10.5005/jp-journals-10016-1137

CASE REPORT

Congenital High Airway Obstruction Syndrome presenting as Nonimmune Hydrops in a 19-week Fetus

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ABSTRACT

A 27-year-old primigravida at 19 weeks of gestation with suspected hydrops fetalis. There were characteristic ultrasonographic features like hyper inflated and enlarged lungs, flattened diaphragm, small centrally displaced heart, and hydrops suggestive of CHAOS. Congenital high airway obstruction syndrome is a rare fetal anomaly causing nonimmune hydrops. Prenatal ultrasound features are characteristic. Meticulous system wise fetal imaging in the presence of hydrops would help in early diagnosis of the condition, helping in appropriate management of pregnancy.

Keywords: Fetal ascitis, Flattened diaphragm, Hyper inflated enlarged lungs, Small centrally displaced heart, Subglottic laryngeal atresia.

How to cite this article: Mundkur A, Nayak SS, Vasudeva A, Katta GM, Kumar P. Congenital High Airway Obstruction Syndrome presenting as Nonimmune Hydrops in a 19-week Fetus. Int J Infertil Fetal Med 2016;7(3):99-102.

Source of support: The authors thank the Indian Council of Medical Research for financial support (BMS No. 54/5/2010).

Conflict of interest: None

Date of received: 14 March 2016

Date of acceptance: 29 June 2016

Date of publication: September 2016

INTRODUCTION

Congenital high airway obstruction syndrome (CHAOS) is a rare cause of fetal nonimmune hydrops (NIH). The true incidence of CHAOS is unknown.[1] High airway obstruction in the fetus could be due to laryngeal or tracheal atresia, laryngeal cysts, and tumors of the oropharynx or cervical region. It is a fatal condition having high mortality in spite of new therapeutic interventions. We report a case of CHAOS diagnosed prenatally, while evaluating a 19-week fetus with NIH. The characteristic sonological pictures suggest upper airway obstruction in the fetus.

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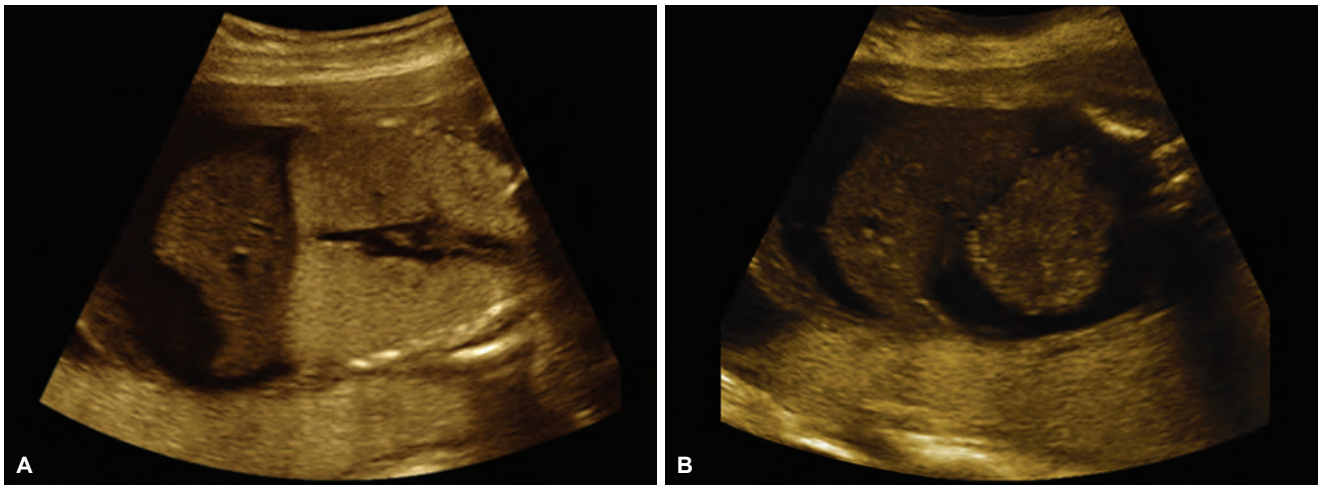
CASE REPORT

A 27-year-old primigravida at 19 weeks of gestation was referred to our tertiary referral center with suspected hydrops fetalis. She had nonconsanguineous marriage with no significant family history and had an uncomplicated antenatal period. She was Rhesus positive. Indirect Coombs test on the maternal blood was negative. Targeted ultrasonography confirmed the presence of fetal NIH as there was severe scalp edema (measuring 22 mm), generalized subcutaneous edema, along with fetal ascites. Fetal anemia was thought to be unlikely as middle cerebral artery peak systolic velocity was well below the 1.5 MoM. Detailed structural survey was carried out, to look for the causes of nonimmune hydrops.

