**TWIN PREGNANCY WITH GASTROSCHISIS IN BOTH TWINS**

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**SUMMARY**

Objective: Gastroschisis is a congenital malformation characterized by an abdominal wall defect located laterally to a normal umbilicus. The cause of gastroschisis is unknown, but most authors consider it exogenous. We describe the case of a woman with a twin pregnancy in which both twins had gastroschisis.

Case Report: A 17-year-old primiparous female was referred to our institution because of a twin pregnancy, with one twin diagnosed with gastroschisis at 34 weeks of gestation. Unfortunately, gastroschisis was noted in both twins, but no other anomalies were observed under level II sonographic evaluation. The twins were delivered by cesarean section at 36+ weeks of gestation because of preterm labor and breech presentation of one fetus. Both twins presented with a 3-cm abdominal wall defect located to the right side of the umbilicus and a large portion of the bowel protruding that was not covered by membrane. Histopathology of the placenta revealed that the twins were diamniotic monochorionic. Chromosomal analysis of cord blood showed normal karyotype (46,XX) in both newborns.

Conclusion: The cause of gastroschisis is unknown, although possible exogenous causes have been studied. The diagnosis of gastroschisis in twin pregnancy is always in late gestation. Therefore, maternal serum alpha fetoprotein screening and a detailed prenatal ultrasound evaluation are recommended in multifetal pregnancies.


Key Words: gastroschisis, maternal serum alpha fetoprotein, twins

Introduction

Gastroschisis is a congenital defect of the anterior abdominal wall that results in the evisceration of abdominal contents. Gastroschisis is a relatively small defect (2 to 4 cm) that involves all layers of the ventral wall. The defect is nearly always located just to the right of the umbilicus, although left-sided defects have been described in rare instances. The incidence of gastroschisis is 0.94 per 10,000 live births [1].

The etiology of gastroschisis is unknown, although several hypotheses have been suggested [2]. Possible exogenous causes have been studied, yet, none has been proven. Younger maternal age and the use of medications, such as aspirin, oral contraceptives and pseudoephedrine during the first trimester of pregnancy, as well as the use of illegal drugs, especially cocaine, heavy alcohol consumption and smoking, were found to be significantly correlated with gastroschisis pregnancies.

A hereditary component has not been found, although familial recurrence has been reported. The hereditary factor may explain the occurrence in monozygotic twin pregnancies.

This report describes the prenatal diagnosis of a case of monozygotic twins complicated with gastroschisis in both newborns.

Case Report

A 17-year-old primiparous female was referred to our medical center because of a twin pregnancy in which
one twin was diagnosed with gastroschisis at 34 weeks of gestation. Unfortunately, gastroschisis was noted in both twins (Figures 1 and 2), but no other anomalies were observed under level II sonographic evaluation at our medical center. There was no family history of abdominal wall defects or any other congenital malformations. During the pregnancy, the mother denied any medication use, smoking or alcohol consumption. However, the 20-year-old father smoked heavily, about 15–20 cigarettes per day for several years. The parents were unrelated and healthy.

The twins were delivered by cesarean section at 36+ weeks of gestation because of preterm labor and breech presentation of one fetus. Twin A (Figure 3) was female and weighed 1,980 g with Apgar scores of 6 and 8 at 1 and 5 minutes, respectively. Twin B (Figure 4) was also female and weighed 2,240 g with Apgar scores of 6 and 9 at 1 and 5 minutes, respectively. Both twins had a 3-cm abdominal wall defect to the right side of the umbilicus with a large portion of the bowel protruding that was uncovered by membrane. There were no other malformations. The placenta weighed 1,060 g, and histopathology revealed that the twins were diamniotic monochorionic. Two hours after birth, both newborns underwent uncomplicated surgery. Both twins stayed in the neonatal unit for 5 weeks. At discharge, they were in good health and thriving.

Chromosomal analysis of cord blood showed normal karyotype (46,XX) in both newborns.

Discussion

Reports of gastroschisis in both twins are very rare. Possible exogenous causes have been studied, but none has been proven, although several risk factors have
been suggested. Maternal medications during the first trimester of pregnancy, such as aspirin, oral contraceptive and pseudoephedrine, as well as use of illegal drugs, especially cocaine, were found to be significantly correlated with gastroschisis pregnancies in different case-control studies. Heavy alcohol consumption has also been suspected. The reports found that pregnant women who smoked cigarettes were at greater risk than non-smokers. The mother of the twins in our case did not use any of these legal or illegal drugs during pregnancy. She also did not drink alcohol or smoke. However, the father had been a heavy smoker for several years. Therefore, the mother was exposed to second-hand smoke.

Gastroschisis has been associated with the abuse of substances that are known to be vasoactive, which supports a vascular origin for the pathogenesis. The nature of gastroschisis formation suggests that a teratogenic agent or vasoactive substance is responsible for initiating the defect. Nicotine, cocaine and pseudoephedrine, theoretically, should increase the risk of gastroschisis development owing to their vasoactive properties [3]. The most widely accepted etiology of gastroschisis, as proposed by DeVries [2], suggests that the defect results from abnormal involution of the right umbilical vein, which normally occurs 28 to 33 days postconception. Other authors suggested that the defect is caused by disruption of the omphalomesenteric artery.

Younger maternal age is consistently reported as a risk factor. The anomaly occurs at four times the baseline rate in women younger than 20 years of age. This may be related to an increased prevalence of cigarette smoking and drug abuse in this population. The mother in the present case was 17 years old. Two well-known risk factors for gastroschisis were presented in this case: young maternal age and exposure to second-hand smoke during pregnancy.

Widespread use of antenatal ultrasound examination has made the detection of gastroschisis feasible during the late first trimester or early second trimester. In our case, the mother was referred to our medical center because of twin pregnancy with one twin diagnosed with gastroschisis at 34 weeks of gestation. Failure to correctly diagnose abdominal wall defects by prenatal ultrasound has occurred mostly in cases associated with small defects, ruptured omphalocele, multiple fetal anomalies, intrauterine fetal death and twin pregnancies and cases referred during late gestation [4]. Prenatal detection of gastroschisis has been improved by current maternal serum alpha fetoprotein (MSAFP) screening as the defect is associated with an elevation in MSAFP level [5]. However, MSAFP screening of the present case was not performed at the local clinic.

There are many conditions that can be associated with an elevated MSAFP, such as neural tube defect, gastroschisis, omphalocele, bilateral renal agenesis and some fetal skin disorders. Sonography is used to differentiate gastroschisis from other conditions that can cause similar serum abnormalities. Therefore, MSAFP screening and a detailed prenatal ultrasound evaluation are recommended in multifetal pregnancies.

Gastroschisis is typically sporadic, although occasional familial cases have been reported, and there has been one report in a pair of monozygotic twins [6]. One study of 127 families suggested a sibling recurrence rate of 3.5%. Hereditary or genetic factors are the most plausible explanation for monozygotic twins with gastroschisis, especially when the mother denies illness or exposure to known teratogenic agents during pregnancy. Further investigation is required to elucidate the role of genetic factors in the etiology of gastroschisis.

References