

Case Report

First Seizure in an Adult: Porencephalic Cyst A Cause?

Dr. Kanakapudi Anil Kumar¹, Dr. Sandeep Chakra², Dr. D. Radha Krishnan³

^{1,2}Postgraduate, ³Professor; Dept. of General Medicine, Andhra Medical College, Visakhapatnam

Corresponding Author: Dr. Kanakapudi Anil Kumar

ABSTRACT

Porencephalic cyst is quite a rare entity in adults with only a few cases reported so far. It is usually congenital and seen in neonates. Here, we report a 45-year-old female who presented with first episode of generalized tonic and clonic seizures. On evaluation, she was found to have a porencephalic cyst brain imaging. She was started on anti-epileptic drugs and is on follow up.

Key words: Porencephalic cyst, seizure.

CASE REPORT

A 45-year-old female presented with two episodes of generalized tonic - clonic seizures. Each episode lasted for about 2-3 minutes with regaining of consciousness during interictal period. Prior to the first episode patient had left sided headache for about 30 minutes during which she had 2 episodes of vomiting. It was not associated with fever, or head trauma. Alcohol and drug history was insignificant. There was no history of similar complaints, perinatal asphyxia, and developmental delay in the past. However she gives a history of headache intermittently, which was dull aching, throbbing type, not associated with any aggravating and relieving factors for the past 2 years. History of cataract surgery 2 years back. Family history was unremarkable

On general examination, patient was conscious, and coherent. Pulse was regular with a rate of 82 beats/minute. All peripheral pulses were palpable. Blood pressure was 110/70 mmHg. Respiratory rate was 16 breaths/minute and regular in rhythm. Temperature was 98.8°F. There was no pallor, icterus, clubbing, cyanosis, lymphadenopathy or oedema.

On neurological examination, Glasgow coma scale was 15/15 (E4V5M6). Cranial nerve examination demonstrated that there is incomplete, incongruous right hemianopia and complete restriction of adduction and abduction with mild restriction of elevation and depression in left eye with sensory neural hearing loss in left ear with gaze evoked nystagmus in the right eye. Ophthalmoscopy was normal. Sensorimotor examination revealed no abnormalities. Other systems examination was unremarkable.

Haematological investigations revealed haemoglobin of 12.4 g/dl with Red Blood Cell Count 3.5 million cells/ μ L, Mean Corpuscular Volume (MCV) 78.8 fL, Mean Corpuscular Haemoglobin (MCH) 28.4 pg, Mean Corpuscular Haemoglobin Concentration (MCHC) 31.6 g/dl, Total Leukocyte Count (TLC) 8600/ μ L, Total Platelet Count (TPC) 2.8 lakhs/ μ L, Erythrocyte Sedimentation Rate (ESR) 50 mm in 1st hour, Differential Leukocyte Count showed neutrophils 78%, lymphocytes 21%, eosinophils 1%, monocytes 0%, basophils 0%. Serological investigations revealed random blood glucose 170 mg/dl, urea 29 mg/dl, creatinine

1.1 mg/dl, Aspartate Aminotransferase (AST) 57 IU/L, Alanine Aminotransferase (ALT) 43 IU/L, Alkaline Phosphatase (ALP) 45 IU/L, total bilirubin 0.6mg/dl, direct bilirubin 0.2 mg/dl, protein 6.8 g/dl, Na⁺(137meq/l), K⁺(3.8meq/l), Ca²⁺(9.8 mg/dl), Mg²⁺(2.1 meq/l). Urine routine and microscopy was normal.

Electroencephalogram (EEG) is to be done. CT scan of the brain was suggestive of a large porencephalic cyst in left fronto parieto-occipital lobe communicating with left lateral ventricular system [figure 1]. MRI showed asymmetric dilation of left lateral ventricle with gross dilation of occipital and temporal horns suggestive of trapped ventricle. The differential diagnoses include cystic lesions such as arachnoid cyst, epidermoid cyst.

She was treated with anti-epileptic drugs, i.v. Dexamethasone, i.v. Mannitol and intravenous fluids and later shifted to oral phenytoin. Neurosurgical consultation was taken, who advised for VP shunting.

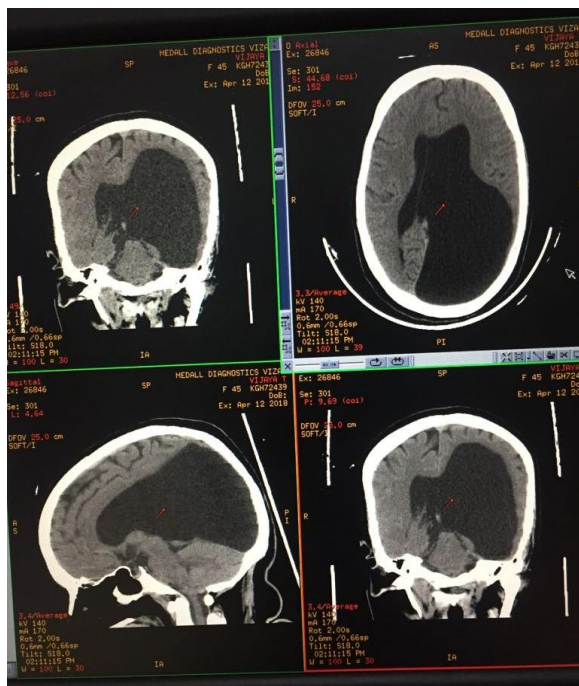


Figure 1:- Cystic enlargement in forebrain.

