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Case report: Antenatal diagnosis of congenital high airway obstruction syndrome – laryngeal atresia

Mukesh Kumar Garg
Department of Radiology, Geetanjali Medical College and Hospital, Udaipur, (Rajasthan), India

Correspondence: Dr Mukesh Kumar Garg, 68, Sukhadia Nagar, University Road, Udaipur-313 001, Rajasthan, India.
E-mail: drmukesh.garg@rediff.com

Abstract

Congenital high airway obstruction syndrome (CHAOS) is a near fatal condition of multifactorial inheritence, in which the fetus has a dilated trachea, enlarged echogenic lungs, an inverted or flattened diaphragm, and ascites. A case of CHAOS, diagnosed antenatally on USG at 28 weeks of gestation, is being reported here.

Key words: Congenital high airway obstruction syndrome; laryngeal atresia

Congenital high airway obstruction syndrome (CHAOS) is a condition in which the fetus has hyperinflated, enlarged, and highly echogenic lungs; an inverted or flattened diaphragm; a dilated tracheobronchial tree; and ascites. It occurs as a result of congenital obstruction of the fetal airway secondary to laryngeal atresia, tracheal atresia, or a laryngeal cyst.[1–2] The disease is generally incompatible with life and, therefore, antenatal USG diagnosis is desirable. I would like to report a case where antenatal diagnosis was possible on USG at 28 weeks' gestation.

Case Report

A 20-year-old multiparous (gravida 2) woman at 28 weeks' gestation was referred for a fetal well-being examination. There was no history of consanguinity and the family history was unremarkable. Her previous pregnancy had been uneventful.

USG showed a dilated trachea [Figure 1], enlarged hyperechoic lungs, inferiorly displaced and flattened diaphragms [Figures 1 and 2], minimal fetal ascites, excessive amniotic fluid volume (amniotic fluid index: 20 cm), and a small heart because of compression by the obstructed lungs [Figure 2]. These findings were diagnostic of CHAOS. We discussed the possible unfavorable outcome of the pregnancy with the parents who chose not to terminate the pregnancy because of religious reasons.

Discussion

Laryngeal atresia is a rare congenital malformation and is usually fatal. The malformation is caused by nondevelopment of the 6th branchial arch during normal embryological development.[3] Smith and Bain[3] have classified laryngeal atresia into three types: type 1, in which there is complete atresia of the larynx with midline fusion of the arytenoid cartilages and intrinsic muscles; type 2, in which there is infraglottic obstruction that is characterized by a dome-shaped cricoid cartilage obstructing the lumen; and type 3, in which there is occlusion of the anterior fibrous membrane and fusion of the arytenoid cartilages at the level of the vocal processes.[3]

Association of laryngeal atresia with partial trisomy 9 and 16, resulting in maternal translocation has also been reported.[4,5]

Antenatal USG shows enlarged hyperechoic lungs, a dilated tracheobronchial tree, ascites, and an inverted or flattened diaphragm. In laryngeal atresia, the trachea is dilated because of nonclearance of fluid (which is normally secreted by the lungs). In high airway obstruction, the nonclearance of fluid from the lungs results in parenchymal hyperplasia, which is apparent on USG as enlarged hyperechoic lungs; this condition was recognized by Dolkart et al.,[6] Morrison et al.,[7] and Liggins.[8] An enlarged lung causes compression of the great veins and the right atrium, and this leads to ascites.[7,9] Compression of the esophagus due to a dilated trachea results in polyhydramnios.[10]

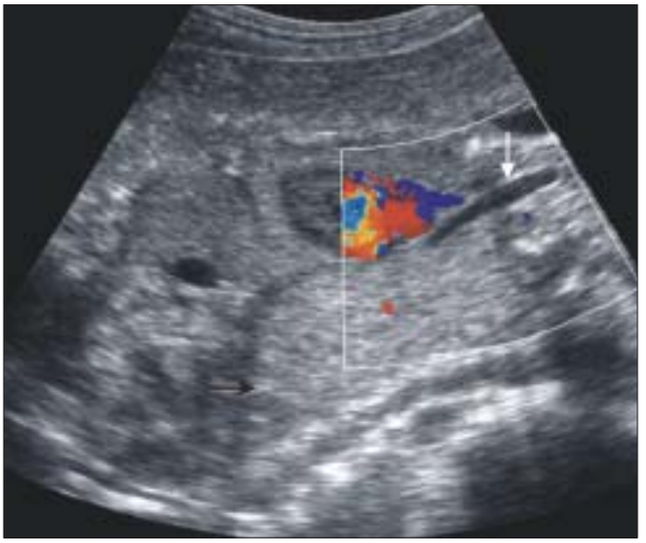

Figure 1: USG of the fetus in the coronal plane, at the level of the thorax shows a dilated trachea (white arrow). The black arrow points towards the flattened diaphragm

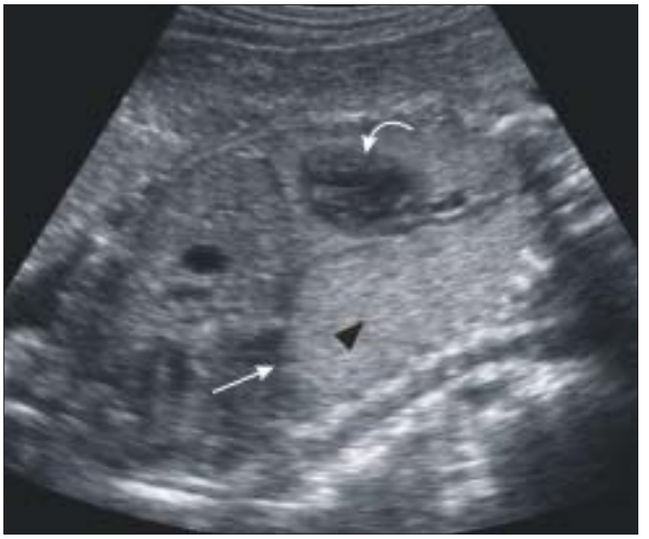

Figure 2: USG of the fetus in the coronal plane shows enlarged hyperechoic lungs (arrowhead) with flattened diaphragms (arrow) and a small heart (curved arrow)

This malformation is generally fatal; however, there are reports of a few cases that have been successfully treated with neonatal interventions such as ex utero intrapartum treatment (EXIT).[11–12]

References

1. Lim FY, Crombleholme TM, Hedric 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.
2. Liechry KW, Cromblehome TM. Management of fetal airway obstruction. Semin Perinatol 1999;23:496-506.
3. Smith II, Bain AD. Congenital atresia of the larynx: A report of nine cases. Ann Otol Rhinol Laryngol 1965;74:338-49.
4. Baker DC Jr, Savetsky L. Congenital partial atresia of the larynx. Laryngoscope 1966; 76:616-20.
5. Van den Boogaard MJ, De Pater J, Hennekam RC. A case with laryngeal atresia and partial trisomy 9 due to maternal 9;16 translocation. Genet Couns 1991;2:83-91.
6. Dolkart LA, Reimers FT, Wertheimer IS, Wilson BO. Prenatal diagnosis of laryngeal atresia. J Ultrasound Med 1992;11:496-8.
7. Morrison PJ, Macphail S, Williams D, McCusker G, McKeever P, Wright C, *et al*. Laryngeal atresia or stenosis presenting as second-trimester fetal ascites: Diagnosis and pathology in three independent cases. Prenat Diagn 1998;18:963-7.
8. Liggins GC. Growth of the fetal lung. J Dev Physiol 1984;6:237-48.
9. Minior VK, Gagner JP, Landi K, Stephenson C, Greco MA, Monteagudo A. Congenital laryngeal atresia associated with partial diaphragmatic obliteration. J Ultrasound Med 2004;23:291-6.
10. Onderoglu L, Saygan Karamursel B, Bulun A, Kale G, Tuncbilek E. Prenatal diagnosis of laryngeal atresia. Prenat Diagn 2003;23:277-80.
11. Crombleholme TM, Sylvester K, Flake AW, Adzick NS. Salvage of a fetus with congenital high airway obstruction syndrome by ex utero intrapartum treatment (EXIT) procedure. Fetal Diagn Ther 2000;15:280-2.
12. Kanamori Y, Kitano Y, Hashizume K, Sugiyama M, Tomonaga T, Takayasu H, *et al*. A case of laryngeal atresia (congenital high airway obstruction syndrome) with chromosome 5p deletion syndrome rescued by ex utero intrapartum treatment. J Pediatr Surg 2004;39:E25-8.

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