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Congenital high airway obstruction syndrome (CHAOS) combined with esophageal atresia, tracheoesophageal fistula and duodenal atresia

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ABSTRACT

Congenital high airway obstruction syndrome (CHAOS) is a rare congenital anomaly and the most common etiology is laryngeal atresia. Recently, an increasing number of cases have survived due to prenatal diagnosis and pre- and peri-natal care including ex-utero intrapartum treatment (EXIT). More than 100 cases of CHAOS have been reported, and about half of them were complicated with associated anomalies.

Here we report a very rare case of prenatally diagnosed CHAOS (laryngeal atresia) complicated with esophageal atresia, tracheoesophageal fistula (TEF) and duodenal atresia, and the patient was saved by EXIT. This combination of anomalies resulted in a very confusing prenatal diagnosis with unique imaging feature of the fetus.

Congenital high airway obstruction syndrome (CHAOS) [1,2] is a rare congenital anomaly that was thought to be fatal about two decades ago but recently more and more cases have survived owing to prenatal diagnosis and pre- and peri-natal care including ex-utero intrapartum treatment (EXIT) [3]. More than 100 cases of CHAOS have been reported previously and about half of them were complicated with associated anomalies in the cardiocirculatory system, digits, musculoskeletal system, genitourinary system, gastrointestinal system, and some other organs [4].

Here we report a very rare case of CHAOS complicated with esophageal atresia, tracheoesophageal fistula (TEF) and duodenal atresia. This combination resulted in a very confusing prenatal diagnosis with unique imaging features of the fetus and may be a distinct entity of the disease.

1. Case

A 21-year-old pregnant female was referred to our institute for further investigation of her fetus. The fetus was suspected of having duodenal atresia at 18 weeks of gestation. Ultrasound examination of the fetus in our institute revealed a single umbilical artery, dilated trachea and dilated lower esophagus (Fig. 1a) that seemed to connect to the lower tracheal part; however, we could not detect the periodic flow of amniotic fluid into the trachea. Both lungs were hyperinflated with a flattened diaphragm and the lungs compressed the heart (Fig. 1a). These findings suggested CHAOS and TEF. Polyhydramnios and double bubble sign were detected

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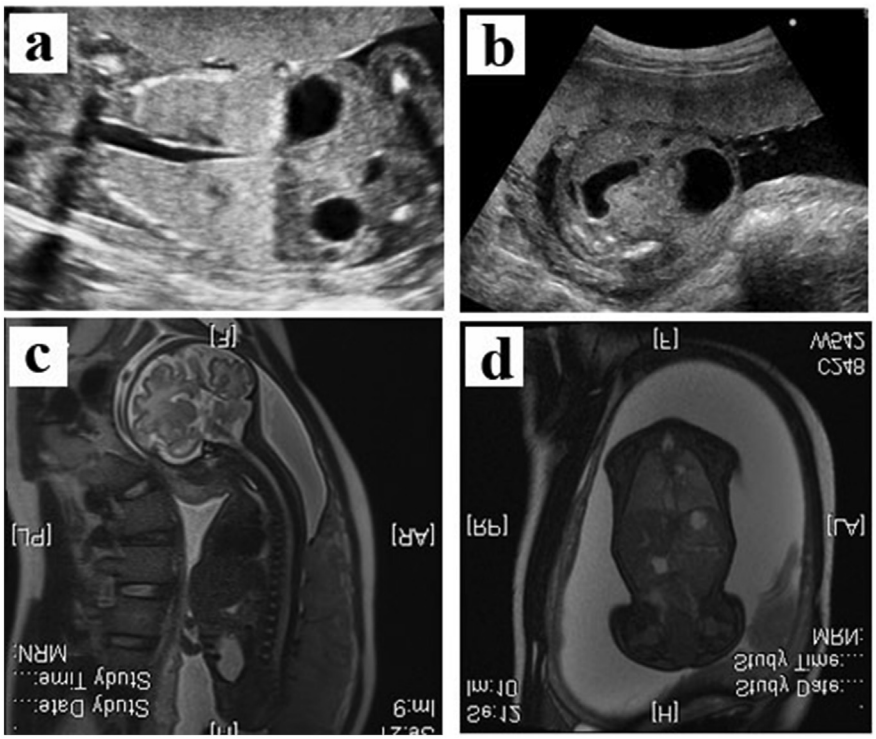

