Case report:

Antenatal Diagnosis of Congenital High Airway Obstruction Syndrome

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Abstract:
Congenital high airway obstruction syndrome (CHAOS) is a near fetal condition of multifactorial inheritance & so need antenatal diagnosis (Biynam). The fetus show typical findings include a dilated, fluid filled trachea, enlarged hyper inflated echogenic lungs, inverted or flattened diaphragm, squeezed heart & ascites. It is the result of congenital obstruction of the airway secondary to laryngeal atresia or a laryngeal cyst (1,2).

Introduction:
Congenital high airway obstruction syndrome (CHAOS) is a rare condition which occurs as a result of congenital fetal airway obstruction which may be complete or partial. The disease is usually incompatible with life and leads to stillbirth or death shortly after delivery. Due to advances in antenatal imaging by ultrasound and magnetic resonance imaging (MRI), more cases are being detected in the antenatal period. CHAOS occurs secondary to atresia or stenosis of the larynx or trachea, which may be partial or complete or due to a laryngeal cyst.

Case report:
21 years old primi gravida came for routine sonography at 20 weeks to r/o anomalies. Her previous sonography was done at 9 weeks & unremarkable. There was no history of consanguinity. On examination lateral ventricles were dilated (width 17mm). No spinal or limb anomaly. Lungs were echogenic & over expanded with inverted domes of diaphragm & tubular compressed heart. Trachea and bronchi were filled with fluid. E/o ascites with cystic structures are seen in it. No other organs were identifiable except liver. Amniotic fluid was excess. Diagnosis of congenital high airway obstruction syndrome was made. After explanation of unfavorable outcome of pregnancy, the parents opted for termination of pregnancy. The aborted fetus showed distended thorax and abdomen. The abdominal cavity showed a large amount of clear yellow fluid. An intrathoracic examination showed the enlargement of the right and left lung. The larynx showed complete obstruction at infraglottic level caused by overgrowth of the cricoid cartilage. Autopsy findings were compatible with laryngeal atresia type II described by Smith and Bain in 1965. Due to non affordability of parents, karyotyping was not done. And due to reluctance of relatives block dissection was not possible.

Discussion:
Laryngeal atresia is rare congenital malformation and is usually fatal. Prenatal diagnosis of CHAOS was possible as early as 15 weeks of gestation (Y Gilboa). The malformation is caused by non development of...
6th brachial arch during normal embryological development(3). Smith and Bain have classified laryngeal atresia into 3 types: Type 1-in which there is complete atresia of the larynx with midline fusion of arytenoid cartilage 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 arytenoid cartilages at level of vocal processes. Many etiologies were proposed including laryngeal or tracheal webs, laryngeal cysts, tracheal stenosis, atresia, laryngeal or tracheal agenesis. However, laryngeal atresia appears to be the most frequent cause. 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)(4,5) fetoscopic laser decompression, while the fetus is still connected to the placenta (T kohl). Several successful cases have been reported. The EXIT procedure is a technique for safely managing airway obstruction at birth, in which placental support is maintained until the airway is evaluated and secured. The sonographic finding of CHAOS include increase in lung size and echogenicity, fluid filled, dilated trachea, fetal hydrops, and polyhydramnios. Antenatal USG shows enlarged hyperechoic lungs, a dilated tracheobronchial tree, Ascites, and an inverted or flattened diaphragm (Garg). In laryngeal atresia, the trachea is dilated because of non clearance of fluid (which is normally secreted by lungs). In high airway obstruction, non clearance of fluids from lungs results in parenchymal hyperplasia, which is apparent on USG as enlarged hyperechoic lungs(6,7,8). An enlarged lung causes compression of great veins and the right atrium and this leads to Ascites(7,9). Compression of oesophagus due to dilated trachea results in polyhydramnios(10). The findings were difficult to differential with congenital cystic adenomatoid malformation (CCAM) type III (Chemsai,Burcu). However, bilateral CCAM was very rare; therefore the diagnosis of CHAOS was considered. Laryngeal atresia may be associated with other structural and genetic abnormalities such as left persistent superior vena cava, single umbilical artery, abnormal fingers and toes, esophageal atresia, or renal agenesis. Association of laryngeal atresia with Partial trisomy 9 and 16 resulting in maternal translocation has also been reported(11,12). The most common associated genetic disorder with CHAOS is Fraser’s syndrome which is inherited by autosomal recessive form and characterized by urogenital defects, laryngeal atresia, syndactyly and cryptophthalmos (Joshi p).

**Conclusion:**

Congenital high airway obstruction syndrome is a rare cause of congenital airway obstruction which is incompatible with life. Antenatal imaging with ultrasound usually shows typical findings which can lead to a diagnosis. MRI has an adjunctive role in demonstrating level of obstruction and excluding extrinsic causes of obstruction.

**Reference:**


