Porencephaly presenting as status epilepticus in adult: A rare case report

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Citation

ABSTRACT
Porencephalic cyst is an uncommon finding in adults as it is usually congenital and seen in neonates. Here, we report a 43-year-old male who was brought to medicine ER in a drowsy state and profuse sweating after binge alcohol drinking the day prior to admission. He was found to have porencephalic cyst in left frontal lobe on imaging of the brain.

Keywords: porencephaly, status epilepticus, encephalopathy

1. INTRODUCTION
Porencephalic cyst is cyst found in intra cranial cavity. It is an unusual to find this in adults. It is mostly congenital but can also be acquired and is seen as a cavity within the cerebral hemisphere. Cerebrospinal fluid is found within the smooth wall lined cavity. The wall is formed by gliotic or spongiotic white matter. This cavity has direct communication with the ventricular system (kumar et al., 2019). It is usually congenital and it causes injury to the uterine vessels which further causes cerebral damage. Intra-uterine cytomegalovirus infection is also known to give rise to congenital porencephalic cysts (Ranjan et al., 2010). Clinical features vary according to size and location of the cyst. It may present with seizures, focal neurological deficits or intellectual disability. Seizures may be partial type or generalized tonic clonic type (Kokkinos et al., 2011). This case report highlights about 43 year old adult male presented as status epilepticus which is probably the first case report.
2. CASE REPORT
Here we are reporting a 43 year old male patient brought to emergency room in a drowsy state with profuse sweating since last 1 hour. Patient was found to be hypoglycemic with RBS of 54 mg%. Patient was shifted to MICU after stabilization. Patient had a binge alcohol drink before this event. He had no h/o headache, vomiting, seizures. Patient was afebrile. No history of chest pain, palpitations. On examination he was mesomorphic, afebrile and had a pulse rate of 96 bpm with oxygen saturation of 96% on room air. BP was 120/80 mm of Hg. Patient was drowsy and was responding to deep pain stimulus. Pupils were 3 mm and normal reacting to light. Plantars were extensor with all other reflexes were normal. His hypoglycaemia was treated with inj. Dextrose 25% i.v.

Lab investigations revealed hb 12.2 gm%, WBC counts of 8900/cu.mm platelet count of 1.18 lacs/cu.mm, urea 21 mg%, creatinine 0.3 mg %, electrolytes and liver function test were normal. RBS 54 mg% (at the time of admission), serum ammonia 62 micro gm/ dL. ABG was normal along with his ECG. His EEG was done which was found to be normal. NCCT brain revealed an ill defined CSF density area noted in left frontal region causing mild ectactic dilatation of frontal horn of left lateral ventricle. Patient developed one episode of generalized tonic clonic seizure during the hospital stay for which he was started on anti epileptic inj levetiracetam. MRI brain was done and there was altered signal intensity in bilateral basal ganglia and in sub cortical areas in bilateral temporo-occipital and high parietal region appearing hyperintense on T2/FLAIR and showing restriction on DWI and blooming on deep grey matter most likely of metabolic encephalopathy and there was well defined multi septated lesion in left frontal region appearing hyperintense on T2 and hypointense on T1 and FLAIR s/o porencephaly (figure 1). Patient was treated on inj mannitol, inj levetiracetam, i.v. antibiotics and inj dexamethasone. Neurosurgical consultation was taken but because of financial constrain it was not done.

Figure 1 T2 weighted MRI showing well defined hyperintense multi septated lesion in left frontal region.

3. DISCUSSION
Porencephalic cyst is a rare finding in adults. It may also be an acquired cavity. The wall of the cavity is lined by gliotic or spongiotic white matter (kumar et al., 2019). The cavity has communication directly with the ventricular system. The size of the cavity varies in different individuals. Its location can be in the cortex and can also be subcortical. It can be found unilaterally or bilaterally. One hypothesis is that porencephalic cysts are a result of diminished blood supply that leads to cerebral damage (Yoneda et al., 2012).

Mutations in COL4A1, which can be inherited or newly formed, have shown a role in the pathogenesis of the porencephalic cyst. COL4A1 encodes for the type IV a1 collagen chain that is necessary for structural integrity of the vascular basement membranes (Yoneda et al., 2012). Intra-uterine cytomegalovirus infection can also lead to congenital porencephalic cysts (ranjan et al., 2010). There are variety of clinical features which vary according to the size and shape. Many patients are symptomless. Others present with seizures, focal neurological deficits or intellectual disability. Seizures may be partial type or generalized tonic clonic type. Motor deficits include hemiparesis and can also cause severe atonic diplegia. There may be no cognitive involvement and the patient can also have severe mental retardation. This condition is commonly associated with microcephaly (ozeki et al., 2010). As seen on MRI
brain the cyst has a well defined white matter margin with or without gliosis. It shows low signal intensity in T1: high signal intensity in T2 as its content being the cerebrospinal fluid.

FLAIR shows reduction of the signal intensity of the fluid while DWI shows no restriction diffusion. Various other cysts can be confused with the porencephalic cyst. They are arachnoid cyst, schizencephaly, and ependymal cyst (kumar et al., 2019). Schizencephaly is like-wise a CSF-filled cavity but it is lined by heterotopic gray matter and extends all the way from the ventricle to the surface of the brain. Ependymal cysts are situated inside the ventricle and have normal surrounding brain tissue (kumar et al., 2019). Anti-epileptic drugs for seizures and a shunt in case of hydrocephalus are some treatment protocols. In patient having intractable seizures despite antiepileptic drugs, surgery is advised. Various surgeries are hemispherectomy or hemispherotomy. They are often performed in children. Also in case of a large porencephalic cyst related to ischemia or trauma surgeries are needed. The surgical treatment of choice is hemispherectomy. Embolization of cerebral arteries can be done. This is a lesser invasive procedure.

This procedure is not yet proven safe or efficacious. The patient in our case report was clinically asymptomatic following antiepileptic drug therapy. Cases of porencephalic cyst presenting as encephalopathy in adults are rarely reported.

4. CONCLUSION
Several disorders of the central nervous system commonly present as seizures and altered sensorium. Porencephaly should be considered as one of the diagnoses for status epilepticus and encephalopathy in adults.

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All the authors of the present study do not have any conflicts of interest.

Informed consent
Written & Oral informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this manuscript.

Data and materials availability
All data associated with this study are present in the paper.

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