Clinical Case Report

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Congenital high airway obstruction with tracheoesophageal fistula

A case report

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Abstract

Rationale: Congenital high airway obstruction syndrome (CHAOS) is defined as complete or partial obstruction of the fetal upper airways. Laryngeal atresia is the most frequent cause.

Patient concern: A male neonate born with poor reactions, weakly spontaneous breathing and cyanosis of the limbs was referred to our hospital.

Diagnosis: CHAOS with tracheoesophageal fistula.

Intervention: A tracheostomy was performed and a 3.0-mm internal diameter tracheostomy tube was inserted.

Outcomes: Neonatal survival depended on our immediate postnatal intervention.

Lessons: In summary, a multidisciplinary team including otolaryngologists, radiologists, obstetricians, and anesthesiologists must be present during the whole diagnosis and treatment process.

Abbreviations: CHAO = congenital high airway obstruction syndrome, EXIT = ex utero intrapartum treatment.

Keywords: congenital high airway obstruction syndrome, ex utero intrapartum treatment, fetal magnetic resonance imaging, laryngeal atresia

1. Introduction

Congenital high airway obstruction syndrome (CHAOS) is defined as complete or partial obstruction of the fetal upper airways.[1] Laryngeal atresia is the most frequent cause, but other etiologies, such as laryngeal cyst, subglottic stenosis or atresia, tracheal atresia, and laryngeal or tracheal agenesis are also included.[2,3] Most cases occur sporadically and the true incidence of HAO in the fetus is unknown. This is a rare and life-threatening condition with high mortality that is reported 80% to 100% of cases.[4] In this article, we report a patient with CHAOS with tracheoesophageal fistula who survived because of our immediate postnatal intervention.

2. Case report

A 35-year-old woman, gravida 3, parity 2, was referred to the hospital at 37-week gestation for cesarean delivery because of breech presentation and polyhydramnios. The male neonate was born with poor reactions, weakly spontaneous breathing and cyanosis of the limbs, and Apgar scores of 5 at both 5 and 10 min. His respiration appeared to be difficult with inspiratory stridor after delivery. He was normothermic, respiratory rate was 62 breaths/min, heart rate was 170 beats/min, and SO_2 (Oxygen Saturation) was 66% to 78% (by pulse oximetry) showed by electrocardiography. Immediately, they attempted to place an endotracheal tube under direct visualization, but this ended in failure. Meanwhile, the neonate was transferred to our department with continuous positive airway pressure. Shortly afterward, he became stridulous and then cyanotic with SO_2 50% shown by pulse oximetry. The blood gas analysis showed mixed acidosis which meant that vital signs were not stable. Intubation was attempted with a 3-, 2.5- and 2-mm internal diameter endotracheal tube but was unsuccessful. A definitive tracheostomy should be performed, or else, dangerous plasma CO_2 levels may build up. A 3.0-mm internal diameter tracheostomy tube was inserted, and the patient was stabilized with pulmonary support overnight (Fig. 1). At postnatal day 2, flexible fiberoptic laryngoscopy revealed laryngeal atresia at the vocal cord plane, and a small hole-like defect was detected in the upper third of the esophagus, through which the tracheal ring could be seen (Fig. 2). To evaluate further the connection between the trachea and

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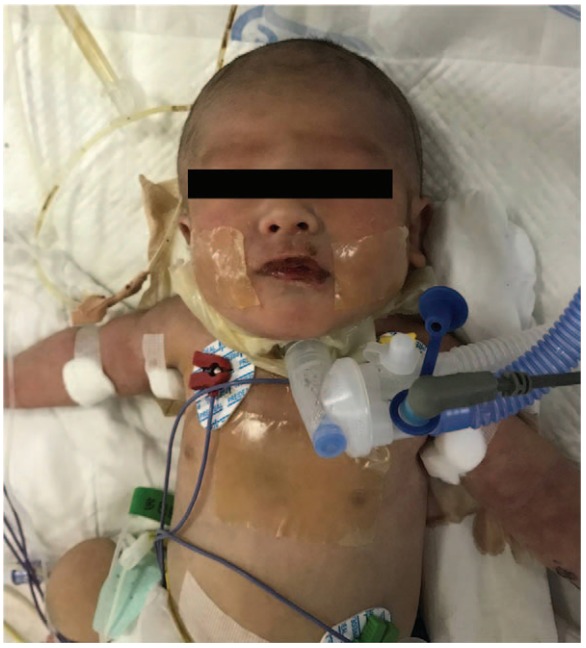

Figure 1. The patient stabilized with pulmonary support under tracheotomy.

esophagus, a computed tomography (CT) scan was performed. In contrast, CT, a 5-mm deep and 3-mm wide air-filled pouch were seen in the midline in the bifurcation, at the level of the tracheal juga, extending caudally between the esophagus and the main bronchi was absence (Fig. 3). The finding was consistent with the hole-like structure seen on fiberoptic laryngoscopy. The parents were counseled regarding the relatively poor prognosis of the syndrome, and the neonate was given up after the consent of the patient and the family members. Autopsy findings were consistent with laryngeal atresia (Fig. 4).