Fetal echocardiography showed normal anatomy of the heart and normal flow patterns in great vessels. Cardiac function was also not compromised, as evidenced by good cardiac performance and normal Doppler patterns of ductus venosus. However, both lungs appeared enlarged, echogenic, occupying major portion of the thoracic cavity (Figs 1A and B), compressing heart to the middle of thorax. Hence, there was reduced cardiothoracic ratio, mesocardia, and a straightened cardiac axis. Diaphragm appeared flattened. No other structural anomalies were seen. The fetus was suspected to have upper airway obstruction. Differential diagnosis was given as congenital pulmonary airway malformation (CPAM), as cause of nonimmune hydrops.

Option of invasive prenatal testing was offered to the couple. They wished to undergo thorough investigations in order to have a definite diagnosis in this affected pregnancy. Cordocentesis was performed. Fetal hematocrit was normal. Karyotype revealed no abnormalities in fetus. There was no evidence of fetal parvovirus infection. Couple was counseled about the probable diagnosis of fetal upper airway obstruction with perinatal and prenatal interventions available in tertiary-level fetal medicine units. However, overall prognosis remains poor despite these interventions.

Couple opted for termination of pregnancy followed by fetal autopsy. The autopsy findings corroborated with



Figs 1A and B: (A) Enlarged, echogenic lungs occupying major portion of the thoracic cavity; and (B) compressing heart to the middle of thorax. Diaphragm appeared flattened

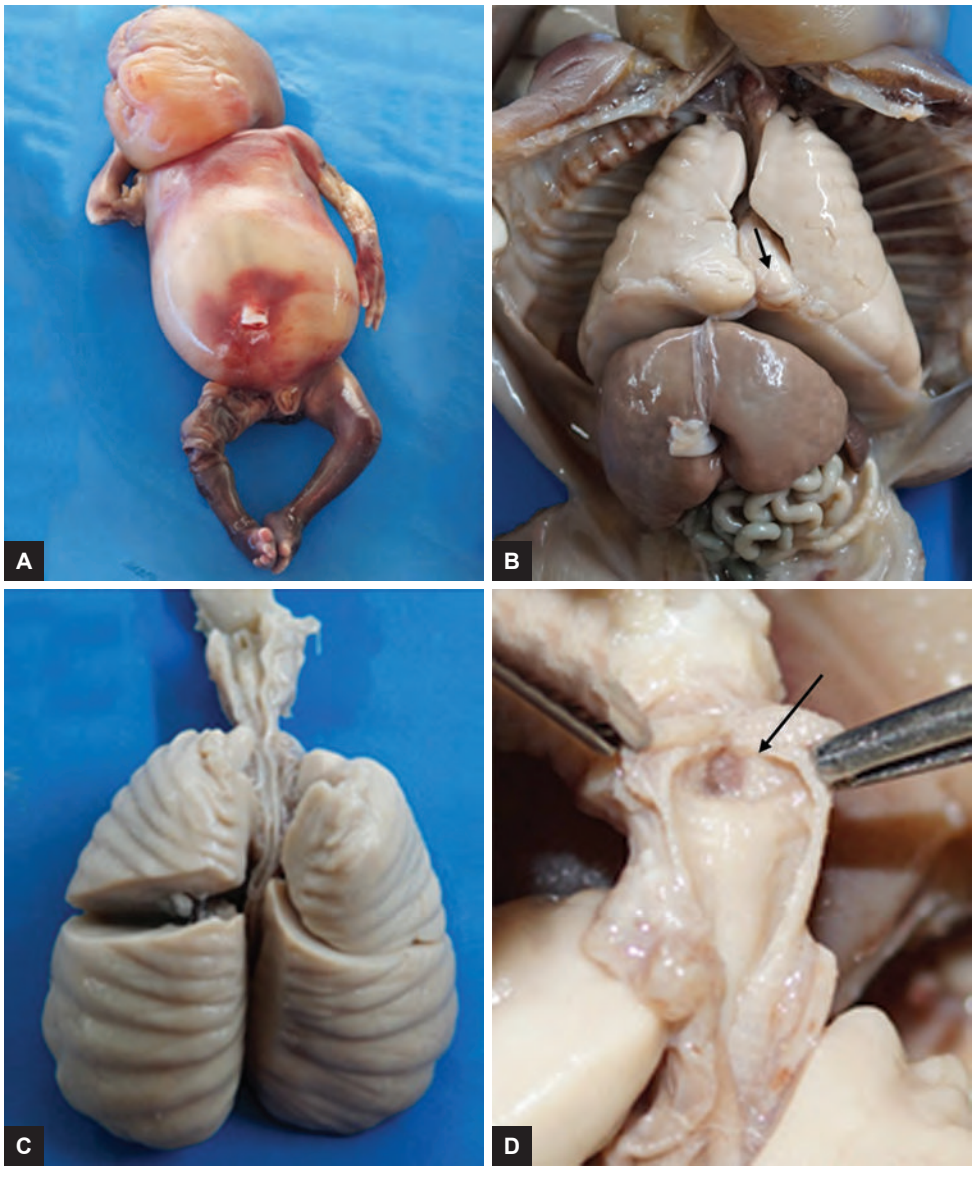
prenatal sonographic diagnosis. Male fetus weighed 763 gm and measured 27 cm in length. There was generalized severe subcutaneous edema of scalp, face, neck, and trunk. Facial features like eyes, ears, nose, and lips were compressed due to severe edema. Distension of thorax and abdomen was noted (Fig. 2A). Overriding of toes was noted on right foot. On internal examination, there was fluid in pleural and peritoneal cavities. Both lungs were bilobed, enlarged with costal impressions on external surface (Figs 2B and C). Right lung weighed 26 gm and measured 6.5 × 3.5 × 3 cm. Left lung weighed 30 gm and measured 7 × 3.5 × 2 cm. Subglottic laryngeal atresia was noted with mildly dilated trachea (Fig. 2D). Esophagus was normal. Heart was compressed and displaced to the midline due to enlarged lungs (Fig. 2B). Diaphragm was flattened. Rest of the anatomy was normal. The autopsy findings were suggestive of CHAOS due to laryngeal atresia. The study has the approval of Institutional Ethics Committee.

