Prenatal Diagnosis of Congenital High Airway Obstruction Syndrome due to Laryngeal Atresia

Reddy Ravikanth*, Vaijnath P. Khanapure

Department of Radiology, St. John's Hospital, Kattappana, Kerala, India

Dear Editor,

A 28-year-old primigravida was referred for routine second-trimester anomaly scan at 20 weeks of gestation. Ultrasonography revealed bilateral echogenic and enlarged lungs with inversion of the diaphragm [Figure 1a]. The heart that was centrally placed appeared to be compressed by the enlarged lungs. There was associated dilatation of the principal bronchi [Figure 1b]. There was no pleural effusion or ascites. There was an associated lumbar meningocele [Figure 1c]. Differential diagnosis considered was bilateral congenital cystic adenomatoid malformation type III (CCAM III) and congenital high airway obstruction syndrome (CHAOS). Detection of obstruction site with distal airway dilatation helped in arriving at a diagnosis of CHAOS. Systemic arterial supply noted in CCAM III is also a differentiating factor. Based on the findings on ultrasonography, a diagnosis of CHAOS was made and the parents were offered counseling regarding the poor prognosis of the condition. Amniocentesis revealed a normal male karyotype. After termination of pregnancy, the postmortem examination findings such as significantly enlarged lungs were compatible with laryngeal atresia as a cause of CHAOS [Figure 2]. The larynx showed complete obstruction at the infraglottic level caused by the overgrowth of the cricoid cartilage. Neither tracheoesophageal fistula nor esophageal atresia was observed. Autopsy findings were compatible with laryngeal atresia type II. CHAOS is partial or complete obstruction of high airway in fetuses caused by stenosis or atresia involving trachea or larynx. Prenatal diagnosis is important as the condition is incompatible with life. CHAOS was first described by Hedrick in the late 1900s.[1] Exact incidence of CHAOS is not known and is a sporadic condition. Genetic syndromes associated with CHAOS are short-rib polydactyly syndrome, Fraser's syndrome, VACTERL association, and Shprintzen–Goldberg omphalocele syndrome.[2] Pathophysiology of CHAOS is outflow obstruction of the fetal lung fluid due to laryngeal or tracheal atresia, leading to pulmonary hyperplasia. Other extrinsic causes of tracheobronchial obstruction such as lymphatic malformations and vascular rings such as double aortic arch are to be excluded. Laryngeal atresia is a fatal congenital malformation and leads to tracheal dilatation due to nonclearance of the fluid in

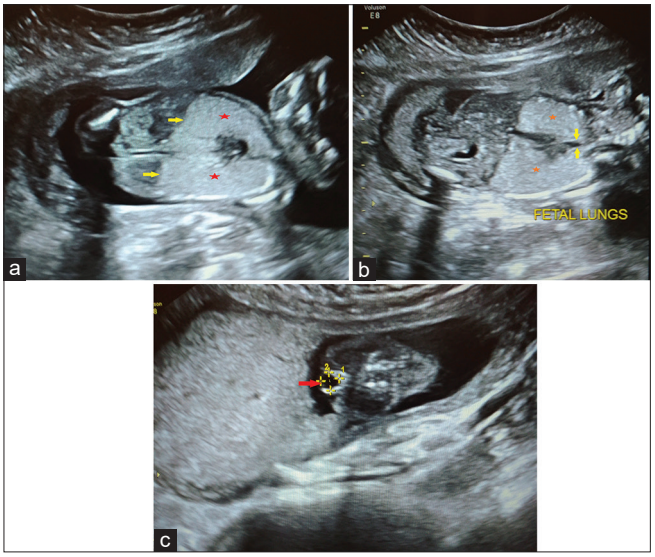

Figure 1: (a) Longitudinal ultrasonography image demonstrating bilateral enlarged hyperechoic lungs (star) with inversion of hemidiaphragm (arrow). (b) Longitudinal ultrasonography image demonstrating midline position of the heart due to compression by enlarged lungs (star) and dilated trachea secondary to laryngeal atresia (arrow). (c) Transverse ultrasonography image of the fetus with congenital high airway obstruction syndrome demonstrating an associated anomaly-lumbar meningocele (arrow)