Fig. 1. Fetal diagnostic images. a) A fetal ultrasonogram taken at 18 weeks of gestation. Coronal section of the fetus is shown. The dilated upper trachea seemed to connect to the lower esophagus. Both lungs were hyper-inflated and showed a high echogenic texture. The diaphragm was flattened. Double bubble sign was clear in the abdominal cavity. b) A fetal ultrasonogram taken at 18 weeks of gestation. Transverse section of the fetal abdomen is shown. The two cystic areas were thought to be the dilated stomach and duodenum. c) Fetal MRI taken at 35 weeks of gestation. Sagittal section of the fetus is shown. The upper esophagus formed a blind pouch and the trachea was directly connected to the lower esophagus. The upper part of trachea seemed to interrupt its luminal continuity and we judged the obstructed level of the airway was at larynx. d) Fetal MRI taken at 35 weeks of gestation. Coronal section of the fetus is shown. The diaphragm was not inverted and had a normal shape. A dilated lower esophagus was detected at the mediastinum. Double bubble sign was seen in the abdominal cavity.

(Fig. 1b), which suggested congenital duodenal atresia. However, fetal magnetic resonance imaging (MRI) taken at 35 weeks of gestation showed that the upper esophagus was dilated and formed an end pouch, and the lower esophagus was also dilated and seemed to connect to the trachea (Fig. 1c). The tracheal continuity seemed to be interrupted at the laryngeal level (Fig. 1-c). There was no characteristic feature that suggested CHAOS (i.e., hyperinflated lungs, inverted diaphragm, and ascites and hydrops) (Fig. 1d). From the chronological changes in the images, it was difficult to explain the fetal pathophysiologic abnormalities but we finally concluded that the fetus was complicated with CHAOS, esophageal atresia with TEF, and duodenal atresia according to the hypothesis of the following mechanisms. The upper alimentary tract formed a closed loop by esophageal atresia and duodenal atresia, and its closed space connected to the trachea and became the buffer space for the massive amount of fluid produced by the fetal lungs. Also the following factors might affect these imaging change; the volume of the lung fluid decreased and the fluid absorption from the stomach might increase, with the progression of gestation. These situations explained why the fetus was complicated with CHAOS but did not show the typical images of lungs affected by CHAOS at 35 weeks. Amniotic fluid was taken and examined at 19 weeks of gestation, and no chromosomal abnormality was detected and the karyotype was 46-XX. The status of the fetus was carefully explained to the mother and she selected to continue her pregnancy.

At 35 weeks and 5 days of gestation the baby was delivered by the EXIT procedure. Before separating the umbilical cord, tracheostomy was performed. The trachea was prominently dilated. Just after incising the trachea, a large amount of dark-greenish, cloudy fluid overflowed from the trachea; this fluid had been stored in the closed space of the upper alimentary tract. After successful tracheostomy by EXIT procedure, the patient was transferred to the next operating room and a plain roentgenogram was taken and double bubble sign was ascertained (Fig. 2a). A nasogastric tube could not be inserted into the stomach. These clinical findings supported our prenatal diagnosis and the patient underwent laparotomy. Esophageal banding and gastrostomy were first performed to prevent perforation of the hyper-dilated stomach, and after the procedure, side-to-side diamond anastomosis of the atretic duodenum was performed.

The infant was transferred to the neonatal intensive care unit (NICU) and intensive care was started. It was difficult to provide respiratory care because intermittent desaturation occurred perhaps due to mucus regurgitation from the dilated lower esophagus (Fig. 2b). We decided to transect the TEF and at 12 days of age the second surgery was performed. Right thoracotomy was performed, and transection of the fistula was successfully performed. However, the presence of a long gap (the gap was about 2.5 vertebral width) between the upper and lower esophagus became apparent and the respiratory condition was not satisfactory, we did not anastomose the esophagus at this time. Laryngoscopy was performed at two months old and laryngeal atresia type 1 was confirmed.

The patient was well cared for in the NICU, and to elongate the upper esophagus the bougienage of the upper pouch was started at