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Case Report

A rare case: Antenatally diagnosed congenital high airway obstruction syndrome

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ABSTRACT

Congenital high airway obstruction syndrome (CHAOS) is a rare life-threatening fetal condition resulting from obstruction of the upper fetal airway which may be partial or complete. Prenatal diagnosis is crucial as it usually results in stillbirth or death after delivery if unrecognized. We report a case of CHAOS that was diagnosed prenatally due to characteristic ultrasound features. We also briefly review literature in light of current management options.

Introduction

Congenital high airway obstruction syndrome (CHAOS) is defined as complete or partial obstruction of the fetal upper airways. This clinical condition was brought into notice firstly by Hedrick in the late 1900s [1]. CHAOS is usually caused by atresia or stenosis of the larynx or trachea. If the syndrome is unrecognized during the prenatal period, it usually results in stillbirth or death shortly after delivery [2]. Fortunately, more cases can be recognized in utero nowadays, as there are significant technical improvements in prenatal imaging. Bilaterally enlarged hyperechoic lungs, dilated airways, and flattened or inverted diaphragm are the typical prenatal sonographic findings. Fetal ascites and nonimmune hydrops may also be associated with the clinical condition. Due to the recently described management options, prenatal definition of fetal airway obstruction has come into prominence with the hope of neonatal outcome improvements [3]. We present a similar case in a 29 year old woman with 21 weeks gestational age.

Case

A 29-year-old woman with a previous two live birth was referred for a routine second trimester antenatal ultrasound at 21weeks gestational age. Ultrasound examination revealed

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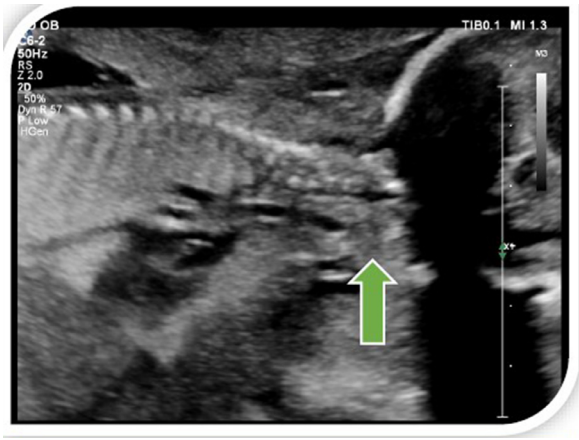

Fig. 1 – Dilatation of trachea is seen with abrupt cut-off at the laryngeal level (possible level of stenosis).

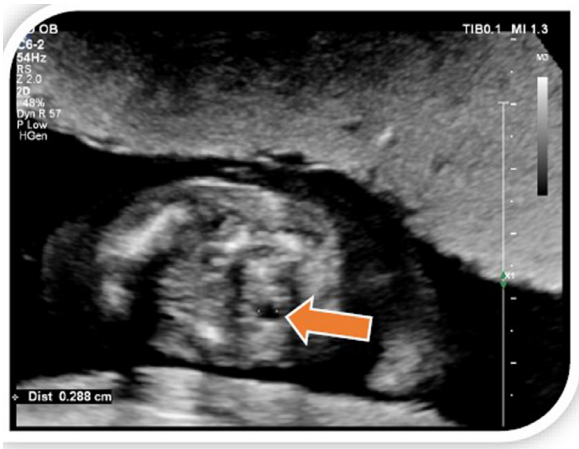

Fig. 2 – Trachea can be visualized just below the laryngeal level.

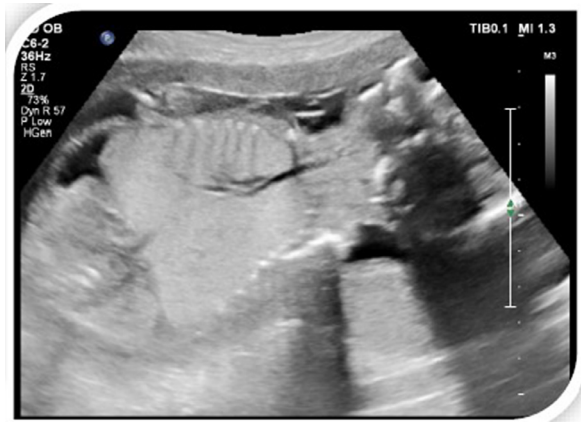

Fig. 3 – Coronal view chows enlarged lungs. inverted diaphragm can be seen.

