Case 753

Schizencephaly and septo-optic dysplasia
Published on 22.03.2001

DOI: 10.1594/EURORAD/CASE.753
ISSN: 1563-4086
Section: Neuroradiology
Imaging Technique: MR
Case Type: Clinical Cases
Authors: R.N.Sener
Patient: 3 years, male

Clinical History:
Poor vision, developmental delay, and hemiparesis.

Imaging Findings:
Presented with poor vision, moderate developmental delay, and right hemiparesis. An MRI examination was performed on a 0.5 Tesla unit, and conventional T1 and T2-weighted sequences were acquired.

Discussion:
Schizencephaly is a form of neuronal migration disorder, in which there is a cleft of the cerebral hemisphere, extending from the lateral ventricle to the pia. Clefts may be unilateral, or bilateral. They are lined by polymicrogyric gray matter. There usually are two types of the condition. Type I consists of a closed cleft, and Type II consists of wide cleft, and the clinical presentation is more severe with wider clefts. A recently proposed theory for the formation of schizencephaly is that this condition is an extreme variant of cortical dysplasia (polymicrogyria), in which the infolding of cortex extends all the way into the lateral ventricle. Congenital cytomegalovirus infection in early gestation can be a cause of schizencephaly. On the other hand, septo-optic dysplasia (deMorsier's syndrome) mainly consists of atrophic optic nerves and tracts, and absence of the septum pellucidum. Hypothalamic-pituitary dysfunction is common in the syndrome, and it can be associated with schizencephaly and the other types of cortical dysplasias.

Differential Diagnosis List: Schizencephaly associated with septo-optic dysplasia

Final Diagnosis: Schizencephaly associated with septo-optic dysplasia

References:
Barkovich AJ. Pediatric neuroimaging. Lippincott Williams & Wilkins, Philadelphia (2000).
Sener RN. Schizencephaly and congenital cytomegalovirus infection.
Sener RN.
Septo-optic dysplasia associated with cerebral cortical dysplasia (cortico-septo-optic dysplasia).
**Figure 1**

**Description:** T1-weighted image reveals a relatively thin cleft lined with abnormal gray matter (probably polymicrogyria). The septum pellucidum is absent. **Origin:**
Description: T2-weighted image reveals the schizencephalic cleft and absence of the septum pellucidum. Cortical structures in the left parietal lobe, posterior to the cleft, also appear abnormal.
Origin: