Antenatal ultrasonography depicting congenital high airway obstruction syndrome with duodenal atresia

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A 25-year-old multigravida was referred to Radiology Department for routine second trimester obstetric ultrasound examination. It was spontaneous conception from a non-consanguineous marriage. Ultrasound examination showed a single live foetus with parameters corresponding to 24 weeks of gestation. Bilateral lungs were enlarged and echogenic with inversion of diaphragm. Dilated trachea and bronchi were noted with heart displaced in midline. There was presence of mild polyhydramnios. There was presence of double bubble sign in the abdomen indicating duodenal atresia. No other foetal anomalies were seen. The diagnosis of congenital high airway obstruction syndrome (CHAOS) was made on antenatal ultrasound (Figure 1). She had an intrauterine foetal demise at 25 weeks. Labour was induced and she delivered a dead female foetus weighing 800 g. Foetal autopsy revealed presence of CHAOS and duodenal atresia.

CHAOS was first described in 1994 by Hedrick et al. [1]. It is a rare entity, defined as partial or complete obstruction of upper airways in the foetus. It can occur in isolation or in association with other syndromes. Causes include laryngeal or tracheal atresia. Irrespective of the cause, clinical and radiological presentation is the same. Obstruction of the high airway leads to the accumulation of fluid in the lungs. Characteristic sonographic features include enlarged bilateral echogenic lungs, dilated lower airway, everted/flattened diaphragm, ascites and polyhydramnios. Differential diagnosis of echogenic lung includes congenital cystic adenomatoid malformation (CCAM), however CCAM is generally unilateral, whereas CHAOS is invariably bilateral [2]. CHAOS is often associated with other foetal anomalies like duodenal atresia in our case. Only one case has been reported in the literature depicting CHAOS with duodenal atresia [3].

Sanford et al. [4] reviewed more than 100 cases of CHAOS and revealed that there is no sex predilection for CHAOS and it is reported equally in males and females. Ultrasound features become evident after 16 weeks of gestation. Laryngeal malformations were the most common cause of CHAOS in their review (74.6%), followed by tracheal malformations (18.6%) and other unspecified causes (6.8%). Anomalies associated with CHAOS include malformations of the digits and musculoskeletal system (22.0%),

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cardiovascular system (17.8%), genitourinary system (17.8%), gastrointestinal system (14.4%) and neurologic system (8.5%).

Previously, CHAOS was considered as lethal anomaly; however, recently survival is reported with ex utero intrapartum treatment procedure and postnatal correction. Kohl et al. [5] reported in utero decompression in foetus with CHAOS. Due to possible survival of these cases, our case has implications for paediatricians and paediatric surgeons. Whenever a case of CHAOS is diagnosed antenataly, the parents should be counselled whether the pregnancy should be continued or not because associated complicated anomalies are associated with very poor quality of life. To conclude, we have described a case of CHAOS and duodenal atresia detected in utero. CHAOS is associated with poor prognosis.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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None.

ETHICAL APPROVAL

Ethics clearance and approval of the study were granted by the ethics committee of our institute. Signed informed consent for participation and publication of medical details was also obtained from the parents. Confidentiality was ensured at all stages.

REFERENCES


Figure 1. Antenatal ultrasonography showing enlarged and echogenic bilateral lungs (L) with inversion of diaphragm (arrows in a). Double bubble sign is seen in the abdomen indicating duodenal atresia (D). Dilated trachea is also noted (arrow in b).