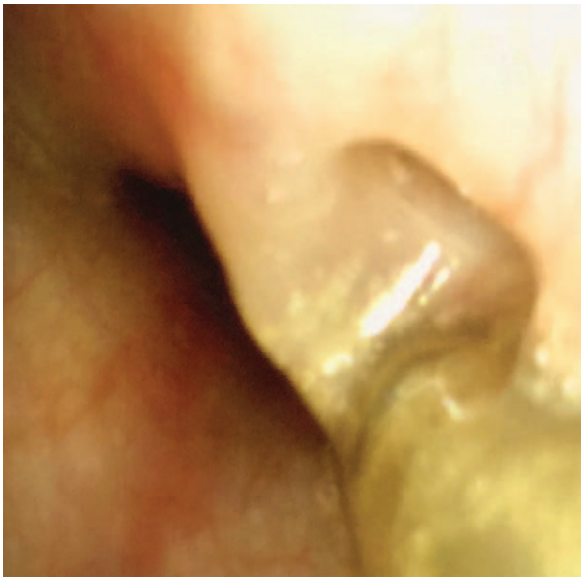

Figure 2. Flexible fiberoptic laryngoscopy revealed laryngeal atresia at the vocal cord plane, and a small hole-like defect was detected in the upper third of the esophagus.

3. Discussion

CHAOS is a rare life-threatening condition characterized by complete or near-complete intrinsic developmental obstruction of the fetal airway and it is usually fatal. This clinical condition was brought to notice firstly by Hedrick in the late 1900s.[5] He reported 4 cases of upper airway obstruction, which exhibited similar findings, including large echogenic lungs, flattened or inverted diaphragms, dilated airways distal to the obstruction, and fetal ascites and/or hydrops, and termed it CHAOS. According to Smith and Bain's classification of laryngeal atresia,[6] there are 3 types of pathology. Type I: complete atresia of the larynx with midline fusion of arytenoid cartilages and intrinsic muscles; type II: infraglottic obstruction where the dome-shaped cricoid cartilage obstructs the lumen; and type III: occlusion of anterior fibrous membranes and fusion of arytenoid cartilages at the level of the vocal process.[7,8] Nowadays, workers such as Hartnick et al[9] have described 3 types according to presentation:

(1) complete laryngeal atresia without an esophageal fistula;
(2) complete laryngeal atresia with a tracheoesophageal fistula; and
(3) near-complete high upper airway obstruction.

Successful treatment of CHAOS requires knowledge of its embryological origin. Although many significant studies have been conducted in recent years, the precise cause is not known. Now, the classic theory claims that the primitive digestive tube (PDT) emerges from the primitive endoderm and subsequently gives rise to the esophagus and trachea. The mesenchymal septum is formed in the coronal plane of the PDT, separating the trachea ventrally and the esophagus dorsally from the distal to the proximal ends of PDT. A failure in this process results in tracheoesophageal malformation such as esophageal atresia, tracheal atresia or tracheoesophageal fistula.[10,11] Then, at about 10-week gestation, an epithelial lamina temporarily occludes the upper airway, and failure of this membrane to recanalize results in laryngeal atresia. In a healthy fetus, fluid secreted by the lungs is absorbed through the tracheobronchial tree. In case of obstruction of the tracheobronchial tree, such as in laryngeal atresia, the lung fluid cannot be cleared, which leads to enlargement of the lungs because of intratracheal pressure by the accumulation of fetal lung fluid. This is just the beginning. The heart appears small compared with the enlarged lungs and positioned towards the midline of the thorax due to compression by the lungs. The diaphragm flattens or inverts according to the severity of the process, and mediastinal shift with decreased venous return and disturbance of cardiac function eventually leads to heart failure and hydrops.[12,13] That is why CHAOS always has similar clinical manifestations.

