A Rare Case of Porencephaly Presented with Status Epilepticus to Emergency Medicine

ABSTRACT

Porencephaly is an extremely rare disorder of the central nervous system involving cysts or cavities in the cerebral hemispheres. Here is a case of porencephaly who presented to the emergency department with status epilepticus. Diagnosis was made by MRI. The objective of current report is to emphasize that porencephaly can present as seizures and should be considered as an important differential diagnosis.

KEYWORDS BP-blood pressure, CT scan-computed tomography, ECG-electrocardiogram, EEG-electroencephalogram, HB-hemoglobin, MRI-magnetic resonance imaging, RR-respiratory rate, TLC-total leukocyte count

INTRODUCTION

Porencephaly is an extremely rare disorder of the central nervous system involving cysts or cavities in the cerebral hemispheres. The term “porencephaly” was coined by Heschl in 1859 to describe a cavity in the human brain. We present a case of 15-year-old girl who presented with a status epilepticus, and on imaging, she had a porencephalic cyst.

CASE REPORT

A 15-year-old girl was brought to Emergency medicine with a history of convulsions since the first day. The patient had 3–4 episodes of generalized tonic clonic seizures with brief periods of disorientation after each episode. There was no bladder bowel incontinence and no focal onset of convulsion, and no history of fever, headache, and vomiting or trauma. The patient was a known case of epilepsy but not on regular treatment. The patient was born of first degree consanguineous marriage and not immunized since birth. She had two sisters and one brother. She did not speak (no speech development). She only made some sounds. She had menarche at the age of 14 and her menstrual cycles were regular. She had mental retardation and delayed milestones.

On examination, the patient was disoriented, restless pulse-98/min, BP-110/70 mmHg in the right upper limb in supine position, RR-16/min. On neurological examination, patient had microcephaly (Head circumference–49 cm). The patient could not speak and was restless and disoriented. Cranial nerves were normal, pupils were bilaterally equal reacting to light. All four limbs were hypotonic; she was moving all four limbs. Bilateral plantars were extensors. The chest was clear and heart sounds were normal, per abdomen was normal, and there was no organomegaly.

Her Hb-11.9 gm%, TLC-10,000, serum protein & albumin were slightly decreased. Renal function, liver function, thyroid function tests, and lipid profile were normal. Her baseline ECG, chest x-ray and ultrasonography of abdomen, and pelvis were normal. On arrival, she was given Inj. Eptoin in loading dose of 600 mg I.V. slowly with Inj. Lorazepam 2 mg I.V. The patient’s convulsion stopped, and later, she regained consciousness after 1 hour. She was then started with Tab. Eptoin 100 mg BD and Tab. Tegretal 200 mg half BD for 3 days, and then half in the morning, and one at night was continued. MRI brain plain was showing partial agenesis of corpus callosum with a porencephalic cyst in right frontal lobe associated with neuronal migrational
abnormalities like adjacent Agyria-pachygria in the right frontal lobe with right-side atrophy of cerebral peduncle with scattered white matter abnormalities seen.

The course of stay in the hospital was uneventful. Neurologically, there was an improvement, and she was discharged from the hospital in satisfactory condition.

DISCUSSION

Malformations of the cerebral cortex are often associated with developmental delay and psychoses. Porencephaly is caused by either local damage from ischemia in the brain hemisphere or most commonly hemorrhage after birth. It can also occur as a consequence of abnormal development before birth\textsuperscript{1,2}. Congenital brain lesion includes genetic porencephaly resulting from maldevelopment during early neuronal migration and encephaloplastic porencephaly, which is late prenatal or perinatal vascular lesion due to arterial ischemic stroke or venous thrombosis. Porencephalic cysts can be located in any lobe or lobes of the two brain hemispheres\textsuperscript{3}.

There are a few case reports in the literature linking porencephaly with psychosis. Pae and Kim (2009) reported the case of a patient who developed a psychotic episode possibly associated with multiple leukoencephalomalacia and porencephalic changes in the brain cortex\textsuperscript{4}.

Ultrasound, CT scan and MRI can detect the cyst, which was previously missed on an air encephalography since porencephalic cyst did not communicate with the ventricular system. EEG taken over porencephalic cysts are characterised by an increased theta and delta bands in the areas surrounding the lesion sites identified by CT scan\textsuperscript{5}.

Our patient had a congenital porencephalic cyst that was diagnosed on MRI. In emergency medicine, this case can present as seizures or status epilepticus as it presented in our case.

We report this case as congenital porencephalic cyst, which is very rare, and there are no cases reported of porencephaly presenting as seizures. As there is no cure for the porencephaly, it is advisable to do proper counselling and follow up of the patients to prevent repeated episodes of seizures. It is important to stress on adherence to antiepileptic medications.

REFERENCES