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Case report

Congenital High Airway Obstruction Syndrome (CHAOS): A perinatal autopsy case report

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ABSTRACT

Introduction: Congenital High Airway Obstruction Syndrome (CHAOS) is a rare anomaly where a partial or complete obstruction of upper airway is seen. As a consequence, the foetus develops enlarged lungs, inverted or flattened diaphragm and ascites. The pathological findings in a case of CHAOS is presented.
Case report: A 28-year-old primigravida presented with a history of 5 months of amenorrhoea. Ultrasonography revealed enlarged echogenic lungs, cardiac mid-position, inverted diaphragm, foetal ascites and talipes of left foot. The parents opted for termination of pregnancy. Foetal autopsy revealed a male foetus with ascites and talipes of left foot. Lungs were massively enlarged, diaphragm was flattened. A dome-shaped cricoid cartilage at the subglottic level was seen as the cause of obstruction.
Conclusion: CHAOS is a rare anomaly. A foetal autopsy can be helpful in establishing the level and nature of the obstruction and in documenting other associated foetal anomalies.

1. Introduction

Congenital High Airway Obstruction Syndrome (CHAOS) is an extremely rare anomaly wherein there is either a partial or complete obstruction of the foetal upper airways, resulting in a constellation of findings. The most common cause is said to be laryngeal atresia [1]. It can be antenatally diagnosed by ultrasonography (USG), which reveals enlarged echogenic lungs, an inverted or flattened diaphragm and foetal ascites [2]. The pathological findings in a case of CHAOS diagnosed at 20 weeks of gestation are presented.

2. Case report

A 28-year-old primigravida of a non-consanguineous marriage presented with a history of 5 months of amenorrhoea. She was non-hypertensive and non-diabetic. There was no significant past history or family history. All blood and biochemical parameters were within normal limits. The USG revealed enlarged echogenic lungs, cardiac mid-position, an inverted diaphragm, foetal ascites and mild talipes of left foot. No pleural or pericardial effusion was noted. The amniotic fluid volume was normal. A diagnosis of CHAOS was made, and the parents were counselled regarding the unfavourable prognosis, following which they opted for termination of pregnancy.

Foetal autopsy revealed a male foetus weighing 685 gm. External examination showed massive foetal ascites and talipes of left foot. [Figs. 1 and 2] Abdominal girth was 28.5 cm and 30 ml of clear yellow fluid was aspirated from the abdomen. All organs were in situ. The lungs were massively enlarged, together weighing 35 gm and showed costal impressions on the external surface. [Fig 3] The diaphragm was flattened. On careful dissection, an airway obstruction was noted at the subglottic level, where a dome-shaped cricoid cartilage was seen as the cause of obstruction. This is consistent with Type II atresia as described by Smith and Bain [3]. [Figs. 4 and 5] No tracheo-oesophageal fistula was observed. Histopathological evaluation showed laryngeal atresia with overgrowth of the cricoid cartilage. Bilateral lungs showed marked dilatation of the alveolar spaces. No other associated anomalies were noted, and no genetic studies were undertaken in this case. Based on the above findings, a diagnosis of CHAOS was confirmed.

3. Discussion

The term CHAOS was introduced by Hedrick et al. in 1994 [4]. CHAOS is a rare anomaly wherein there is an obstruction of the foetal upper airways. Laryngeal atresia is the most frequent aetiology. Other causes include laryngeal webs and cysts,