CHAOS can be diagnosed as early as 15-week gestation on transvaginal ultrasound, before ascites development. However, most cases are recognized in utero because of significant technical improvements in prenatal imaging such as sonography and magnetic resonant imaging (MRI).[14] The main diagnostic tool for prenatal diagnosis of CHAOS is sonography, which has typical findings on evaluation. It shows large echogenic lungs that flatten or invert the diaphragm, and small, compressed, centrally replaced heart due to compression by the lungs. Ascites and nonimmune hydrops are always present in ultrasound images. However prenatal identification of the level of atresia is difficult. Nowadays, MRI is popular for the diagnosis of fetal disease due to its absence of radiation and can be performed when any fetal

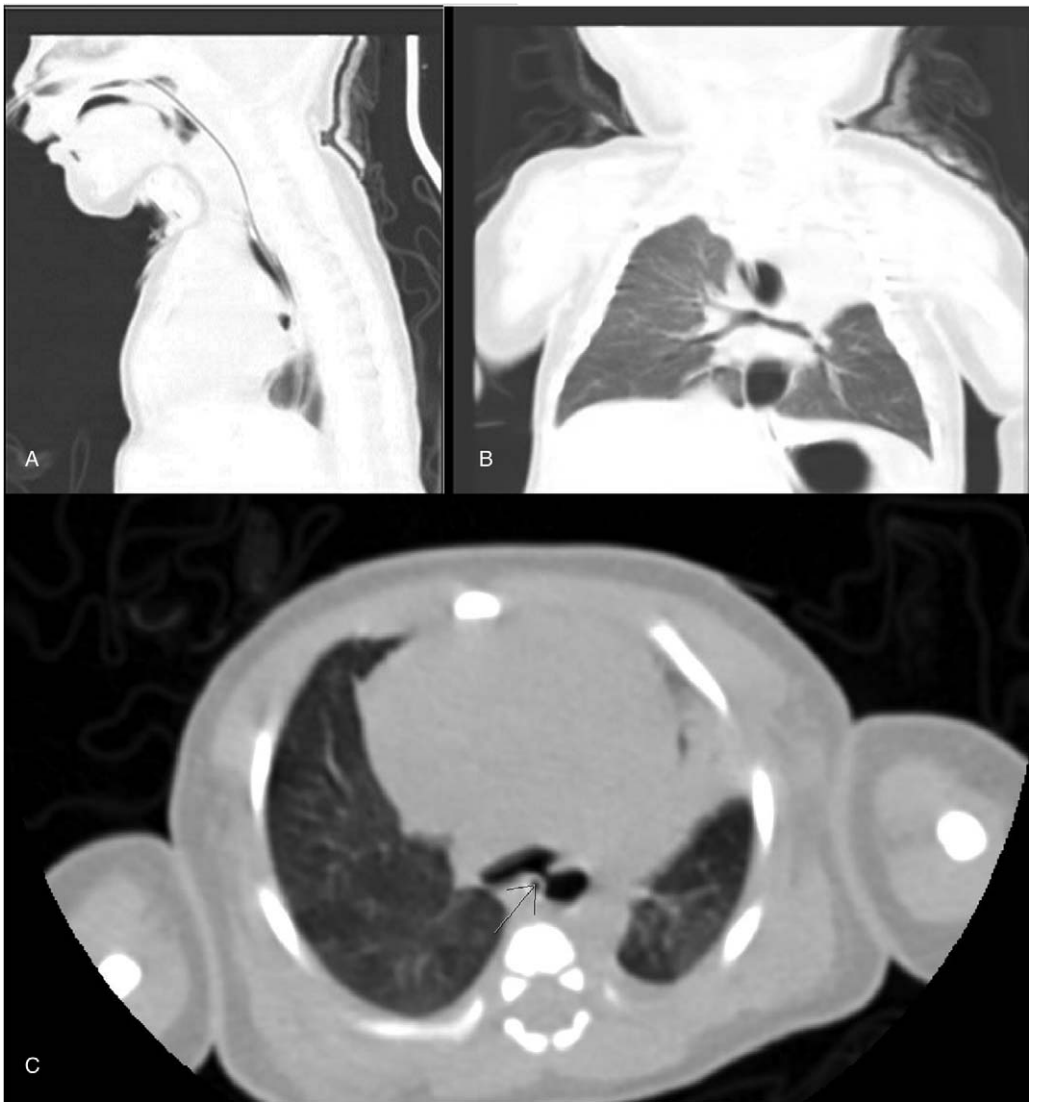

Figure 3. Contrast CT: sagittal position (A) and coronary position (B) revealed the main bronchi were absent; cross-section (C) revealed a 5-mm deep and 3-mm wide air-filled pouch in the midline in the bifurcation, at the level of the tracheal juga, extending caudally between the esophagus and the trachea. CT = computed tomography.

