CASE REPORT

Fetus with Lateral Gastroschisis Born to a Celiac Mother

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ABSTRACT

Gastroschisis is a rare anterior abdominal wall defect which develops secondary to an early embryonic vascular defect. We report here a rare site of gastroschisis involving the posterior aspect of lateral abdominal wall on the right side, born to a celiac mother. There is no reported literature on any association between gastroschisis and celiac disease. We propose here possible role of palmitic acid pathway in this index case.

Key words: Gastroschisis; Lateral gastroschisis; Celiac disease

INTRODUCTION

Gastroschisis has been traditionally thought to be caused due to early gestation vascular accident. The defect spares the midline and does not involve the umbilicus; it is usually paramedian and is more common on the right side. We report here a rare site of gastroschisis involving the posterior aspect of lateral abdominal wall on the right side in a fetus, born to a celiac mother.

CASE REPORT

Baby X was born to a 28-year-old primiparous mother at 27 (+5) weeks of gestation by vaginal delivery. Mother was a biopsy-proven case of celiac disease on dietary restrictions and was a booked case at another hospital. Level II ultrasound done at 22-week gestation showed severe oligohydramnios with absent diastolic flow; no gross congenital malformation was detected. Unfortunately, no subsequent visits were made to the treating hospital and mother went into spontaneous labor about 6 weeks later to give birth to a macerated stillborn baby. The birth weight was 600 g. Physical examination revealed herniation of the loops of small intestine from a defect in the posterior aspect of the right lateral wall of abdomen (Figure 1). No membrane or sac was present over the herniating intestinal loops. The umbilicus was normal in position. Anal opening and external genitalia were normal with no other gross congenital anomaly.

DISCUSSION

Gastroschisis is an important anterior abdominal wall birth defect that has shown an increased prevalence in recent decades. Factors possibly increasing the risk for gastroschisis include maternal age <20 years, cigarette smoking, alcohol and exposure to aspirin, cocaine, amphetamines, or the agricultural chemical atrazine [1-3]. We are not aware of any reported association of gastroschisis with celiac disease.

The gastroschisis defect does not have any covering; however, exposure to amniotic fluid elicits an inflammatory response which may give the appearance of a membrane. The normal position of the umbilicus, separate from the defect, reliably differentiates it from an omphalocele. Various theories regarding the development of gastroschisis include abnormal involution of the right umbilical vein, failure of mesoderm to form body wall, rupture of amnion around umbilical ring, and disruption of the right vitelline artery.

Lubinsky, in 2012, hypothesized estrogen-induced thrombosis for pathogenesis for gastroschisis. He rationaled that high maternal estrogen during early pregnancy causes thrombosis of fetal vessels; pal-

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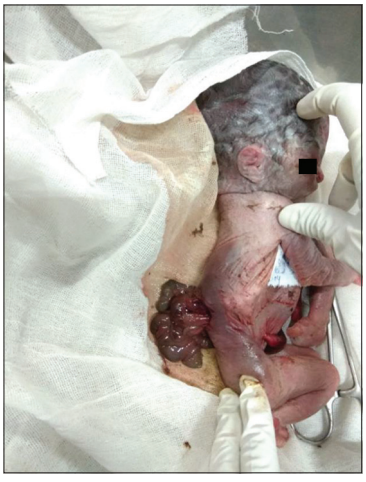

Figure 1: Defect on the right posterolateral aspect. No membrane covering bowel loops. Umbilicus away from defect in normal position

mitic acid, which is a byproduct of thrombosis, affects cell signaling and prevents fusion of body wall folds [4]. Lubinsky's hypothesis gave explanation for most of the etiological factors earlier thought to be responsible for gastroschisis [5].

Based on Lubinsky's hypothesis, we propose here the likelihood cause in our case. The celiac disease patients tend to have higher omega-6 polyunsaturated fatty acid levels [6], which is a precursor of palmitic acid through cyclooxygenase pathway. This palmitic acid can cross placenta and may lead to causation of gastroschisis by preventing cell signaling and fusion of body wall.

The fact that a Level II ultrasound done at 22-week gestation missed this anomaly is noteworthy. Barisic et al., in their data collected by 19 congenital malformation registries from 11 European countries, reported an 83% sensitivity of antenatal ultrasound for detecting gastroschisis and 75% for omphalocele [7]. Mean gestational age at the first diagnosis was 20 ± 7 weeks for gastroschisis and 18±6 weeks for omphalocele. Saller et al. found that median alpha-fetoprotein (AFP) levels in the second trimester were significantly higher in pregnancies with gastroschisis (nearly all cases) and omphalocele (most cases) as compared to unaffected pregnancies [8]. A focused ultrasound after an elevated maternal serum AFP is less likely to be false negative vis-à-vis a routine scan. As the cauliflower-like floating and inflamed bowel loops are the predominant sonographic findings rather than the defect itself, abnormal site of gastroschisis in our case is unlikely to have contributed to the false-negative ultrasound report.

The prognosis for gastroschisis is marred by associated IUGR and oligohydramnios, which frequently complicate antenatal course. Loss of protein and nutrients through the exposed intestines may be responsible for IUGR in these fetuses. IUGR may be overestimated if formulae relying on abdominal circumference are used. In fetuses with abdominal wall defects, special formulae to determine fetal weight, which are based on biparietal diameter, occipitofrontal diameter, and femur length should be used [9].

The fertility period in celiac disease is shortened and celiac women on a normal diet suffer from spontaneous abortions and other complications of pregnancy more often than those maintaining a gluten-free diet [10]. Our case was born stillbirth, may be affected by non-compliance of gluten-free diet or late diagnosis of celiac disease.

To conclude, we present this case with very unusual location of defect in the posterior aspect of lateral abdominal wall. This site of defect cannot be explained with vascular perfusion deficit. We hypothesize the external disruption as cause for this unusual site. Furthermore, its presence in celiac mother may open window to future research. We hypothesize the likelihood role of palmitic acid in this index case.

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