

Schizencephaly

Vishal Kalia, Kavita Saggar, Vibhuti

Schizencephaly is a rare congenital disorder of cell migration with defect in sulcation. It is characterized by gray matter lined clefts. Clinically the patients presents with intractable seizures and variable developmental delay. Two types have been described in the literature - Closed Lip (Type I) which consists of fused cleft without hydrocephalous and Open Lip (Type II) consisting of open cleft with hydrocephalous. Diagnosis is made by imaging and magnetic resonance imaging is the most sensitive modality in detecting clefts and the associated anomalies like pachygyria, polymicrogyria, heterotropias, septo-optic dysplasia and absent septum pellucidum (1,2). This anomaly should be recognized early because of its genetic implications and MRI is the modality of choice.

We present a case of 16 year old male patient who presented with history of intractable seizures since last three years. No other complaints were there. The antenatal, intranatal and perinatal periods were uneventful. However, recently schizencephaly has been shown to be associated with bipolar disorder along with congenital hemiparesis, mental retardation and seizures in an isolated case reported from Jammu (3). The laboratory investigations were within normal limits. MRI study of the brain revealed close lip (Type I) schizencephaly with gray matter hetrotropias.

References

1. Barkovich AJ, Norman D. MR Imaging of schizencephaly. *Am J Roentgenol* 1988; 150(6): 1391-96.
2. Dubey AK, Gupta RK, Sharma P, Sharma RK. Schizencephaly Type I. *Indian Pediatrics* 2001; 38: 949-51.
3. Arora M, Praharaj SK, Sina VK, Sarkar S. Schizencephaly with bipolar disorder. *Singapore Med J* 2009; 50 (2): 79-80

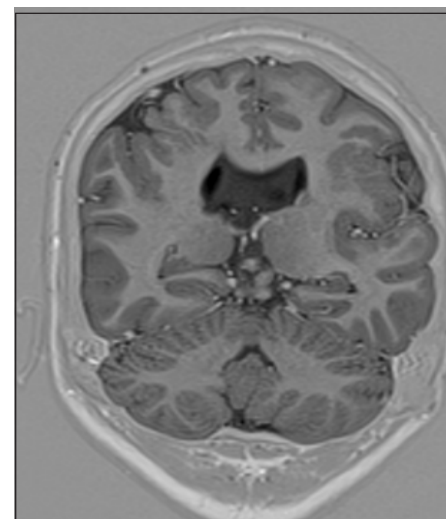
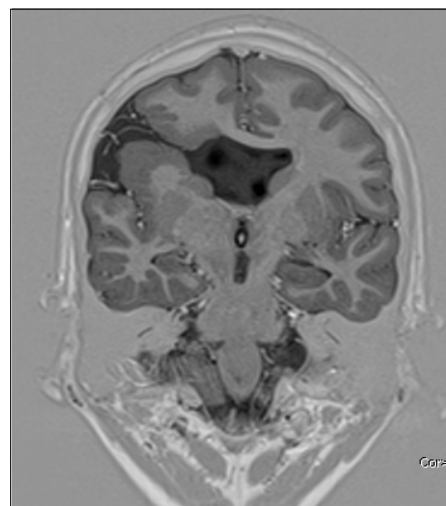


Fig. 1 Coronal Inversion Recovery MR images showing (A) a CSF filled narrow cleft lined by heterotopic gray matter in right frontal region extending from cortical surface to the lateral ventricle. Subependymal heterotopia is also noted in roof of right lateral ventricle. (B) Cortical cleft lined by thickened gray matter not reaching till the ventricle is seen in the left frontal region. Note the associated polymicrogyria.

From the Department of Radiodiagnosis, Dayanand Medical College & Hospital Ludhiana- Punjab, India

Correspondence to : Dr Vishal Kalia 464-Sector 8/II Urban Estate Karnal, Haryana-India