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The cortical bite sign: a midtrimester sonographic marker of unilateral cortical focal dysplasia

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Objective

To describe the “cortical bite sign”, a specific sonographic feature of cortical focal dysplasia which is consistently seen at midtrimester axial brain ultrasound in fetuses with complete agenesis of the corpus callosum (ACC).

Methods

Retrospective analysis of prospectively collected data from 2018 to 2021, including patients referred to five fetal medicine centers in the second trimester (19^{+0} – 22^{+0} weeks of gestation) with suspected ACC. All cases with the diagnosis of complete ACC were submitted to an axial sonographic assessment of the fetal brain on the transventricular plane. On this scanning section, the mesial profile of both cerebral hemispheres at the level of the fronto-parietal cortex was investigated. In this area, the operator looked for an abnormal invagination of the cortical surface along the widened interhemispheric fissure, which was referred to as the “cortical bite sign”. All fetuses were submitted to dedicated antenatal magnetic resonance imaging (MRI) to reassess the ultrasound findings. Cases with additional brain anomalies which did not involve the cortex were excluded. The final diagnosis was confirmed at postnatal brain MRI or at postmortem examination, for cases undergoing termination of pregnancy (TOP). The primary outcome of this study was to evaluate the presence and the laterality of the “cortical bite sign” in fetuses with complete ACC. at antenatal ultrasound and MRI.

Results

In the study period, 64 cases of ACC were selected for the study purposes, of those 50 (78.1%) underwent TOP and 14 (21.9%) resulted in a live birth. The “cortical bite sign” was detected at ultrasound in 13/64 (20.3%) cases, and at targeted MRI in two additional cases (23.4%), all of which were electively terminated. Moreover, the “cortical bite sign” was found to be exclusively unilateral and on the left cerebral hemisphere in all the cases. There was a predominant majority, albeit non-significant, of male fetuses (80.0% vs 20.0%, P=0.06) in the group of complete ACC with “cortical bite sign”. No case of genetic or chromosomal anomaly was documented in our population.

Conclusion

The “cortical bite sign” is a specific marker of focal cortical dysplasia which seems to characterize a large group of fetuses with complete ACC. The etiology and prognostic role of the “cortical bite sign” remains to be investigated.