surgery or intervention is planned to establish the level of obstruction or to exclude extrinsic causes of obstruction.[15] It is better at identifying the level of obstruction due to higher intrinsic soft tissue contrast.[14,16]

CHAOS may not be harmful for the fetus till delivery, but it may lead to neonatal death soon after birth. So, following multiple discussions and counseling sessions with the patient elective termination of pregnancy is always suggested and undertaken. However, it has recently become possible to bypass the airway obstruction and establish adequate ventilation while the fetus is still connected to the placenta. The most developed intrapartum fetal therapeutic procedure is called ex utero intrapartum treatment (EXIT).[17] De Cou et al[18] successfully used EXIT to treat a case of CHAOS in 1988 due to laryngeal atresia. After that, several successful cases have been reported. However, it is not an easy way to deal with multiple complications during the procedure. Several most important aspects of EXIT can be summarized. First of all, it is important to have a skillful anesthesiologist with a specific anesthetic technique to obtain significant uterine relaxation and fetal anesthesia, while maintaining adequate placental perfusion. Secondly, it is significant to maintain fetal airway securement with different laryngologic procedures.

In summary, of course, early prenatal diagnosis of patients with CHAOS is necessary so that management can be undertaken in a planned way in a subset of patients. But in some remote areas, prenatal diagnosis is not so developed. When a neonate appeared to be difficult with inspiratory stridor after delivery, an immediate intubation even a definitive tracheostomy should be performed as soon as possible. When the patient was stabilized afterward, contrast CT and fiberoptic laryngoscopy were helped to make a definitive diagnosis. It is required a multidisciplinary team including otolaryngologists, radiologists, obstetricians, and anesthesiologists during the whole diagnosis and treatment process.

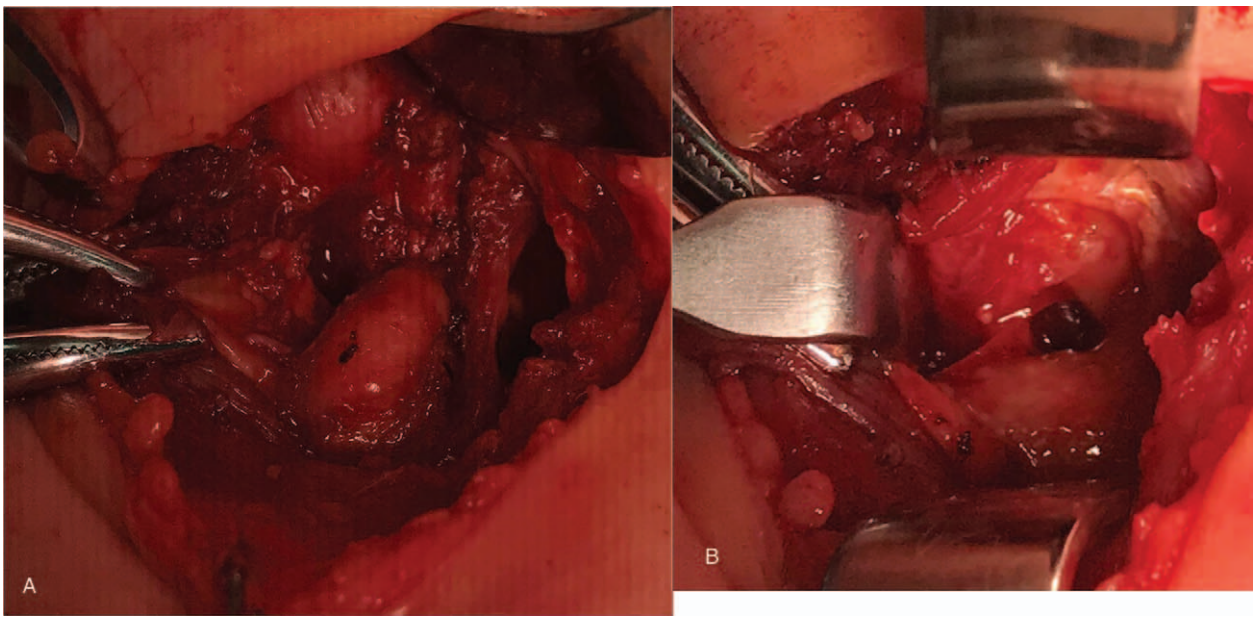

Figure 4. Autopsy revealed laryngeal atresia.

Author contributions

Data curation: Ying Wang.
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Investigation: Limin Zhao.
Writing – review & editing: Xiaoyan Li.

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