

Closed Lip Schizencephaly With Septum Pellucidum Agenesis: A Rare Case Report

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Abstract: Schizencephaly is a rare brain malformation disease with either congenital or acquired cause (neonatal stress). It may either be closed lip (Type 1) or open lip (Type 2)⁽¹⁾. Patients present usually with the first decade with either or combination of symptoms of mental retardation, delayed milestones or seizures. We present a case of Type 1 schizencephaly with temporal lobe heterotopia who presented to us with a chief complaint of intractable seizures⁽²⁾.

Keywords: Schizencephaly, nipple sign, heterotopias, septum pellucidum agenesis, mental retardation, delayed milestones, seizures.

I. Introduction

Schizencephaly is a cleft lined with gray matter and connecting the subarachnoid CSF spaces with the ventricular system. Schizencephaly results from injury involving the entire thickness of the developing hemisphere during cortical organization so can be either congenital or acquired⁽²⁾. The injuries are due to prenatal infection, ischemia, or chromosomal abnormalities. Clinical manifestations of schizencephaly most often include varying degrees of developmental delay, motor impairment, and seizures. It might be associated with septo-optic dysplasia, optic nerve hypoplasia or the absence of septum pellucidum, pachygyria, PMG, heterotopia, and arachnoid cysts.

Schizencephaly appears as a cleft filled with CSF and extending medially from the subarachnoid space into the lateral ventricle. The wall of the cleft is lined with dysmorphic gray matter. The gray matter sometimes extends into the lateral ventricle in the form of subependymal heterotopias. The cleft may be small or large, unilateral or bilateral. The anomaly may be of the open-lip or closed-lip type. Closed-lip schizencephaly is characterized by gray matter-lined lips, which are in contact with each other (type 1) (Nipple sign)⁽³⁾. Open-lip schizencephaly has separated lips and a cleft of CSF extending to the underlying ventricles (type 2)⁽¹⁾

Case Report

A 26 year old male patient presented with seizures for last 15 years which is poorly controlled by antiepileptics (phenytoin). Patient was advised MRI scan. Imaging findings:- T2W axial images show CSF attenuating cleft in left parietal lobe (anterior to rolandic sulcus), reaching upto the lateral margin of left lateral ventricle. The CSF attenuating communication is not well visualized near the subependymal part. Dysmorphic T2W hyperintense and T1W hypointense grey matter is seen lining the cleft throughout its course. (Fig1a,1b,2) A short nipple shape outpouching is seen in the lateral part of left lateral ventricle—*nipple sign*. (Fig1) Post Gadolinium enhance scan show few cortical vessels along the cleft in parietal lobe. Absence of septum pellucidum is also seen. No structural abnormality was seen in rest brain parenchyma. Presence of CSF attenuating cleft between left lateral ventricle and subarachnoid space of left parietal lobe with heterotropic grey matter seen along tract which is ill defined in subependymal location with presence of nipple sign is indicative of closed lip schizencephaly with septum pellucidum agenesis⁽⁵⁾. Patch of Dysmorphic grey matter with hazy GM-WM interface is seen in left temporal lobe suggestive of focal cortical dysplasia (Fig 4). The heterotopias can be a cause of refractory epilepsy as well⁽³⁾.

Bibliography

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Figures

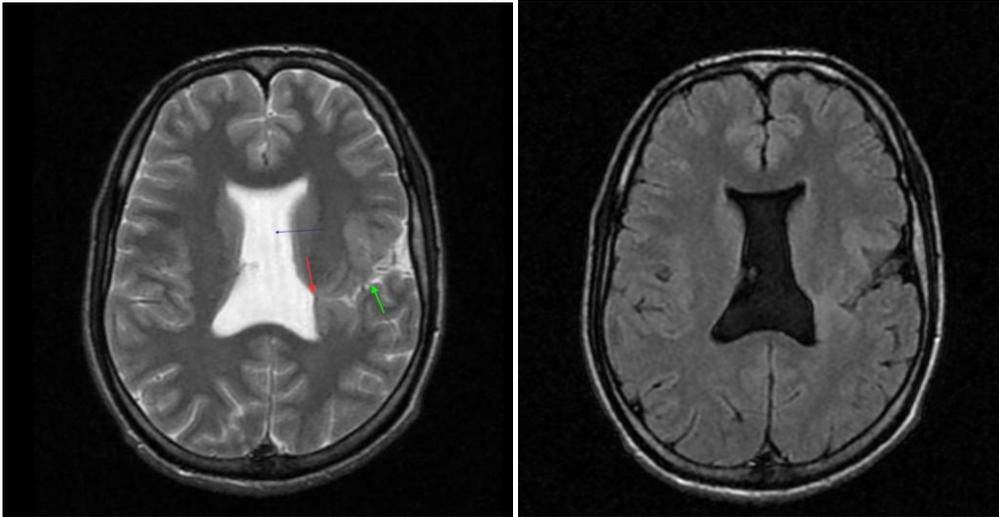


Figure 1a – Axial T2W axial images show CSF attenuating cleft in left parietal lobe reaching upto the lateral margin of left lateral ventricle (green arrow) with dysmorphic grey matter lining it. Nipple sign (Red arrow). No septum pellucidum is seen (purple arrow). Figure 1b show the same findings in an axial FLAIR section of the brain at the same level.

