

CASE REPORT

ANTENATAL DIAGNOSIS OF CONGENITAL HIGH AIRWAY OBSTRUCTION SYNDROME (CHAOS)

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ABSTRACT

Abstract: congenital high airway obstruction syndrome is a near fatal condition of a multifactorial inheritance, in which the fetus has a dilated trachea, enlarged echogenic lungs, an inverted or flattened diaphragm and ascites. A case of CHAOS, diagnosed antenatally at 18 weeks of gestation is being reported here.

Keywords: Congenital high airway obstruction syndrome, laryngeal atresia

1.INDRODUCTION

CHAOS is a condition in which the fetus has hyperinflated, enlarged and highly echogenic lungs, an inverted or flattened diaphragm, dilated trachea bronchial 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. 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 18 weeks of gestation.

2.CASE REPORT

a 22 year old primi woman at 18 weeks of gestation was referred for a fetal well-being examination. There was no history of consanguinity and the family history was unremarkable.

USG showed a dilated trachea (figure 1) enlarged hyperechoic lungs (figure 2) inferiorly displaced and flattened diaphragm (figure 3) fetal ascites (figure 4) and excessive amniotic fluid index. These findings were diagnostic of CHAOS. We discussed the possible unfavourable outcome of the pregnancy with the parents.

3.DISCUSSION

laryngeal atresia is a rare congenital malformation and is usually fatal. The malformation is caused by non-

development of 6thbranchial arch during normal embryological development.

Smith and Bain (1) have classified laryngeal atresia into three types. 1. In which there is complete atresia of larynx with midline fusion of arytenoid cartilages and intrinsic muscles. 2. In which there is infraglottic obstruction that is characterized by a dome shaped cricoid cartilage obstructing the lumen. 3. In which there is occlusion of anterior fibrous membrane and fusion of arytenoid cartilages at the level of vocal process.

Association of laryngeal atresia with partial Trisomy 9 and 16, resulting in maternal translocation has also been reported (2,3).

Antenatal USG shows enlarged hyperechoic lungs, a dilated trachea-bronchial tree, ascites and an inverted or flattened diaphragm. In laryngeal atresia, the trachea is dilated because of non-clearance of fluid(which is normally secreted by lungs). In high airway obstruction, the non-clearance of fluid from lungs result in parenchymal hyperplasia, which is apparent on USG as enlarged hyperechoic lungs. This condition was recognized by Dolkat et al (4). An enlarged lung causes compression of great veins and right atrium and these leads to ascites(5,6). Compression of oesophagus due to dilated trachea results in polyhydramnios (7).

The malformation is generally fatal, however there are reports of few cases that have been successfully treated with neonatal interventions such as ex uterointrapartumtreatment(EXIT) (8,9).

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Figure 1: image showing dilated Trachea



Figure 3: image showing flattened diaphragm



Figure 1: image showing dilated Trachea



Figure 4: image showing fetal ascites

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