An Antenatal Diagnosis: Congenital High Airway Obstruction

Abstract

Congenital high airway obstruction (CHAOS) is a rare lethal fetal malformation characterised by obstruction to the fetal upper airway, which can be partial or complete. Antenatal diagnosis of CHAOS is important due to recent management options. Diagnosis is made with secondary changes such as hyperechoic enlarged lungs resulting in mediastinal compression, ascites, hydrops, flattened or everted diaphragms and dilated distal airways. We reported a case of CHAOS, antenatally on ultrasonography (USG) at 20 weeks of gestation.

Keywords: *Ascites, atresia, congenital high airway obstruction syndrome, echogenic lung*

Miital S, Mittal A, Singal R, Singal S[1], Sekhon G

Departments of Radiodiagnosis and Imaging and [1]Surgery, M.M. Institute of Medical Sciences and Research, Mullana, Haryana, India

Introduction

Congenital high airway obstruction syndrome (CHAOS) is defined as complete or partial obstruction of fetal upper airways due to either intrinsic atresia or extrinsic compression. Fetal ascites and nonimmune hydrops may also be seen due to cardiac compression and obstructed venous return.[1]

Case Report

A 28-year-old primigravida was referred for a routine second trimester antenatal ultrasound at 20-week gestational age. There was no history of consanguinity and the family history was unremarkable. Ultrasound examination revealed that the fetus had bilateral large echogenic lungs [Figure 1]. The leaflets of diaphragm were inverted. The fetus had hydrops as thickened subcutaneous fat and ascites [Figure 2]. The heart was centrally placed and seemed to be compressed by the enlarged lungs [Figure 3]. Amniotic fluid was normal. No additional anomaly could be demonstrated. Based on ultrasound findings, diagnosis of CHAOS was made. The parents were counseled regarding the relatively poor prognosis of the syndrome, and the pregnancy was terminated after the consent of the patient and the family members. On autopsy, findings were of laryngeal atresia.

Discussion

CHAOS can be diagnosed as early as 15th week of gestation on transvaginal ultrasound, before ascites develop.[2] Magnetic resonant imaging (MRI) can be done when any fetal surgery or intervention is planned to know the level of obstruction or to exclude extrinsic cause of obstruction. Ultrasonography findings of voluminous lungs, inverted diaphragms, compressed centrally placed heart can also be appreciated on MRI.[1]

CHAOS is difficult to differentiate from congenital cystic adenomatoid malformation, especially type III and pulmonary sequestration, which is a rare condition.[3] Recently, CHAOS was proposed to be a part of tracheal agenesis, congenital cardiac abnormalities, radial ray defects, and duodenal atresia. Other congenital abnormalities such as gastroschisis or omphalocele should also be ruled out.[4] Exit procedure involves partial delivery of fetus (fetal head and

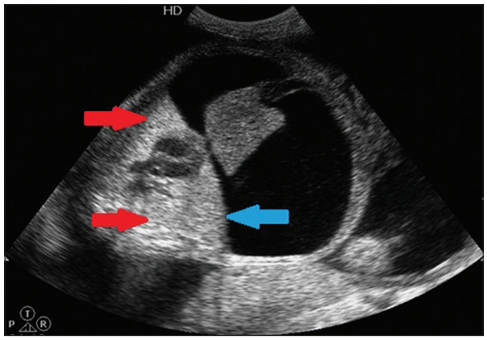

Figure 1: Coronal section through the fetal chest and abdomen revealed bilateral echogenic lungs (marked with red arrows) and flattened diaphragm (marked with blue arrow)

Address for correspondence:
Prof. Singal R,
Department of Surgery,
M.M. Institute of Medical Sciences and Research,
Mullana - 133 203,
Haryana, India.
E-mail: singalrikki@yahoo.com

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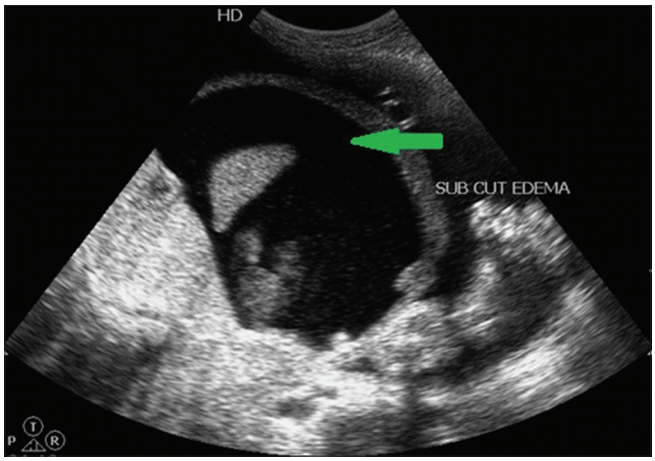

Figure 2: Antenatal ultrasound showing massive ascites (marked with arrow) and subcutaneous edema

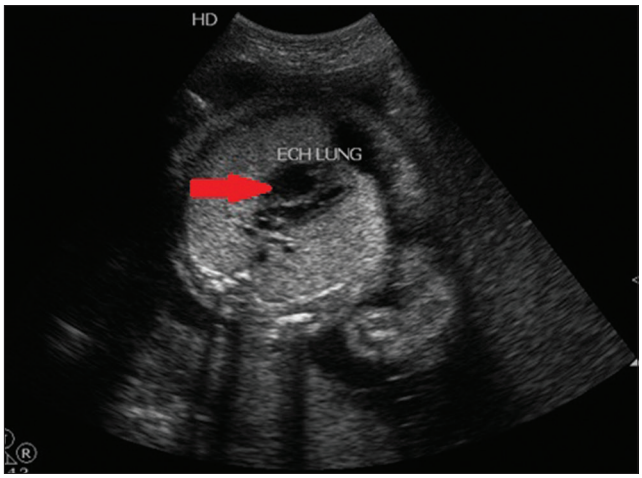

Figure 3: Axial section of ultrasound image of fetal chest showing hypervoluminous echogenic lungs with centrally placed compressed heart (marked with arrow)

chest) while placenta and umbilical cord remains intact to maintain uteroplacental circulation.[3]

Conclusion

Sonography is the modality of choice to diagnose CHAOS with specific characteristic findings, but MRI is superior in providing additional information of level of obstruction when planning for fetal surgery or intervention.

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Conflicts of interest

There are no conflicts of interest.

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