Address for correspondence: Dr. Reddy Ravikanth, Department of Radiology, St. John's Hospital, Kattappana - 685 515, Kerala, India.
E-mail: ravikanthreddy06@gmail.com

Received: 15-01-2020 **Revised:** 02-03-2020 **Accepted:** 02-06-2020 **Available Online:** 14-07-2020

Access this article online

Quick Response Code:

Website: www.jmuonline.org

DOI: 10.4103/JMU.JMU_5_20

How to cite this article: Ravikanth R, Khanapure VP. Prenatal diagnosis of congenital high airway obstruction syndrome due to laryngeal atresia. J Med Ultrasound 2021;29:298-9.

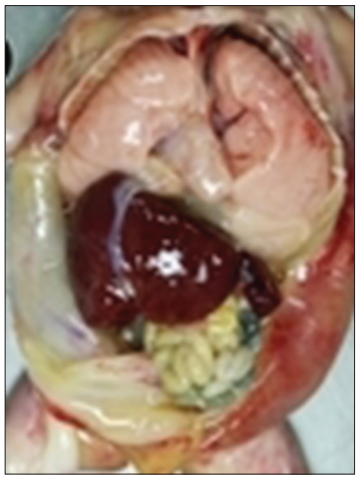

Figure 2: Autopsy image of the fetus at 20 weeks demonstrating voluminous lungs and dilated airway secondary to laryngeal atresia

CHAOS. Smith and Bain classified laryngeal atresia into three types.[3] Type I is complete laryngeal atresia with the fusion of arytenoids cartilages in the midline. Type II is characterized by a dome-shaped cricoid cartilage causing obstruction in the infraglottic location. Type III is characterized by fused arytenoids cartilages and anterior fibrous membrane, causing obstruction at the level of vocal processes. Typical prenatal sonological findings of CHAOS include bilateral enlarged hyperechoic lungs causing flattening or inversion of the diaphragm and dilated airways. Heart is positioned toward the midline due to compression by the enlarged lungs. Bilateral CCAM Type III is a fetal lung condition that closely mimics CHAOS on imaging. CCAM is a non airway obstructing lesion usually lobar and mostly left sided. CCAM does not involve the entire lung, hence will show a compressed lung rim. However, this feature is not seen in CHAOS. Pulmonary sequestration appears as an echogenic, homogenous lung mass with 90% incidence in the left hemithorax. Detection of a systemic artery from the aorta to the fetal lung lesion is a pathognomonic feature of fetal pulmonary sequestration. Bilaterality in CCAM and pulmonary sequestration is very rare. CHAOS may be associated with nonimmune fetal hydrops and fetal ascites.[4] With latest technical improvements in antenatal imaging, more number of cases are being detected nowadays and the management options are being developed to improve outcomes. CHAOS can be diagnosed as early as 15th week of gestation on transvaginal ultrasound, before ascites develop. In the past, CHAOS was equivalent to certain fetal death. Nowadays, if CHAOS due to incomplete obstruction is diagnosed in the late second or in the third trimester and if severe hydrops has not occurred yet, *ex utero* intrapartum treatment (EXIT) procedure can be offered. Timely detection of CHAOS during the antenatal period is important, especially if intervention is being considered, and a controlled near-term cesarean section with a well-planned EXIT intervention may improve the chances of neonatal survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Roybal JL, Liechty KW, Hedrick HL, Bebbington MW, Johnson MP, Coleman BG, *et al*. Predicting the severity of congenital high airway obstruction syndrome. J Pediatr Surg 2010;45:1633-9.
2. Garg MK. Case report: Antenatal diagnosis of congenital high airway obstruction syndrome-laryngeal atresia. Indian J Radiol Imaging 2008;18:350-1.
3. Smith II, Bain AD. Congenital atresia of the larynx. A report of nine cases. Ann Otol Rhinol Laryngol 1965;74:338-49.
4. Kuwashima S, Kitajima K, Kaji Y, Watanabe H, Watabe Y, Suzumura H. MR imaging appearance of laryngeal atresia (congenital high airway obstruction syndrome): Unique course in a fetus. Pediatr Radiol 2008;38:344-7.