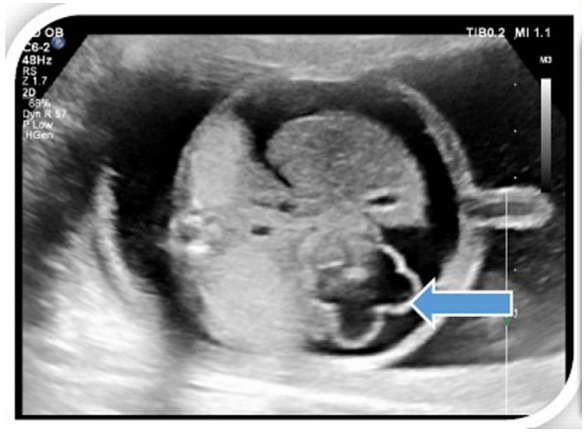

Fig. 4 – Axial view shows moderate fetal ascites and floating bowel loops. Gastric bubble can be appreciated (arrow).

that the fetus had bilateral large echogenic lungs. The principal bronchi appeared dilated (Fig. 1). No cystic lesions were seen in the lungs. The diaphragm was inverted. The heart was centrally placed and seemed to be compressed by the enlarged lungs, and there was moderate ascites. However, amniotic fluid was normal. The intra-cranial fetal structures, cerebellum, upper lip, spine, both kidneys, stomach bubble, urinary bladder, and the limbs were normal. The placenta was normal appearing. The femur length and bi-parietal diameter corresponded to 20 weeks of gestation while the abdominal circumference was corresponding to 25 weeks due to the presence of ascites. (Figs. 1-5)

Based on ultrasound findings, the diagnosis of CHAOS due to laryngeal atresia was made.

The parents were counselled regarding the relatively poor prognosis of the syndrome, however, they wanted to continue the pregnancy. The parents later visited after few weeks and the pregnancy was terminated because of still birth.

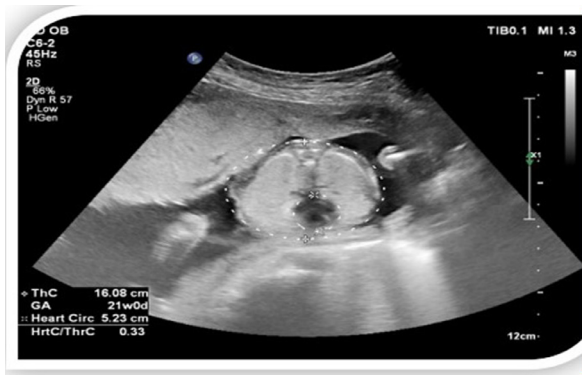

Fig. 5 – Axial image of fetal thorax shows bilateral enlarged echogenic lungs and centrally placed heart. The Cardio-thoracic ratio is decreased (0.33)

Discussion

Tracheal atresia is a very rare congenital malformation which takes place by deficient recanalization of the upper airways around the 10th week of gestation resulting in a clinical spectrum [1]. In a healthy fetus, the fluid secreted by fetal lung is absorbed through the tracheobronchial tree. However, in case of any obstruction in the tracheobronchial tree, this fluid is unable to be cleared.

The accumulation of the fetal lung fluid results in gradual increase of intratracheal pressure leading to enlargement of the lungs. It is the beginning of a chain reaction: the enlarged lungs cause compression of the heart and great veins. Due to the compression, the heart replaces centrally and becomes small and dysfunctional. Decreased venous return and dysfunctional cardiovascular system end in ascites and hydrops.

