

POREENCEPHALIC CYST IN ADULTS : A RARE CASE WITH UNUSUAL PRESENTATION

Neurosurgery

Prof. Dr. Mrinal Bhuyan

Professor and Head, Department of Neurosurgery, GMCH GUWAHATI.

Dr Ankur Anand*

Senior Resident, Department of Neurosurgery, GMCH GUWAHATI.*Corresponding Author

ABSTRACT

Porencephalic cysts are rare conditions of cerebrospinal fluid accumulation in the brain parenchyma that is usually related to perinatal vascular events, penetrating trauma and cranial surgery. These intracranial intraxial cysts communicating to either subarachnoid space or ventricles are mainly seen in paediatrics age group and very rarely in adults. We present a rare case of 26 year old female without any significant past medical or surgical history presented to our emergency department in unconscious state following new onset generalized tonic clonic seizures on further evaluations with CT scan and MRI imaging of head. It revealed a large porencephalic cyst in left parieto-occipital region, hence we are reporting this case for its rarity and its consideration as differential for new onset seizure.

KEYWORDS

cystic lesion of brain, benign lesions, porencephalic cyst, new onset Seizure

Introduction :

Porencephalic cyst is a rare entity affecting the central nervous system, whose etiopathogenesis has not yet been clearly defined. It is a cavity within the cerebral hemisphere¹ with smooth wall lined by gliotic or spongiotic white matter, which contains cerebrospinal fluid². The cavity usually communicates directly with the ventricular system and is often seen in territories supplied by the cerebral arteries². Epidemiological point of view, porencephalic cysts are described in newborns with an estimated incidence of 3.5 per 100,000 live births, and only very rarely they are reported in adults³.

CASE REPORT :-

We present a case of 26 year old female without any history of trauma. Her past medical and surgical history was also non significant. She presented to our emergency department in unconscious state following an episode of seizure. It began as focal onset motor seizure involving the right upper Limb and lower Limb then gradually became generalized tonic clonic seizure associated with loss of consciousness. The patient was then brought to our casualty with impaired consciousness and thoroughly examined to rule out any lesions in skin or mucosa and biochemical evaluations were within normal limits. CT scan of head was done and it revealed a large porencephalic cyst present in left Parieto-occipital region communicating with the left lateral ventricle. There were no sign of intra parenchymal or intra cystic hemorrhages and there were no sign of raised ICP. Patient was managed conservatively in neuro surgical ward with anti-epileptic medications. An EEG test was done which showed no any epileptic foci or seizure like activity. MRI was done which showed intra axial intraparenchymal porencephalic cyst present in the left parieto-occipital region communicating to left lateral ventricle. Patient should gradual improvement on day 2 regaining full consciousness and her neurological status where examined which was completely normal. On further History she denied any past episodes of mild recurrent headaches and consumption of any abusive substances. She didn't had any history of perinatal intra cerebral hemorrhage and neither she was on any chronic medications and her cognitive and development parameters are well within normal limits. Patient was discharged from the hospital on day 3 with Levetiracetam 500 mg twice daily dose and was followed up in neurosurgery OPD.

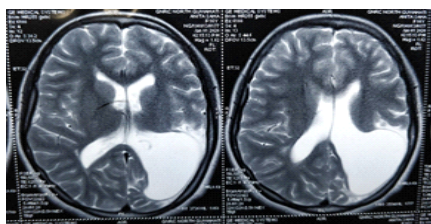


Fig 1:- T2 Axial MRI Sequence showing large single intraparenchymal cyst present in left Parieto-occipital region, communicating with the ventricular cavity.

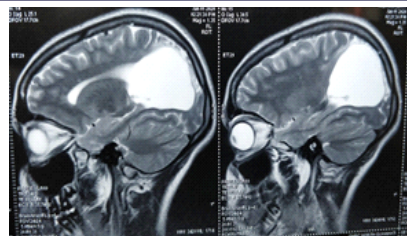


Fig 2 :- T2 Sagittal MRI Sequence showing cystic cavity present in left Parieto-occipital region communicating with the left lateral ventricle.

Discussion:-

Porencephalic cysts can be classified as congenital or acquired: it has been suggested that they are caused by a disturbance of vascular supply leading to cerebral degeneration⁴. Congenital porencephalic cysts result from intra-uterine vascular injury leading to cerebral ischemia or intra-parenchymal hemorrhage. Intra-uterine infectious injury by a virus like cytomegalovirus can also give rise to congenital porencephalic cysts⁵. Acquired porencephalic cysts are secondary to injury later in life due to trauma, surgery, ischemia, or infection⁶. De novo or inherited heterozygous mutations in COL4A1, which encodes the type IV $\alpha 1$ collagen chain, have been reported in individuals with porencephaly⁴.

