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Case Report Open Access

Atypical Double-Bubble in MRI of a Fetus with Double Atresia Involving Esophagus and Jejunum

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Abstract

Double-bubble sign is seen in fetuses with various alimentary tract obstructions, and typically with duodenal atresia. Atypically enlarged and clear double-bubble with enlarged distal esophagus on MRI was observed in a fetus with polyhydramnios at gestational week (GW) 27 suggesting double atresia of the esophagus and duodenum. Postnatal investigations and surgical procedures performed in this otherwise healthy male neonate weighing 2794 g at GW 36 revealed double atresia involving the esophagus (no fistula to trachea) and proximal jejunum. He transiently required tracheal intubation because an increased intra-abdominal pressure caused insufficient respiration. Following surgical repair, the patient left hospital at 69 days of age with excellent clinical course. Differential diagnoses should include a rare anomaly of double atresia in the esophagus and proximal jejunum in infants with double-bubble sign.

Keywords: Double-Bubble sign; Esophageal atresia; Double atresia; Fetal diagnosis; Neonatal management

Introduction

As fetal scanning is key to the diagnosis of various congenital anomalies, prenatal ultrasound is widely used in obstetrical practice in Japan. We encountered recently a pregnant woman with atypically enlarged and clear double-bubble in the fetal abdomen. This case indicated that differential diagnoses should include a rare anomaly of double atresia occurring in the esophagus and proximal jejunum in fetuses with double-bubble sign.

Case Report

A 28-year-old nulliparous Japanese woman presented with double-bubble sign in the fetal abdomen and polyhydramnios on ultrasound study at gestational week (GW) 25. Atypically enlarged and clear double-bubble with enlarged distal portion of the esophagus on MRI study at GW 27 in the fetus suggested double atresia of the esophagus and duodenum in this patient (Figure 1). Chromosomal analysis using amniotic fluid obtained at amnioreduction at GW 33 for treatment of polyhydramnios revealed normal male karyotype. An otherwise healthy infant was born vaginally at GW 36 weighing 2794 g, but required tracheal intubation because of insufficient respiration caused by the abdominal distension. He was diagnosed as having double atresia, including jejunal atresia 3 cm distal to the ligament of Treitz and esophageal atresia. The distance between the esophageal pouches was less than 1 cm and there was no tracheoesophageal fistula (TEF). These were surgically repaired at 1 and 36 days of age, respectively. The patient left hospital at 69 days of age with an uneventful postoperative course.

Discussion

To our knowledge, there have been no reports on double atresia involving the esophagus and jejunum showing double-bubble sign. The double-bubble sign, originally described on plain radiography, but now also appreciable on ultrasound and MRI, is a result of excessive fluid-filled structures in the abdomen. Therefore, double-bubble sign is seen prenatally in fetuses with various alimentary tract pathologies [1-5]. Although the most frequent pathology for the double-bubble sign is duodenal stenosis/atresia [1], it is not exclusively pathognomonic for duodenal atresia. Pathologies other than duodenal stenosis/atresia presenting with a double-bubble sign include cystic biliary atresia [2], colonic duplication [3], malrotation with midgut volvulus [4], and triple gut atresia [5].

As duodenal atresia is present in 3 – 6% of patients with esophageal atresia [6,7], double atresia of the esophagus and duodenum was suspected in the present case. In such cases with double atresia, an increased intra-abdominal pressure due to gastric distension hampers

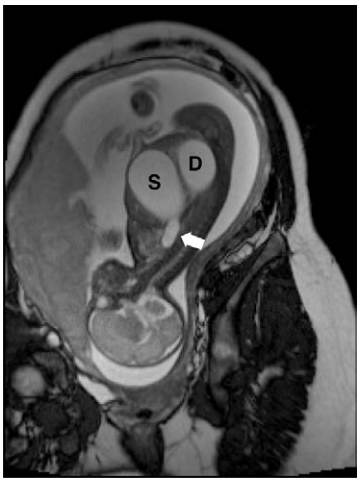

Figure 1: Sagittal steady-state free-precession MR Image at gestational week 27. The white arrow indicates the dilated caudal esophageal pouch. S, stomach; D, Duodenun.

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the movement of diaphragm causing an insufficient respiration of neonates. A nasogastric tube is helpful for the decompression of intra-abdominal pressure in neonates with duodenal atresia alone, but not in those with the esophageal atresia. Although the present case did not have TEF, the presence of TEF predisposes the patient to respiratory compromise from aspiration. A placement of gastrostomy tube is recommended for gastric decompression before all surgical procedures [8]. Therefore, both the tracheal intubation and gastric paracentesis had been prepared and the former, but not the latter was indeed required in the present case. Thus, the antenatal suspicion of double atresia involving the esophagus and lesions distal to the esophagus contributed to the safe management in this patient.

The present case indicated that double atresia involving the esophagus and jejunum should be added to the differential diagnosis of causative pathologies leading to the double-bubble sign.

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