The patient in this case report provided written informed consent for the writing and publication of the report.

Disclaimer: None.

Conflict of Interest: The authors declare that they have no competing interests..

Source of Funding: None.

References

- Pereira RG, Ribeiro BNF, Hollanda RTL, de Almeida LB, Simeão TB, Marchiori E. Non-neoplastic intracranial cystic lesions: not everything is an arachnoid cyst. *Radiol Bras* 2021;54:49-55. doi: 10.1590/0100-3984.2019.0144.
- Tambuzzi S, Gentile G, Zoja R. Porencephalic cyst in adult. *Autops Case Rep* 2022;12:e2021351. doi: 10.4322/acr.2021.351.
- Guey S, Hervé D. Main features of COL4A1-COL4A2 related cerebral microangiopathies. *Cereb Circ Cogn Behav* 2022;3:e100140. doi: 10.1016/j.cccb.2022.100140.
- Teunissen MWA, Kamsteeg EJ, Sallevelt SCEH, Pennings M, Bauer NJC, Vermeulen RJ, et al. Biallelic Variants in the COLGALT1 Gene Causes Severe Congenital Porencephaly: A Case Report. *Neurol Genet* 2021;7:e564. doi: 10.1212/NXG.0000000000000564.
- Siddardha SS, Ghule A, Garikapati A, Kakade Y, Kumar S. Porencephaly presenting as status epilepticus in adult: a rare case report. *Med Sci Discov* 2020;24:4639-2.
- Al-Mosawi AJ. Congenital Externally Communicating Porencephaly Presenting as Hemiplegic Cerebral Palsy: Imaging Study of a Rare Condition. *SunKrist J Neonat Pediatr* 2021;3:1013.
- Kumar KA, Chakra S, Radha Krishnan D. First seizure in an adult: porencephalic cyst a cause? *International Journal of Research and Review (IJRR)* 2019;6:251-3.
- Marchi A, Pennaroli D, Lagarde S, McGonigal A, Bonini F, Carron R, et al. Epileptogenicity and surgical outcome in post stroke drug resistant epilepsy in children and adults. *Epilepsy Res* 2019;155:106155. doi: 10.1016/j.eplepsyres.2019.106155.
- Wynne D, Abdul Jalil MF, Dhillon R. Endoscopic Fenestration of a Symptomatic Porencephalic Cyst in an Adult. *World Neurosurg* 2020;141:245-6. doi: 10.1016/j.wneu.2020.06.092.
- Stern JM, Salamon N. Porencephaly. In: Stern JM, Salamon N, eds. *Imaging of Epilepsy: A Clinical Atlas*. Los Angeles: Springer, 2022; pp 213-215.
- Lu D, Tan J, Xu H. Ventriculoperitoneal shunt for giant porencephaly: a case report and literature review. *Front Surg* 2024;11:e1389050. doi: 10.3389/fsurg.2024.1389050.
- El Hasbani G, Balaghi A, Assaker R, Rojas A, Troya M, Kofahi A, et al. Intraparenchymal hemorrhage and cerebral venous thrombosis in an adult with congenital porencephalic cyst presenting for generalized tonic-clonic seizures. *Radiol Case Rep* 2019;15:95-9. doi: 10.1016/j.radcr.2019.10.028.
- Muro I, Ramos C, Barbosa A, Vivancos J. Haemorrhage within the cavity of a porencephalic cyst: a haemorrhagic complication in a patient with COVID-19. *Neurologia (Engl Ed)* 2022;37:504-7. doi: 10.1016/j.nrleng.2021.09.003.
- Dahal R, Gurung P, Kayastha J, Malla S, Badinski T, Rajbhandari P, et al. Intraoperative electrocorticography-guided resection of the epileptogenic zone in an unusual porencephalic cyst: case report and literature review. *Ann Med Surg (Lond)* 2024;86:2309-13. doi: 10.1097/MS9.0000000000001871.

AUTHOR'S CONTRIBUTION:

MYA: Design, concept, scientific writing and final approval.

DCH: Data acquisition of clinical data, critical revision and final approval.

GVS: Data interpretation, writing, critical revision and final approval.