Case 1027

Semilobar holoprosencephaly

Published on 12.07.2001

DOI: 10.1594/EURORAD/CASE.1027
ISSN: 1563-4086
Section: Neuroradiology
Technique: MR
Case Type: Clinical Cases
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Patient: 5 months, female

Clinical History:

Mild hypotelorism, mental motor retardation

Imaging Findings:

The patient was presented with mild hypotelorism and mental motor retardation. MR imaging was performed with a 1.5 T MR scanner. Spin-echo T1-weighted, turbo spin-echo T2-weighted, FLAIR (fluid attenuated inversion recovery) sequences on three planes were performed for the patient.

Discussion:

Holoprosencephalies are a group of disorders characterized by a failure of differentiation and cleavage of the prosencephalon. Holoprosencephaly is a relatively common brain malformation occurring in 5-12/100,000 live births. DeMeyer has divided holoprosencephaly into three subcategories: alobar, semilobar, and lobar forms. These categories are useful for classifying of different severities. Semilobar holoprosencephaly is the intermediate form. Patients with semilobar holoprosencephaly usually have normal faces, but they occasionally have mild facial anomalies. The interhemispheric fissure and falx cerebri are usually partially formed posteriorly, however anterior portions of the brain remain fused and underdeveloped. The thalami are partially separated, resulting in a small third ventricle. The hippocampal formation remains rudimentary, the temporal horns of lateral ventricles are incompletely formed. The septum pellucidum is completely absent. The anterior portions of the corpus callosum are always absent, but the splenium is present. Holoprosencephaly is the only brain anomaly described in which the posterior corpus callosum forms in the absence of anterior callosal formation. In addition to midline anomalies, the patients commonly have disordered neuronal migration, particularly pachygyria.

Differential Diagnosis List: Semilobar holoprosencephaly

Final Diagnosis: Semilobar holoprosencephaly

References:

Barkovich AJ. Apparent atypical callosal dysgenesis: analysis of MR findings in six cases and their relationship to
Description: Sagittal SE T1 weighted image shows abnormal gyral pattern and hypoplasia of frontal lobes, splenium of the corpus callosum is present posteriorly. Origin:
Figure 2

**Description:** Axial FSE T2 weighted image shows absence of an interhemispheric fissure and falx cerebri anteriorly, resulting in fusion of frontal lobes. Thalami are partially fused with rudimentary third ventricle. **Origin:**

**Description:** Coronal SE T1 weighted image shows that hippocampal gyri are malformed. **Origin:**