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Letters to the Editor

Prenatal ultrasonographic appearance of isolated cutis verticis gyrata

Cutis verticis gyrata (CVG) is a rare disorder of the scalp characterized by the presence of ridges and furrows that resemble the surface of the brain and that are usually associated with a syndrome such as Beare–Stevenson cutis gyrata or Turner syndrome. It has an estimated prevalence of 1 in 100 000 of the male population and 0.026 in 100 000 of the female population[1–4]. Primary CVG is commonly associated with neuropsychiatric problems such as schizophrenia, mental retardation, and epilepsy. Although Hsu *et al.*[5] have previously reported the prenatal sonographic appearance of Beare–Stevenson cutis gyrata syndrome, there was no sonographic image of the CVG. We present here the prenatal appearance of isolated CVG.

A 36-year-old Turkish woman, gravida 2, para 1, with no risk factors other than advanced maternal age, attended for antenatal care of her second pregnancy at our unit. Her first ultrasound examination, using a Logiq 9 (General Electric, Milwaukee, USA), was performed at 9 weeks of gestation for confirmation of intrauterine pregnancy. Ultrasound evaluation at 16 weeks of gestation prior to amniocentesis, revealed a fluid collection 18 mm in length and 13 mm in depth beneath the scalp in the frontal region (Figure 1). There was no brain tissue within the lesion. We were unable to clearly identify bone tissue between the intracranial structures and the lesion, raising the suspicion of the presence of a bone defect in the skull. On ultrasound examination at 20 weeks of gestation the fluid collection beneath the scalp had disappeared. Examination of the cranium showed that the skull in this region was intact. Detailed ultrasound examination of all organ systems revealed no abnormal findings, and the karyotype was normal female.

At 38 weeks of gestation spontaneous delivery of a 3350-g (50 cm in length) female occurred. Physical examination of the newborn was normal except for CVG measuring 3 × 2 cm in the frontal region (Figure 2). The lesion's skin fold depth was approximately 7 mm. Follow-up at 2 years of age revealed that the infant was mentally and physically normal apart from the clinically non-significant finding of a 5-mm discrepancy in the size of her feet, the right foot being larger than the left.

Since we did not perform a late first-trimester ultrasound examination, we could not ascertain when the lesion first appeared. The lesion was characterized by a focal fluid collection between the cranial bone and the scalp, which disappeared around 20 weeks of pregnancy. It is likely that when the fluid that had collected beneath the skin was absorbed, the skin folds were not large enough to be demonstrable by ultrasonography. Our initial differential diagnosis included cephalocele or encephalocele. However, brain tissue was not observed within the lesion. The greatest diagnostic dilemma was determining whether there was a cranial bone defect between the sac and the intracranial structures. However, on consecutive ultrasound scans performed following the disappearance of fluid at 20 weeks of gestation, a true bone defect was excluded when it was found to be acoustic shadowing.



Figure 1 Prenatal ultrasonographic appearance of the cutis verticis gyrata.



Figure 2 Neonatal picture of the cutis verticis gyrata.

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Three-dimensional sonography of unilateral Tessier number 7 cleft in a mid-trimester fetus

Facial clefts are among the most common malformations with an estimated frequency of one in 700–1000 births. They typically occur between the middle and the lateral third of the superior lip, along the ideal line connecting the nostril to the oral cavity, occasionally involving the alveolar ridge and palate, and are either unilateral or bilateral. However, in about 3% of cases, clefts occur in different portions of the face. Obstetric sonologists are familiar with the median cleft because of its association with holoprosencephaly, a condition with a high fatality rate that is not infrequent in embryos and fetuses but is rarely seen after birth[1]. The most common atypical facial cleft found at birth is the lateral cleft, which has an estimated incidence of one in 3000–5600 live births. A unilateral cleft, where the left side is more commonly affected[2,3], is characterized by a variable degree of widening of the oral commissure (macrostomia) and is frequently associated with variable hypoplasia of the lateral skeleton of the face (maxilla, zygomatic bone, ascending branch of the mandible) and external ear. According to the well-established Tessier's classification, this defect is categorized as a cleft number 7[4].

