

MIDDLE INTERHEMISPHERIC HOLOPROSENCEPHALY WITH SUBDURAL HEMATOMA

A.K. Poyraz, M.R. Onur¹

Key-word: Holoprosencephaly

Background: A 4-month-old boy with prenatal sonography findings consistent with corpus callosum agenesis and ventriculomegaly presented. Patient was born at 37-weeks from 18-year-old woman, gravida 1 para 1. There was no history of teratogen exposure in utero such as radiation, alcohol, drug or postnatal trauma. There was no family history of brain malformations nor chromosomal abnormalities.

Clinically no facial abnormality was observed. The mother had gestational diabetes during pregnancy.

The baby was delivered by means of cesarean section because of severe eclampsia at 37 weeks. Postnatal routine physical examination of patient was normal.

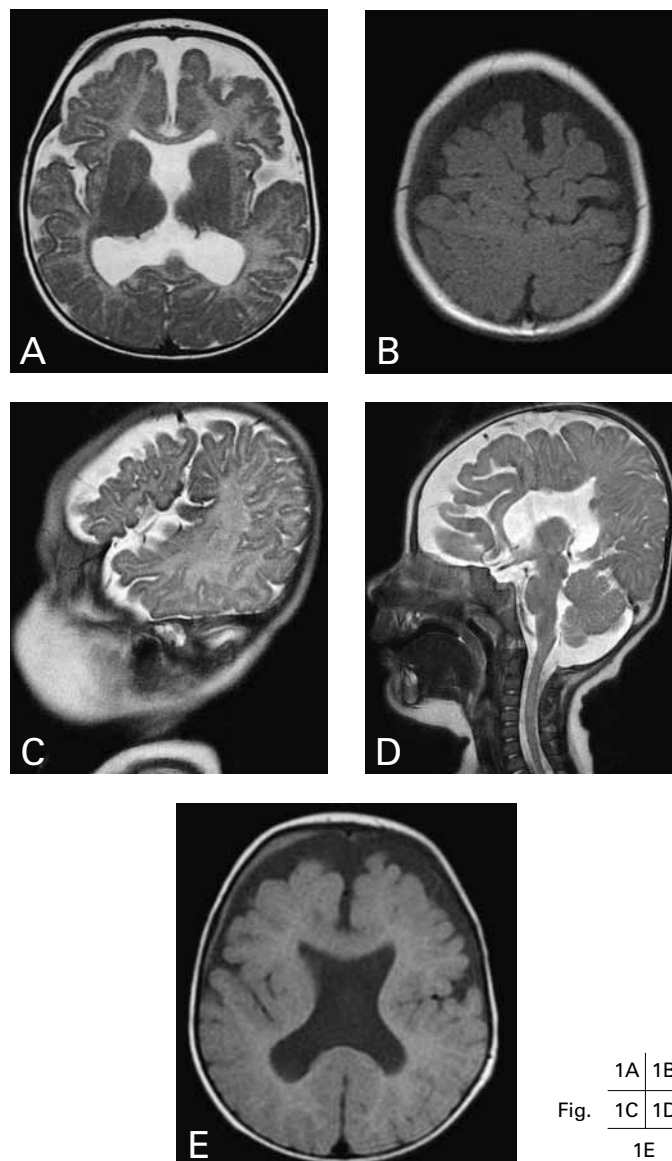


Fig.

| | |
|----|----|
| 1A | 1B |
| 1C | 1D |
| 1E | |

1. Department of Radiology, Firat University Hospital, 23119, Elazig, Turkey

Work-up

MRI of the brain (Fig. 1) showed on axial T2-weighted image (A) absence of septum pellucidum and dilated lateral ventricles. Axial T1-weighted image (B) demonstrates an interhemispheric fissure defect and continuity of cerebral white-gray matter of posterior frontal and parietal lobes. Sagittal T2-weighted image (C) visualizes a sylvian fissure extending to vertex. On sagittal T2-weighted image (D), an absent corpus callosum body with preserved genu, rostrum and splenium is seen. Axial T2 FLAIR image (E) shows a subacute subdural hematoma with intermediate intensity, at right frontal region.

Radiological diagnosis

Based on the MRI-findings the diagnosis of *syntelencephaly* (middle interhemispheric variant holoprosencephaly) *with subdural hematoma* was made. Bilateral sylvian fissures extending to vertex with perisylvian cortical dysplasia (bilateral perisylvian syndrome) and subdural hematoma were additional findings.

Discussion

Holoprosencephaly is a congenital forebrain anomaly spectrum characterized by lack of formation of the midline structures. Forebrain development fails to divide into two separate hemispheres and ventricles. Incomplete cleavage occurs in cerebral hemispheres.

According to severity of anomaly, holoprosencephaly is classified into 4 subtypes: alobar, semilobar, lobar and middle interhemispheric holoprosencephaly variant (MIHV).

Barkovich and Quint suggested that the middle interhemispheric variant of holoprosencephaly may be a spectrum of holoprosencephaly. MIVH is characterized with incomplete cleavage of the

posterior frontal and parietal lobes, dysgenesis of the corpus callosum, absent septum pellucidum with normal basal ganglia and thalami.

Anterior portions of the frontal lobes, occipital lobes and basal ganglia are normally separated in MIVH. The callosal genu and splenium appear relatively spared in this condition, but the callosal body is absent or hypoplastic.

The presence of focal posterior frontal and parietal hemispheric fusion with sparing of other lobes and absent body of corpus callosum and preserved rostrum, genu and splenium parts are findings consistent with syntelencephaly.

Gestational diabetes is a proven cause of malformations in the developing human fetus. There is a two- to fourfold elevated risk for all birth defects in gestational diabetes.

The most common malformations are sacral agenesis, complex cardiac defects, anencephaly, and holoprosencephaly. MIVH is a variant of holoprosencephaly which can appear with different forms. Because of faint signs and symptoms, MRI should be performed in all holoprosencephaly cases.

Bibliography

1. Cohen H.L., Sivit C.J.: Holoprosencephaly. *Fetal and Pediatric Ultrasound: A Casebook Approach*. McGraw-Hill, 2001, New York, NY.
2. Lewis A.J., Simon E.M., Barkovich A.J.: Middle interhemispheric variant of holoprosencephaly: a distinct clinicon neuroradiologic subtype. *Neurology*, 2002, 59: 1860-1865.
3. Pulitzer S.B., Simon E.M., Crombleholme T.M., et al.: Prenatal MR findings of the middle interhemispheric variant of holoprosencephaly. *AJNR*, 2004, 25: 1034-1036.
4. Robin N.H., Ko L.M., Heeger S., et al.: Syntelencephaly in an infant of a diabetic mother. *Am J Med Genet*, 1996, 66: 433-437.