DISCUSSION

The condition congenital high airway obstruction was first described by Hedrick et al[2] in the year 1994 due to laryngeal atresia by the ultrasonographic findings of large echogenic lungs, flattened diaphragm, dilated airway distal to obstruction, and fetal ascites or hydrops.[3] Congenital high airway obstruction syndrome is a result of obstruction of the upper airway at various levels. It may be because of atresia or stenosis of larynx or trachea that appears to be the most common cause, laryngeal agenesis, cysts, or tumors of the cervical region.[4] Here we report a case of CHAOS due to laryngeal atresia at 19 weeks of gestation presented with hydrops. However, there have been reports of diagnosis of CHAOS as early as 15 weeks of pregnancy by transvaginal sonography.[5]

Congenital high airway obstruction syndrome takes place by deficient recanalization of the upper airways around the 10th week of gestation. The obstruction causes decreased clearance of fluid secreted by fetal lung, which lead to accumulation of fluid within the tracheobronchial tree, resulting in increased tracheal pressure and hyper inflated and enlarged lungs.[2] These enlarged lungs cause compression of the heart with central displacement. Reduced venous return and dysfunctional cardiovascular system end in ascites and hydrops. Enlarged lungs cause flattening of the diaphragm. Differentiating CHOAS from bilateral lung masses, such as a bilateral CPAM or other causes of extrinsic airway obstruction, such as a double aortic arch is crucial. In the present case there were characteristic ultrasonographic features like hyper inflated and enlarged lungs, flattened diaphragm, small centrally displaced heart, and hydrops suggestive of CHAOS.

Congenital high airway obstruction syndrome is associated with some genetic syndromes and chromosomal abnormalities. In our case there were no associated malformations and karyotype was normal. However, the prognosis of the condition is poor. There have been reports of exutero intrapartum treatment (EXIT) procedure.[6] This EXIT procedure is done in an attempt to create an intact upper airway, while maintaining placental circulation at cesarean delivery. Despite technical advancements in the field, overall neonatal prognosis remains poor, as the larynx and trachea are severely atretic.[7] Many attempts have been done to surgically correct the airway obstruction, while fetus is *in utero*. Overall prognosis remains poor again. However, fetal medicine experts have made attempts to prenatally diagnose milder forms of upper airway obstruction, like a web in the upper trachea with an intact larynx and vocal cords. In this case, a successful fetoscopic ablation of the tracheal web was performed,



Figs 2A to D: Clinical photographs of fetus show generalized severe subcutaneous edema of scalp, face, neck, and trunk: (A) Enlarged lungs with costal impression; (B, C) centrally displaced and compressed heart; [(B) arrow] and subglottic laryngeal atresia; and [(D) arrow]

lung pathology reverted back to normal, and intact child survival was reported.

In developing countries, the options of EXIT procedure or prenatal interventions are not available. Hence, till date, poor prognosis has to be conveyed to these couple. Therefore, it is important to diagnose the condition well before 20 weeks, so that medical termination of pregnancy can be offered in countries like India. In addition, this is one of the causes of nonimmune hydrops with low risk of recurrence, indicating good prognosis in future pregnancy. Hence, a complete workup would help in counseling the couple.

CONCLUSION

Congenital high airway obstruction syndrome is a rare fetal anomaly causing nonimmune hydrops. Prenatal ultrasound features are characteristic. Meticulous system-wise fetal imaging in the presence of hydrops would help in early diagnosis of the condition, helping in appropriate management of pregnancy.

ACKNOWLEDGMENT

The authors are grateful to the patient and family for consenting to participate in the study.

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