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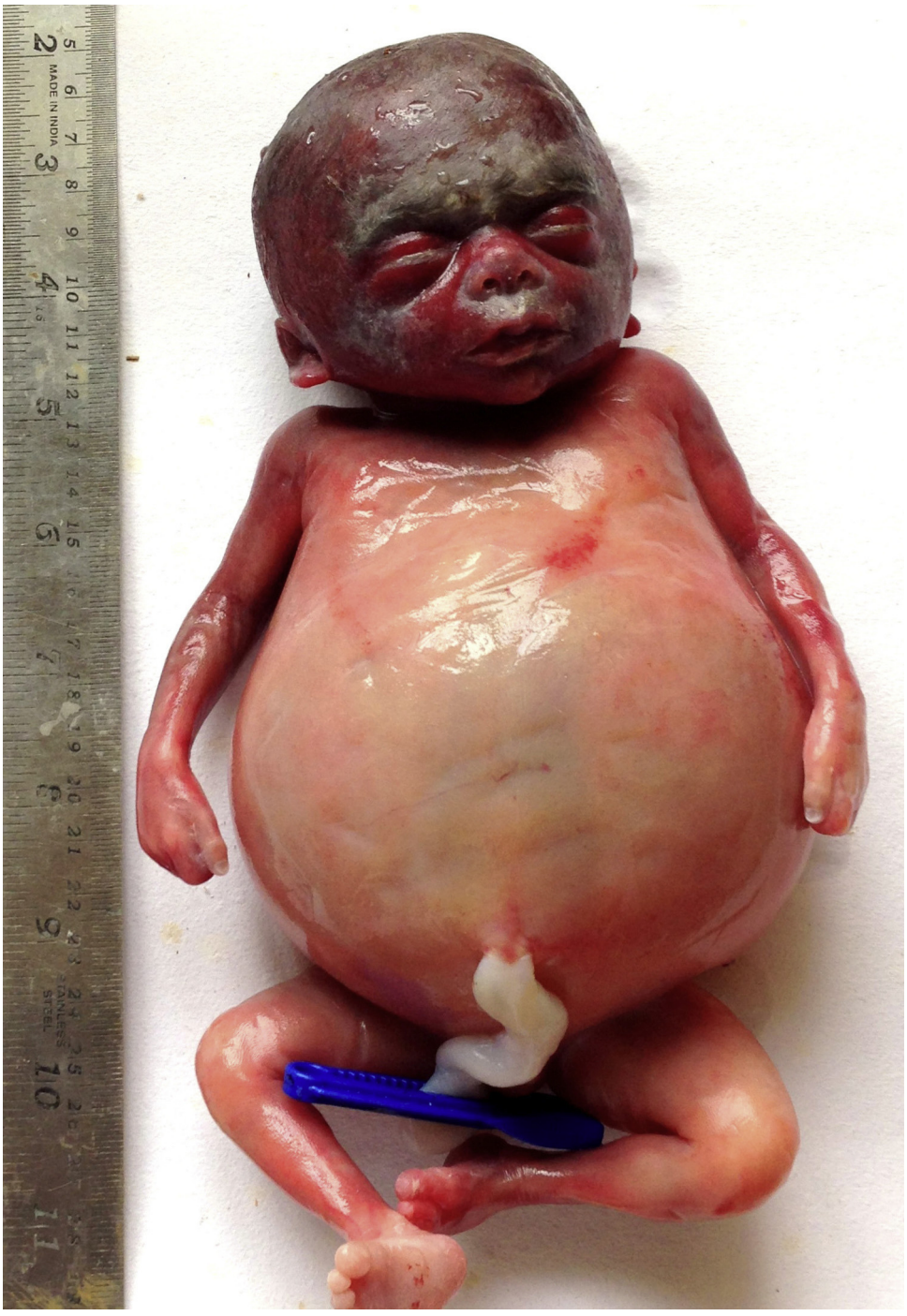

Fig. 1. Foetus shows massive ascites.

tracheal agenesis or atresia. Laryngeal atresia occurs due to the non-development of the 6th branchial arch and has been classified into three types by Smith and Bain in 1965 [3]

- Type I Complete atresia with midline fusion of the arytenoids cartilages
- Type II Infraglottic obstruction with a dome-shaped cricoid cartilage obstructing the lumen
- Type III Occlusion of the anterior fibrous membrane and fusion of the arytenoids at the level of the vocal processes.

The exact incidence of CHAOS is not known, as it most commonly sporadic in nature. Until 2007, about 52 cases had been documented [5]. CHAOS is also known to be associated with several genetic disorders, the most common being Fraser's syndrome, which is transmitted in an autosomal recessive fashion and is characterized by urogenital defects, laryngeal atresia, syndactyly and cryptopthalmus [2]. Other associated syndromes include Cri-du-Chat syndrome, short rib polydactyly syndrome, velocardiofacial syndrome, Shprintzen-Goldberg omphalocoele syndrome and TACRD (Tracheal agenesis, complex cardiac anomalies, radial ray defects, duodenal atresia) [5]. Chromosomal anomalies like trisomy 9, trisomy 16 and chromosome 5p deletion have also been documented