Three possible presentations include:

(a) complete laryngeal atresia without an oesophageal fistula,
(b) complete laryngeal atresia with a trachea-oesophageal fistula, and
(c) near-complete high upper airway obstruction

As a natural conclusion of the pathological process, bilateral large hyperechoic lungs, small, compressed, and centrally replaced heart, flattened or inverted diaphragm, and ascites are characteristic findings on sonographic examination [4, 5]. The diagnostic tool for prenatal diagnosis of CHAOS is ultrasound as it has typical findings on evaluation [6]. Regarding the amniotic fluid index, compression of the esophagus by dilated airways may lead to polyhydramnios, as the fetal swallowing of the fluid is disrupted. On the hand, impaired swallowing of the fetus may also cause oligohydramnios. The gestational age at the diagnosis may affect the amniotic fluid quantity. Polyhydramnios may not be present due to the examination in the early 2nd trimester in our cases.

CHAOS is a close differential to bilateral congenital cystic adenomatoid malformation (CCAM) [7]. CCAM (especially type III) and upper airway obstruction secondary to intrinsic causes such as tracheal or laryngeal atresia or stenosis and tracheal webs similarly have bilateral uniform hyperechogenic appearance of the fetal lungs on sonographic examination. In order to make a differentiation between CHAOS and sequestered lung, the obstruction site with distal airway dilatation (present in CHAOS) and the systemic arterial supply (present in sequestered lung) must be clearly seen. Congenital high airway obstruction syndrome should be also differentiated from extrinsic causes of trachea-laryngeal obstruction. Some of these extrinsic causes are lymphatic malformation, cervical teratoma, and vascular rings like double aortic arch.

CHAOS is mostly sporadic, and the exact incidence is not known [1,2]. In utero death of the affected cases; being a part of some genetic syndromes; better detection rate of the anomalies by the means of technical improvement of imaging tools; and data only from isolated cases instead of studies consisting of large series may explain this indefinable incidence. The most common associated genetic disorder with CHAOS is Fraser's syndrome which is inherited by autosomal recessive

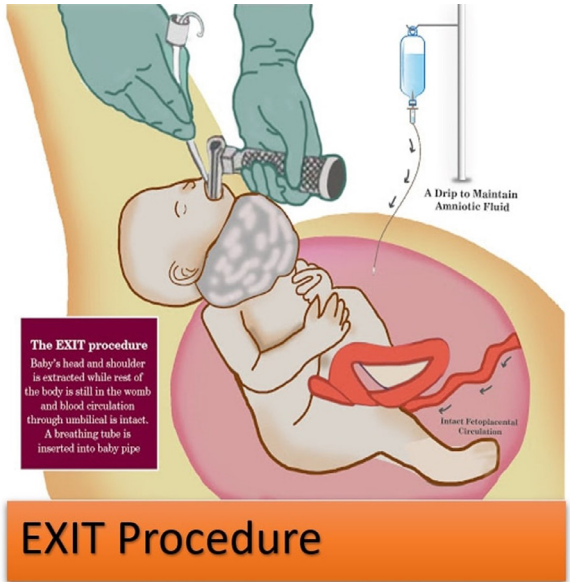

Fig. 6 – Exit procedure.

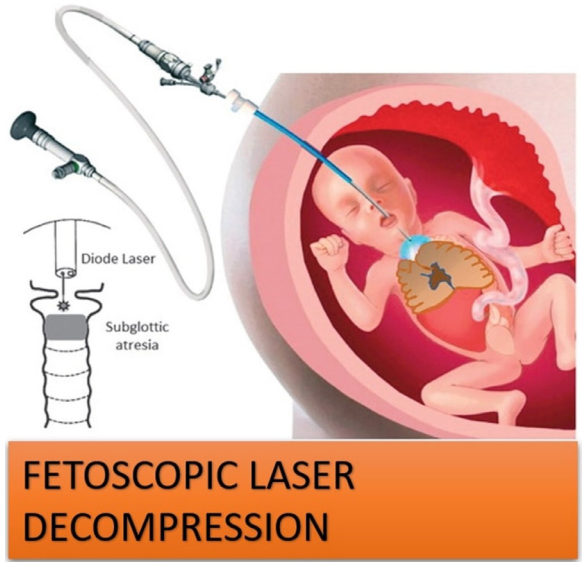

Fig. 7 – Fetoscopic laser decompression.

form and characterized by urogenital defects, laryngeal atresia, syndactyly, and cryptophthalmos [8,9]

It has been proposed that CHAOS may be a part of an association, which has been given the acronym TACRD (Tracheal Agenesis, complex Cardiac anomalies, Radial ray defects, and Duodenal atresia). This is distinct from the more common VACTERL (Vertebral, Anorectal, Cardiac, Tracheo-Esophageal, Renal, Limb anomalies) association which has trachea- oesophageal fistula and not tracheal agenesis as a component. Hence, a suspicion of CHAOS on imaging should prompt a search for associated anomalies too [8].

