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Prenatal MR Diagnosis of a Thick Corpus Callosum

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Summary: Fetal sonography revealed cerebral, facial, and genitourinary abnormalities, prompting MR at 33 weeks' gestational age. Cerebral MR confirmed a thickened corpus callosum and showed open sylvian fissures, abnormal gyri in the frontal lobes, and presumed neuronal heterotopias. An abortion was performed at 34 weeks' gestational age, and pathologic findings corresponded well to the MR manifestations.

Index terms: Corpus callosum, abnormalities and anomalies; Fetus, magnetic resonance

Magnetic resonance (MR) imaging is being used increasingly as a complementary diagnostic technique for prenatal diagnosis of fetal brain abnormalities showed at sonography (1–3). We report a case in which prenatal sonography detected a thick corpus callosum, and subsequent MR imaging showed this unusual finding to be associated with abnormal gyral pattern and anomalous neuronal migration, worsening the prognosis.

Case Report

At 10 weeks into her first pregnancy, a 28-year-old woman had transabdominal sonography, which showed a single fetus with transitory cervical cystic hygroma. The fetal measurements at 24 weeks' gestation corresponded to the 50th percentile for the age. The fetal corpus callosum was abnormally thick, with measurement of the callosal body reaching 7 mm. Apart from the thickened corpus callosum, fetal cerebral anatomy was normal; however, retrognathia was also detected. In the fetal abdomen, sonography showed bilateral ureteral duplication with cystic dysplasia of the right upper pole. A second sonogram obtained at 31 weeks' gestation confirmed these findings. Results of trophoblastic biopsy and amniotic puncture showed normal 46,XX karyotype.

MR imaging was performed at 33 weeks' gestation on a 1.5-T unit after maternal premedication with 1 mg of fluni-

trazepam given orally 30 minutes before the examination. Section thickness was 5 mm, with a 2.5-mm intersection gap. Fast spin-echo T2-weighted (4000/114 [repetition time/echo time]) and gradient-echo T1-weighted (240/6, 110° flip angle) sequences were obtained in axial, coronal, and sagittal planes of the fetal head. MR images confirmed the abnormal thickness of the fetal corpus callosum (Fig 1A); the genu and body were 9 mm and 8 mm thick, respectively. The length of the corpus callosum was normal, with the ratio of the length of the corpus callosum to the anteroposterior cerebral diameter reaching 0.38. Furthermore, MR images showed abnormal gyri with largely open sylvian fissures (Fig 1B and C). The cortex of the frontal lobes had an abnormal cauliflower appearance, indicating possible polymicrogyria (Fig 1B). The lateral ventricular walls showed irregular thickening, hypointense on T2-weighted sequences, corresponding to probable neuronal subependymal heterotopia (Fig 1C). T1-weighted gradient-echo images did not show any acute or subacute hemorrhage.

Owing to the associated cerebral anomalies, abortion was performed at 34 weeks' gestation. Pathologic examination of the fetus showed moderate retrognathia with a palatine cleft. There was bilateral pyeloureteral duplication with dilatation and dysplasia of the right upper pole. On macroscopic examination, the sylvian fissures were abnormally open (Fig 1D). Irregular, narrow gyri were observed along the upper lip of both sylvian fissures and along the frontal lobes (Fig 1D). The corpus callosum was more than 1 cm thick and multiple nodular heterotopias were dispersed throughout the white matter. On histologic examination, the alignment of the callosal fibers was irregular, and neuronal heterotopias were again seen diffusely throughout the white matter. The macroscopic abnormal gyri corresponded to polymicrogyria, with four layers of cortex seen on histologic sections.

Discussion

Prenatal diagnosis of a thick corpus callosum is an exceptional finding. Normal sonographic