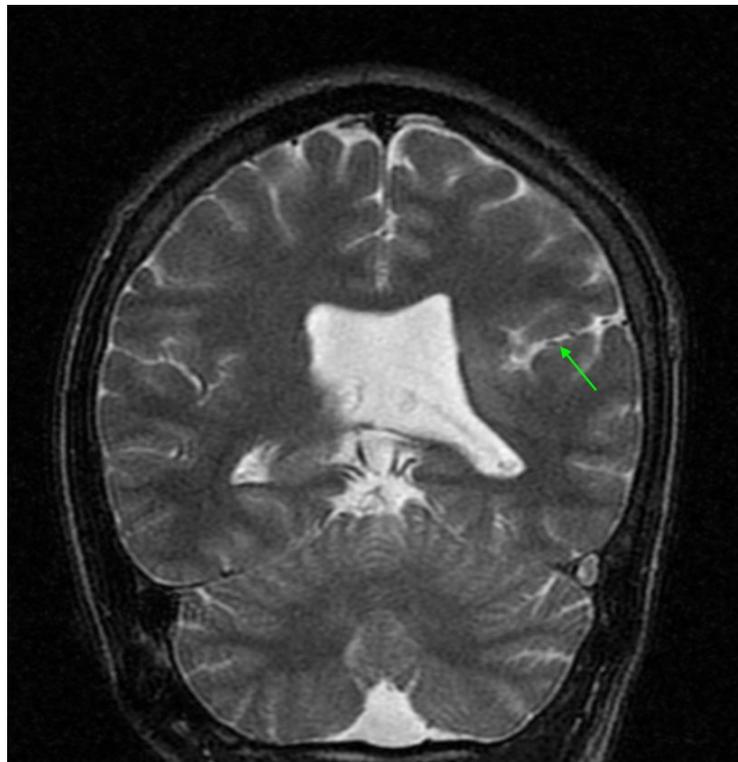


Figure 2- coronal T2 weighted image show the cleft (green arrow) with surrounding dysmorphic grey matter. Absence of septum pellucidum is also seen in the coronal image.

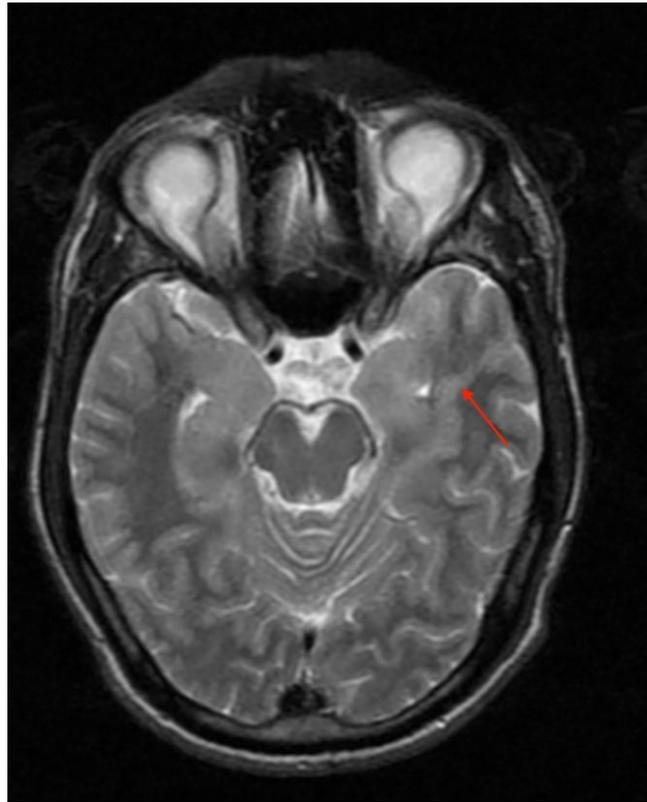


Figure 3. this axial T2 weighted image show heterotrophic grey matter in the left temporal lobe(red arrow)