We herein report the case of a primigravida, 29-year-old woman who was referred for an anomaly scan at 21 weeks' gestation. There were no specific risk factors for fetal malformations and the pregnancy had been unremarkable. First-trimester screening by nuchal translucency measurement and biochemistry suggested a very low risk of chromosomal aberrations.

The sonogram was performed with a standard two-dimensional (2D) approach and revealed a single umbilical artery and skin tags on both cheeks of the fetus. On first observation, facial anatomy appeared otherwise unremarkable. However, the left oral commissure was unusually deep and there was the impression of an asymmetry between the left and the right side of the face (Figure 1). The remaining fetal anatomy and biometry was normal, including the amniotic fluid volume and placental insertion.

At 22 weeks' gestation a three-dimensional (3D) sonogram was performed with a GE Voluson 730 (General Electric Medical Systems, Milan, Italy). Surface rendering confirmed the presence of left macrostomia and bilateral skin tags. The left cheek appeared sunken and there was anterior displacement of the left external ear (Figure 2). A diagnosis of Tessier cleft number 7 was made. The patient had a consultation with a plastic surgeon and eventually decided to terminate the pregnancy.

There is an ongoing controversy regarding the usefulness of 3D ultrasound in the diagnosis of typical facial clefts[1,5]. We suggest that 3D ultrasound is particularly valuable for atypical clefts, and for lateral clefts in particular. The central portion of the face, nose,

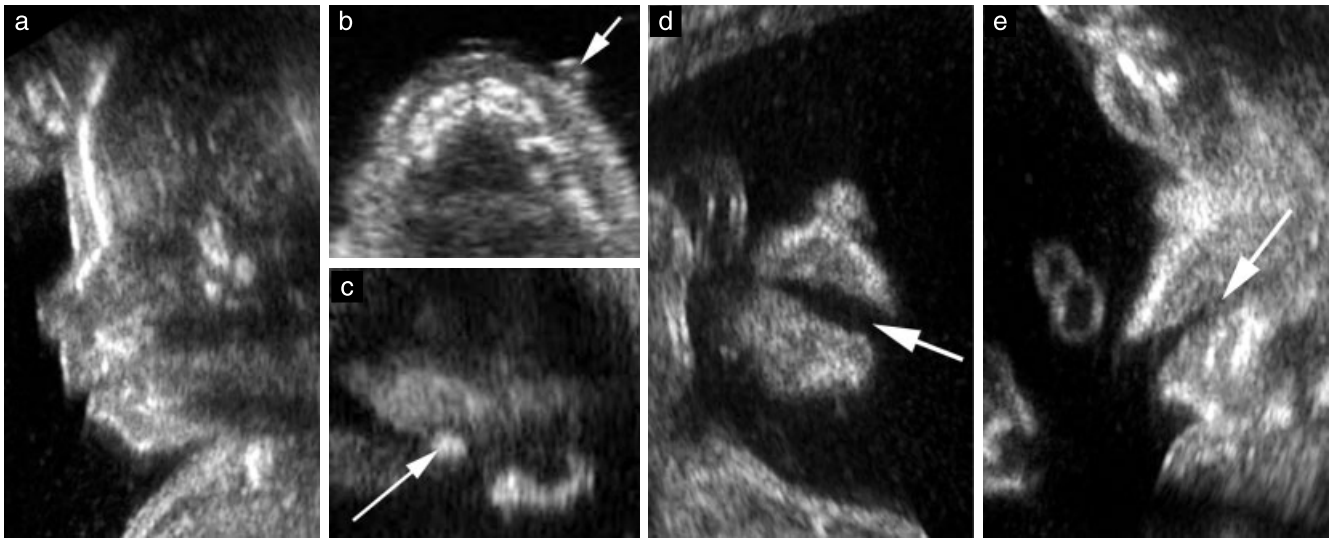


Figure 1 A composition of two-dimensional sonograms. (a) The fetal profile appears unremarkable. (b,c) Arrows point to skin tags on both cheeks. (d) A coronal view of the lips suggests that the superior lip is intact; however, there is the impression of asymmetry in the commissures of the mouth suggesting left macrostomia (arrow). (e) The impression of left macrostomia is strengthened by a lateral view of the cheek (arrow); there is only one umbilical artery in the cross-section of the cord.

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