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Prenatal MRI Image of a Fetus with Semilobar Holoprosencephaly

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صور الرنين المغناطيسي لانْدِمَاجُ مُقَدَّم الدِّماغ نصف الفصي عند جنين أثناء الحَمَل

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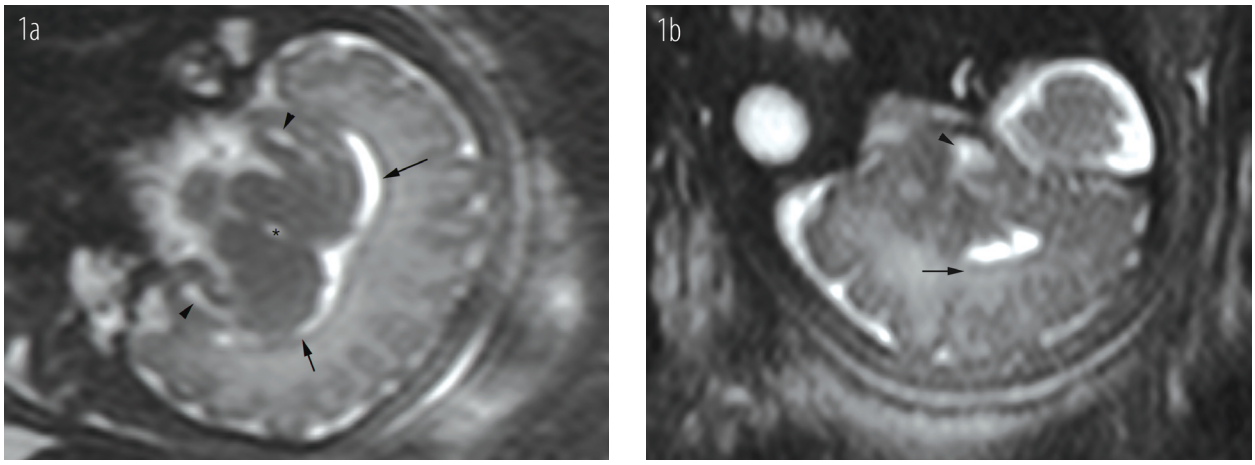

Figure 1a: ***Heavily T2W (HASTE) images in the fetal cranial coronal and sagittal planes.***

1a. ***Coronal plane at the level of the thalami: Central horseshoe-shaped single monoventricle (arrows) with absent frontal horns, absent anterior midline falx and inter-hemispheric fissure, absent septum pellucidum with failure of cleavage of frontal and parietal lobes anteriorly; rudimentary temporal horns (arrowheads); thalami are partially separated with rudimentary third ventricle (marked *)***

1b. ***Sagittal plane: Corpus callosum (arrow) is absent in the uncleavaged frontal region. Temporal horn (arrowhead) is identified. Note the proptosis.***

A 30 year old multigravida presented to the Gynecology Department of Sultan Qaboos University Hospital, Oman, at 32 weeks pregnancy. Both parents were healthy and the marriage was nonconsanguineous. There was no family history of birth defects. An antenatal ultrasound study,

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at 32 weeks pregnancy, raised the suspicion of a brain malformation, but it was suboptimal due to maternal habitus. An MRI of the fetus, at 34 weeks pregnancy, demonstrated semilobar holoprosencephaly. The baby was born at term with microcephaly, proptosis, and dysmorphic features. The diagnosis was confirmed by a postnatal computed tomography (CT) scan.

COMMENT

Holoprosencephaly (HP) is a congenital anomaly characterized by lack of cleavage of the prosencephalon. Although relatively rare, it is the most common anomaly that involves both the brain and the face. Prenatal diagnosis of this anomaly using ultrasonography, particularly of the less severe forms, is difficult. Magnetic resonance imaging (MRI) has recently become an important complement to ultrasound in prenatal diagnosis of central nervous system anomalies.[1]

HP is the most common anomaly affecting the ventral forebrain, occurring in 1/250 embryos and 1/8300-16,000 live births.[2, 3]

HP refers to a spectrum of disorders resulting from absent or incomplete cleavage of the forebrain (prosencephlon) during early embryologic development (days). HP is usually categorized as alobar, semilobar or lobar depending on the degree of forebrain cleavage. [4] Alobar is the most severe form with complete failure of cleavage of the two cerebral hemispheres. It results in a monoventricular cavity; fusion of the thalami; absence of the corpus callosum; falx cerebri; optic tracts and olfactory bulbs. Semilobar HP shares many of these same features, but demonstrates partial segmentation of the ventricles and incomplete fusion of thalami. Septo-optic dysplasia, the least severe type of HP, results in separation of the ventricles and thalami and absence of the septum pellucidum.[5] The advent of high-resolution real-time ultrasound imaging equipment has allowed detection of the group of holoprosencephalies, but lack of familiarity with uncommon forms may lead to diagnostic confusion. Coronal sonograms of the fetal head, in addition to standard axial projections, should be performed whenever an intracranial cystic abnormality is identified.[6] Several characteristic midline facial malformations are associated with holoprosencephaly, including hypotelorism. The degree of facial dysmorphism tends to parallel the severity of holoprosencephaly and, therefore, sonographic evaluation of facial morphology may aid in prenatal diagnosis.[7] Recently, diffusion tensor imaging and fiber tracking have revealed white matter structures not apparent on routine MRI imaging sequences, which are in agreement with pathologic descriptions of the holoprosencephalic brain.[8]

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