The use of fetal MRI allows exquisite definition of fetal anatomy and facilitates treatment planning as it more effectively depicts the dilated airways, the level of obstruction, its

relation to trachea, evaluate facial involvement and aids in exclusion of extrinsic pathologies leading to obstruction.

Antenatally diagnosed cases of CHAOS may be offered an EXIT (ex utero intrapartum treatment) procedure (Fig. 6) which consists of delivery of the fetal head and chest to secure an airway while simultaneously maintaining the uteroplacental circulation, with tracheostomy being necessary in most cases [10]. If the airway is not adequately visualized then rigid bronchoscopy is attempted. If an endotracheal tube is unable to be passed, tracheotomy is the final option. Cases of spontaneous antenatal improvement in CHAOS due to spontaneous perforation also suggest that intrauterine fetoscopic laser laryngotomy (Fig. 7) may be beneficial in a small subset of these patients [8,10].

Conclusion

CHAOS is a rare and fatal cause of congenital airway obstruction if unrecognized during prenatal period. Antenatal sonographic imaging shows typical findings which can lead to a diagnosis. MRI is superior to sonography in demonstrating the level of obstruction and in assisting in the differential diagnosis by excluding extrinsic causes of obstruction can be offered.

Patient consent

Written and informed consent was obtained from the patient for publication of this case report and any accompanying images.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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 [Internet] 2010;45:1633–9 doi:. doi:10.1016/j.jpedsurg.2010.01.022.
[2] J S, Saket RK D, Singh P. Congenital High Airway Obstruction Syndrome (CHAOS) a rare fetal malformation diagnosed on antenatal ultrasonography. Int J Clin Med Imaging 2015;02:2176–8.
[3] Courtier J, Poder L, Wang ZJ, Westphalen AC, Yeh BM, Coakley F V. Fetal tracheolaryngeal airway obstruction: prenatal evaluation by sonography and MRI. Pediatr Radiol 2010;40:1800–5.
[4] Nolan HR, Gurria J, Peiro JL, Tabbah S, Diaz-Primera R, Polzin W, et al. Congenital high airway obstruction syndrome (CHAOS): natural history, prenatal management strategies, and outcomes at a single comprehensive fetal center. J Pediatr Surg [Internet]. 2019;54:1153–8 doi:. doi:10.1016/j.jpedsurg.2019.02.034.
[5] Afzal S, Fatima K, Ambareen M. Antenatal ultrasound diagnosis of congenital high airway obstruction syndrome: a case report and review of literature. Cureus 2019;11.
[6] Kadakola B, PS S, Jain S. Antenatal ultrasonographic diagnosis of congenital high airway obstruction syndrome - a chaotic state: a case report. Int J Reprod Contraception, Obstet Gynecol. 2015;4:1650–2.
[7] SJ K, DW P, S W. Fetal echogenic lung lesions: Prenatal ultrasound diagnosis and outcome. Pediatr Radiol 1995;25:208–10.
[8] Joshi P, Satija L, George RA, Chatterjee S, D'Souza J, Raheem A. Congenital high airway obstruction syndrome-antenatal diagnosis of a rare case of airway obstruction using multimodality imaging. Med J Armed Forces India [Internet]. 2012;68:78–80 doi:. doi:10.1016/S0377-1237(11)60111-1.
[9] Lim FY, Crombleholme TM, Hedrick HL, Flake AW, Johnson MP, Howell LJ, et al. Congenital high airway obstruction syndrome: Natural history and management. J Pediatr Surg 2003;38:940–5.
[10] Guimaraes CVA, Linam LE, Kline-Fath BM, Donnelly LF, Calvo-Garcia MA, Rubio EI, et al. Prenatal MRI findings of fetuses with congenital high airway obstruction sequence. Korean J Radiol 2009;10:129–34.