DISCUSSION

Porencephalic cyst is an uncommon intra-cranial cyst in adults. It is a congenital or acquired cavity within the cerebral hemisphere. It contains cerebrospinal fluid

with smooth wall lined by gliotic or spongiotic white matter. [1] It usually communicates directly with the ventricular system. It varies greatly in size. It can be cortical or sub-cortical, unilateral or bilateral and often seen in territories supplied by the cerebral arteries. It has been suggested that, porencephalic cysts are caused by a disturbance of vascular supply leading to cerebral degeneration. [2] Congenital porencephalic cysts result from intra-uterine vascular injury leading to cerebral ischemia or intra-parenchymal haemorrhage. Intra-uterine infectious injury by a virus like cytomegalovirus can also give rise to congenital porencephalic cysts. [3-5] Amygdalar-hippocampal atrophy often co-exists with congenital porencephaly (95%), and the atrophy may be bilateral despite unilateral cysts. [6] Acquired cysts are secondary to injury later in life due to trauma, surgery, ischemia, or infection. [7] De novo or inherited heterozygous mutations in COL4A1, which encodes the type IV a1 collagen chain that is essential for structural integrity for vascular basement membranes, have been reported in individuals with porencephaly. [2] Our case depicts a porencephalic cyst in adulthood but the cause is unknown.

Clinical features are variable as the cysts vary in size and location. Patients may be asymptomatic or may present with epilepsy, focal neurological deficits or mental retardation. 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. [4,5] EEG may help in the diagnosis, but the findings are not specific. CT scan brain reveals a hypodense intracranial cyst with a well defined border and central attenuation the same as cerebrospinal fluid. Usually there is no mass effect on the adjacent parenchyma, but occasionally very large cysts do result in local mass effect. It does not show enhancement with contrast. On MRI brain

cyst appears well defined and lined by white matter with or without gliosis. It contains cerebrospinal fluid signal with low signal intensity in T1: high signal intensity in T2. FLAIR shows suppression of fluid signal intensity and DWI with no restricted diffusion. The differential diagnosis for the porencephalic cyst includes arachnoid cyst, schizencephaly, and ependymal cyst. Arachnoid cysts are extra-axial and displace the brain cortex away from the adjacent skull. Schizencephaly is a CSF-filled cavity that is lined with heterotopic gray matter and extends all the way from the ventricle to the brain surface. Ependymal cysts are typically intra-ventricular with normal surrounding brain tissue. Treatment may include physical therapy, anti-epileptic drugs for seizure disorders, and a shunt in case of hydrocephalus. Surgery is advised in the patients with anti-epileptic drug resistant epilepsy. This includes hemispherectomy and hemispherotomy, although usually performed in children and in cases of large porencephalic cysts related to ischemia or trauma. Hemispherectomy is currently the surgical treatment of choice for intractable seizures associated with large, unilateral hemisphere porencephalic cysts and a neurologic deficit. An alternative minimally-invasive approach is permanent endovascular balloon occlusion in which the desired cerebral arteries are embolized. But this approach has yet to prove its safety and efficacy. In our case; the patient was clinically asymptomatic following anti-epileptic drug therapy.

CONCLUSION

Seizure is one of the most common neurological symptoms that can arise from any insult to brain. Porencephalic cyst has diverse clinical features. It may be

asymptomatic or present with epilepsy or spastic quadriparesis or mental retardation. Cases of porencephalic cyst in adults are rare and are seldom reported. An atypical case of this entity is discussed. Hence, porencephalic cyst should be considered among other differential diagnoses for seizures, mostly in children but also in adults.

REFERENCES

1. Osborn AG. Brain. Vol. 1. Salt Lake City, Utah: Amirsys; 2004. Choroid plexus cyst. Diagnostic imaging; pp. 7–30.
2. Yoneda Y, Haginoya K, Arai H, Yamaoka S, Tsurusaki Y, Doi H, et al. De novo and inherited mutations in COL4A2, encoding the type IV collagen $\alpha 2$ chain cause porencephaly. The American Journal of Human Genetics. 2012;90(1):86–90.
3. Ranjan M, Arivazhagan A, Chandrajit P, Chandramouli BA. Unusual manifestation of an orbital roof fracture in a child: Acute orbital leptomenigeal porencephalic cyst. Neurology India. 2010;58(6):974–75.
4. 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. Pediatric Neurosurgery. 2011; 47(3):204–09.
5. 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.
6. Ho SS, Kuzniecky RI, Gilliam F, Faught E, Bebin M, Morawetz R. Congenital porencephaly: MR features and relationship to hippocampal sclerosis. American Journal of neuroradiology. 1998;19(1):135–41.
7. Osborn AG. Brain. Vol. 1. Salt Lake City, Utah: Amirsys; 2004. Porencephalic cyst. Diagnostic imaging; pp. 7–36.

How to cite this article: Kumar KA, Chakra S, Radha Krishnan D. First seizure in an adult: porencephalic cyst a cause? International Journal of Research and Review. 2019; 6(2):251-253.
