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Case Report

A rare case of congenital high airway obstruction syndrome

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Abstract

Congenital high airway obstruction syndrome is one of the rare life threatening conditions and usually has lethal outcome. High airway obstruction can be at the level of larynx or trachea and is due to developmental absence or blockage of airway. We are here by presenting a very rare case of congenital high airway obstruction.

Key words: Congenital high airway obstruction syndrome (CHAOS), Laryngeal atresia, Ex-utero intrapartum treatment, Fetal MRI

1. Introduction

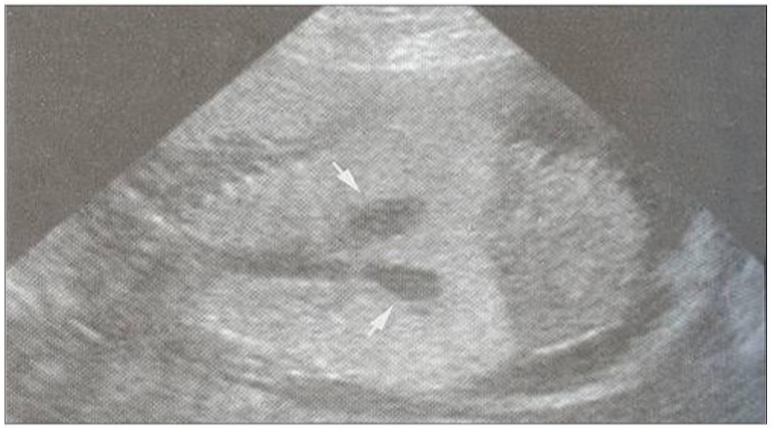
Congenital high airway obstruction is a rare life threatening condition reported since 1989. High airway obstruction can be at the level of larynx or trachea and is due to developmental absence or blockage of airway. CHAOS is diagnosed prenatally by ultrasound[1]. We report one such rare case of congenital high airway obstruction and its prognosis.

2. Case Report

An 18 year case of primigravida with second degree consanguineous marriage presented with 24 weeks period of gestation for regular ANC check up. She had regular menstrual cycle, had spontaneous conception. Her clinical examination revealed 24 weeks Period of gestation. 1st and 2nd trimesters were uneventful. Routine ante-natal investigations were within normal limits. Blood group and Rh typing - A positive.

Anomaly scan at 24 weeks revealed bilateral lungs enlarged and echogenic with dilated fluid filled trachea and major bronchus with polyhydramnios. Fetal growth was corresponding to gestational age, no other abnormalities were made out. (Figure-1)

Figure 1: Bilateral lungs enlarged and echogenic with dilated fluid filled trachea and major bronchus.

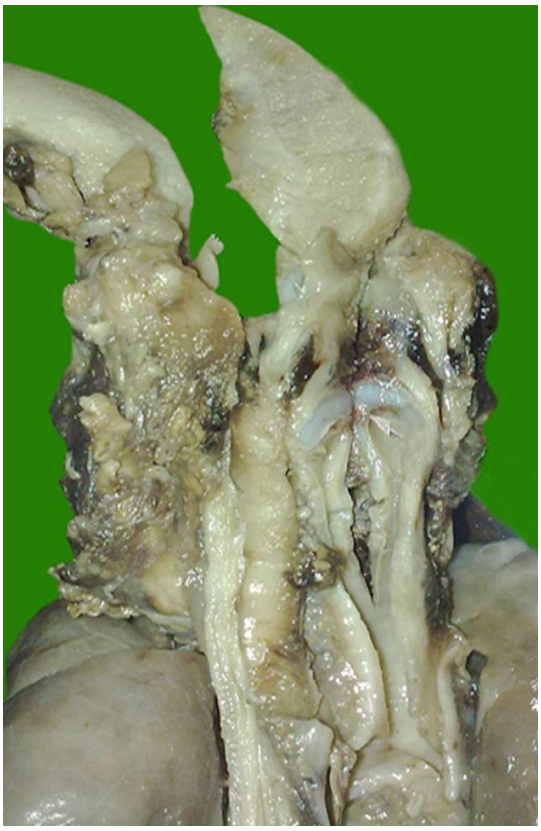


The lethal prognosis of fetus was discussed with the patient and relatives; concern was to continue the pregnancy.

Pregnancy was continued with serial ultrasound scans for growth monitoring and looked for hydrops. Delivery was planned electively at term for neonatal tracheostomy, but the patient went into spontaneous labour at 36 weeks of gestation admitted in advanced labour, delivered quickly, a live female baby, weighing 2000grams. No respiratory effort present. Endotracheal intubation failed and tracheostomy was done, in spite of the effort, baby succumbed after half an hour. External examination of the neonate revealed no dysmorphic features.

Autopsy was performed after taking consent from parents, that revealed subglottic stenosis of trachea and no other abnormalities were present (Figure-2).

Figure 2: Autopsy-subglottic stenosis of trachea.



3. Discussion

Congenital high airway obstruction syndrome is one of the rare life threatening conditions and usually has lethal outcome. Exact cause is not known, and most of the cases sporadic, though commonly caused by laryngeal atresia, subglottic stenosis or laryngeal web. However, several genetic syndromes have been linked with CHAOS and most commonly associated with Fraser's syndrome characterised by laryngeal atresia, cryptophthalmos, syndactyly, renal agenesis, abnormalities of ear and external genitalia.[1]

In CHAOS, the obstruction of the larynx or trachea that results in retention of bronchial secretions and pulmonary distension by the retained fluid. Over inflation of the lung with flattening or inversion of the diaphragm, cause compression of the heart and inferior vena cava, impairing venous return. Resulting in fetal hydrops and ascites. A detailed malformation scan is warranted to exclude additional malformation.[2,3,4,5]

Non-immune fetal hydrops is reported, though it is not present in our case.[2] Few cases have been successfully managed with neonatal intervention such as Extra-utero intrapartum therapy procedure (EXIT) and neonatal tracheostomy[6,7]. Very few babies are reported to be alive beyond the neonatal period. The long term outcome is unknown.[7]

To conclude CHAOS remains indeed very complex and challenging in the prenatal management of airway. Accurate diagnosis is the key to establish the diagnosis of CHAOS, fetal MRI being better for localisation of the level of obstruction. The accurate diagnosis of level of obstruction and planned neonatal airway management, either EXIT procedure or neonatal tracheostomy can salvage the neonate, which is otherwise succumbed to this lethal condition.

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