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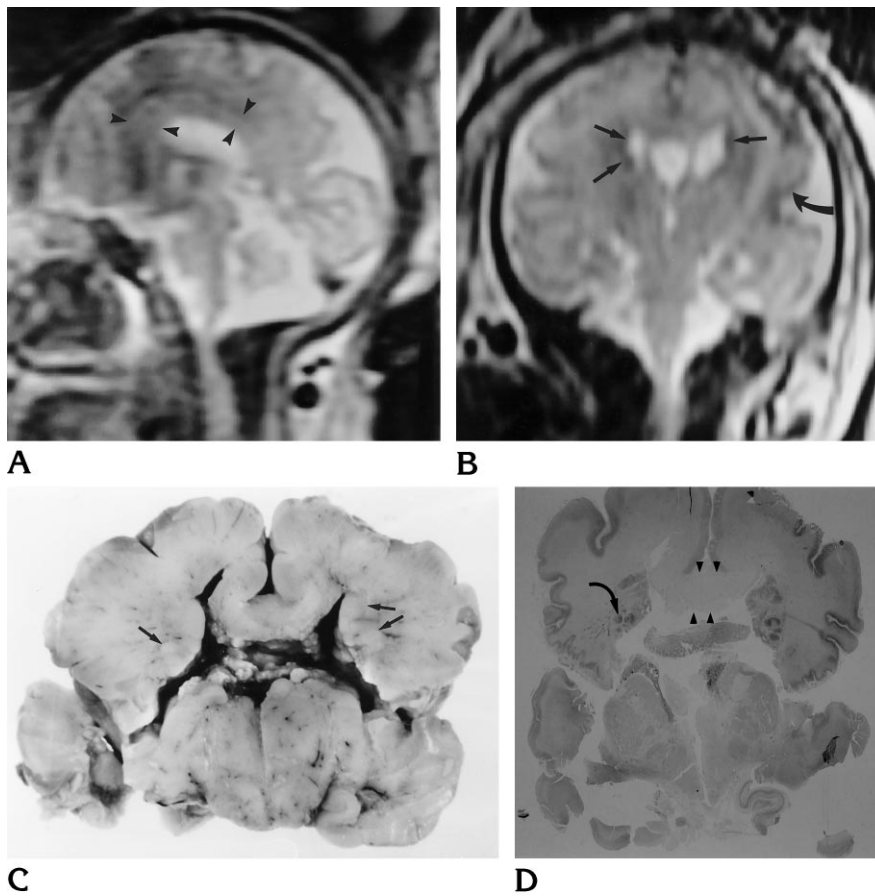


Fig 1. MR and pathologic findings in a fetus with a thickened corpus callosum.

A, Sagittal fast spin-echo T2-weighted (4000/114) MR image of the fetal head shows an unusually thick corpus callosum (arrowheads).

B, Coronal fast spin-echo T2-weighted MR image of the fetal head shows abnormally open sylvian fissure with an irregular aspect of the frontal cortex (curved arrow), which corresponded to polymicrogyria at histologic examination. Periventricular neuronal heterotopias are also well delineated (straight arrows).

C, Coronal section of the fetal encephalon shows diffuse subependymal heterotopias (arrows) and the abnormally thick corpus callosum.

D, Pathologic specimen of the coronal section of the fetal brain shows the thickened corpus callosum (arrowheads) as well as subependymal heterotopias (curved arrow) and polymicrogyria.

standards for the appearance and growth of the corpus callosum were defined by Malingier and Zakut (4). These authors showed that at 19 weeks' gestation, the genu and the body of the corpus callosum are approximately 2 mm and 1 mm thick, respectively, and that they reach a thickness of 4 to 5 mm and 3 mm, respectively, near term. All the measurements obtained in our subject were above these norms; yet the length of the corpus callosum appeared normal. Indeed, the ratio of the length of the corpus callosum to the anteroposterior diameter of the brain increases slowly from 0.29 at 18 weeks' gestation to 0.38 at term, as shown by endovaginal sonography (4).

Prognosis of a thick corpus callosum is unknown. To our knowledge, callosal hypertrophy has been described only in adults with schizophrenia (5). A precise embryological explanation for this anomaly is not readily available. The formation of the corpus callosum occurs between 8 and 20 weeks' gestation, the same period in which neuronal migration occurs (6). Therefore, callosal anomalies are often associ-

ated with other brain malformations, such as neuronal migration disorders (Aicardi syndrome).

One example of hamartomatous overgrowth of brain tissue is unilateral megalencephaly. In this malformation, overgrowth of brain tissue, including white matter, is associated with neuronal migration anomalies such as polymicrogyria, pachygyria, and heterotopia. Our case may represent such an overgrowth of white matter localized to the corpus callosum.

A precise prognosis for this pregnancy was determined by means of fetal cerebral MR imaging, which was performed successfully after maternal premedication only. Abnormal gyri were easily established by comparing the MR appearance with that in an atlas of fetal development (7). At 33 weeks' gestation, the insulae are normally deep (7). In our subject, subependymal neuronal heterotopia appeared as hypointense, irregular thickening of the lateral walls of the lateral ventricles. At 33 weeks' gestation, these signals could no longer correspond to a normal germinal matrix (7). Polymicrogyria

was suspected on the basis of the particular aspect of the cortical surface of the frontal lobes and by the presence of subependymal heterotopias and abnormal gyral pattern. This major cerebral dysgenesis was confirmed by pathologic examination.

The associated cerebral malformations detected here are impossible or difficult to diagnose by sonography; yet, the presence of associated malformations modifies the fetal prognosis and therefore the obstetric management. MR imaging is thus complementary to fetal sonography, as it can show unsuspected associated cerebral anomalies. Indeed, discovery of associated malformations by MR imaging worsens the prognosis and may be a useful indication for abortion.

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