Porencephalic cysts in adults without any history of trauma, surgery, ischemia and infection are very rare and are rarely reported in literature.

Clinical features are highly variable and are dependent upon cysts that vary in size and location. Patients are mostly asymptomatic or may present with mild headache, epilepsy, focal neurological deficits or mental retardation with or without signs of raised Intracranial Pressure. Seizures may be partial or generalized. Motor deficits range from hemiparesis to severe atonic diplegia. Cognitive deficits vary from normal or slight learning disability to severe mental retardation. Microcephaly is usually associated^{5,7}.

Pertaining to diagnosis of porencephaly, the electroencephalogram may help but the findings are not specific. Patients with porencephalic cysts, the background may be normal, or show slowing which may be focal or generalized⁸.

Radiological evaluations are necessary for pointing towards conclusive diagnosis. A cerebral CT scan shows a hypodense intracranial cyst, with a well-defined border and central attenuation due to the cerebrospinal fluid. Mass effect on the adjacent parenchyma is not usually observed, but very large cysts can cause it locally. Brain MRI shows a well-defined, white matter-coated brain cyst with or without gliosis, with the cerebrospinal fluid signal. Fetal MRI is also

helpful to differentiate this entity from extra-axial lesions such as arachnoid cysts but also rare tumors.⁹

Patients are mostly Managed conservatively, those presenting with seizures are generally managed with antiepileptics, and surgery is reserved for refractory cases².

Hence, this case report may help Clinicians to have a broad differential while managing case having a new-onset generalized seizures.

References :-

1. Wynne D, Abdul Jalil MF, Dhillon R. Endoscopic fenestration of a symptomatic porencephalic cyst in an adult. *World Neurosurg.* 2020;141:245-6. <http://dx.doi.org/10.1016/j.wneu.2020.06.092>. PMID:32569761.
2. Oommen AT, Sethy G, Minz NT, Patra J, Panda SS. Unusual presentation of porencephalic cyst in an adult. *J Clin Diagn Res.* 2017;11(2):OD12-3. <http://dx.doi.org/10.7860/JCDR/2017/22654.9374>. PMID:28384919.
3. Qureshi A, Jehangir A, York EP. Porencephalic cyst: a rare cause of new-onset seizure in an adult. *J Community Hosp Intern Med Perspect.* 2018;8(2):92-3. <http://dx.doi.org/10.1080/20009666.2018.1454788>. PMID:29686797.
4. Yoneda Y, Haginoya K, Arai H, et al. De novo and inherited mutations in COL4A2, encoding the type IV collagen $\alpha 2$ chain cause porencephaly. *Am J Hum Genet.* 2012;90(1):86-90. <http://dx.doi.org/10.1016/j.ajhg.2011.11.016>. PMID:22209246.
5. Kokkinos V, Garganis K, Kontogiannis K, Zountsas B. Hemispherotomy or lobectomy? The role of presurgical neuroimaging in a young case of a large porencephalic cyst with intractable epilepsy. *Pediatr Neurosurg.* 2011;47(3):204-9. <http://dx.doi.org/10.1159/000330546>. PMID:22122950.
6. Osborn AG. *Brain*. Vol. 1. Salt Lake City, Utah: Amirsys; 2004. p. 7-36. Porencephalic cyst- Choroid plexus cyst. Diagnostic imaging.
7. Ozeki M, Funato M, Teramoto T, Ohe N, Asano T, Kaneko H, et al. Reversible cerebrospinal fluid oedema and porencephalic cyst, a rare complication of ventricular catheter. *Journal of Clinical Neuroscience.* 2010;17(5):658-61.
8. PORENCEPHALIC CYST [Internet]. Available from : <https://www.epilepsydiagnosis.org/aetiology/porencephalic-cyst-eeg.html>
9. Abergel A, Lacalm A, Massoud M, Massardier J, des Portes V, Guibaud L. Expanding porencephalic cysts: prenatal imaging and differential diagnosis. *Fetal Diagn Ther.* 2017;41(3):226-33. <http://dx.doi.org/10.1159/000447740>. PMID:27409647