A 23 year old man presented with generalized tonic–clonic seizures. The patient had history of seizures since childhood. Patient had spastic hemiparesis of left side of the body since birth. He also had mild to moderate mental retardation, speech and behavioural abnormalities.

Computed Tomography of brain was taken. CT revealed, a large cystic lesion isodense to CSF continuous with right lateral ventricle medially and with the convexity CSF spaces laterally suggestive of open lip (Type II) Schizencephaly (Fig. 1). A hypodense tram line shadow noted in left posterior parietal region segmenting the brain suggestive of a closed lip (Type I) Schizencephaly also noted (Fig. 2).

Schizencephaly or split brain is a rare congenital disorder characterized by a full thickness cleft with in the cerebral hemisphere, lined with grey matter. Schizencephaly most commonly involve parasylvian region. Schizencephaly is thought to represent a defect in neuronal migration. The leading theory indicates that schizencephaly results from an early, focal destruction of germinal matrix and surrounding brain before the hemispheres are fully formed. The term ‘schizencephaly’ was coined by Yakovlev and Wadsworth. Large portion of the cerebral hemisphere may be absent and replaced by cerebrospinal fluid. The population prevalence was 1.54 in 100,000.

Two types have been described, depending on the size of the area and the separation of the cleft lips. In closed lip or type I Schizencephaly, the cleft walls are in apposition. This fused cleft forms a furrow in the developing brain and is lined by polymicrogyric grey matter. In type II Schizencephaly or open lip, there is a large holohemispheric cleft in the cerebral cortex filled with fluid and lined by poly microgyric white matter.

The clinical manifestations depend on the severity of lesion. Patient with type I Schizencephaly are often normal and may have spasticity and seizures. Type II Schizencephaly may present with seizures, mental retardation, hypotonia, inability to walk and speak, and blindness. The identification of grey matter lining the cleft is the pathognomonic finding in differentiating Schizencephaly from porencephaly. This is best demonstrated by MRI. In Porencephaly; the CSF space is lined by gliotic white matter.

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