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Congenital Choanal Atresia – A Case Report

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Introduction

Choanal atresia (CA) is a rare developmental failure of the posterior nasal cavity to communicate with the nasopharynx[1]. Congenital choanal atresia was first described by Roederer in 1755[2]. Incidence of this developmental defect varies from 1 in 5000 to 1 in 8000 births[3,4]. Bilateral atresia is more common than unilateral[5-7].

Finding of cyclical cyanosis in newborn points towards the diagnosis of bilateral congenital choanal atresia.

Types of choanal atresia include bony and membranous type with bony atresia being more common[8]. Approximately 10% to 50% of patients with congenital CA have additional congenital anomalies including the cluster of defects known as "CHARGE" syndrome[1,3,5,6,9-16].

The "CHARGE" association is a poly-malformative disease associating - Coloboma, Heart disease, Atresia of choana, Retarded growth and development, Genital hypoplasia and Ear anomalies. "CHARGE" being an acronym based on these different malformations.

Data about the incidence and presentation of this congenital anomaly among Bangladeshi population is limited. In this paper, we report a unique case of bilateral congenital choanal atresia in a 2 hours old female newborn.

Case Report

A 2 hours old female newborn baby weighing 1.8 Kg was referred from Department of Gynae and Obstetrics to the neonatal unit of the Department of Paediatrics, Dhaka Medical College Hospital for birth asphyxia. Baby was delivered by caesarian section due to prolonged 1st stage with face presentation. On subsequent enquiry, it was found that during antenatal period, the mother was healthy and there was no history of taking any drugs.

The baby was the 1st issue of her non consanguineous parents and family history was unremarkable.

After admission, the baby was found to have respiratory distress and cyanosis, which was relieved by crying. Nasogastric tube could not be passed through the nose into the nasopharynx. She did not have any other apparent problems (Fig.-1).

The baby was investigated with X-ray of the chest and posterior choanogram. Although, computed tomography (CT) accurately delineates the abnormal anatomy of the nasal fossae, it was not possible to do because of financial constraints.

However, X-ray chest was normal. On posterior choanogram, no contrast was seen in the nasopharynx, but was seen in nasal cavity, which was suggestive of bilateral choanal atresia.

Subsequently, oto-laryngologist was consulted for further management. The membrane of both choana was perforated with a urethral dilator under general anaesthesia. A plastic tube was placed in situ and fixed with silk in both nasal passages (Fig. -2).

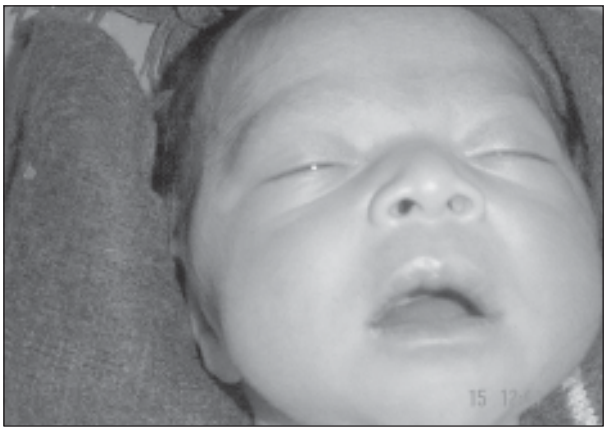

Fig.-1: *Apparently normal baby sleeping with opened mouth*

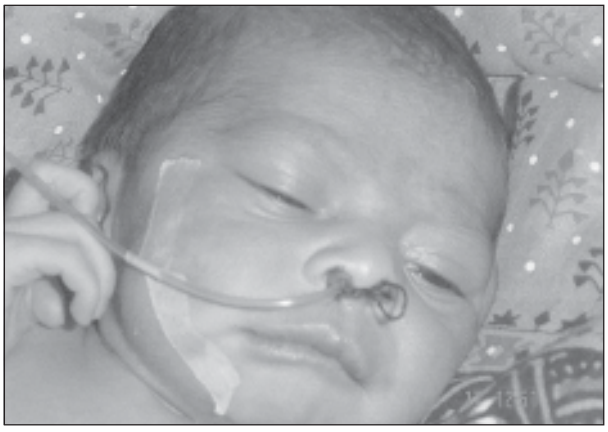

Fig.-2: *After surgery plastic tube placed in situ*

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Parenteral antibiotic was given in post-operative period. Irrigation of the tube was done with saline everyday to keep the tube patent and establish normal breathing. The tube was kept in situ for 2 weeks. No surgical complication was noted.

On 15th day of operation, the baby was discharged with an advice to come after 7 days. On subsequent follow up, the baby was found normal, was gaining weight properly and did not have any breathing difficulty.

Discussion

Although the most common form of nasal obstruction in neonates is soft tissue oedema, congenital nasal deformities are being recognized as an important cause of newborn airway obstruction[17,18]. Choanal atresia is considered as the most common congenital anomaly of the nose[17].

The incidence of CA is about 0.3 /1000 live birth[19]. There is no statistically significant difference between races in rates, even though white infants have a higher rate than those of other races[3]. Girls affected twice as common as boys[19] which is consistent with the present case.

The genetic aspects of choanal atresia have not been clearly defined. The defect is probably a mutifactorial trait as in cleft lip and palate[12]. The appearance of the condition in both single and successive generations supports this contention[20].

Some other authors hypothesized that the condition is of autosomal dominant inheritance with reduced penetrance[21,22]. They have suggested that incomplete penetrance and variable expressivity is caused by a defect of neural crest. Brenner[18] reported occurrence of CA in 2 siblings and other author described a familial tendency in most of the cases of CA[23]. The case presented in this paper was the 1st issue of her parent and no other family member had this type of problem.

A significantly higher mean paternal age at conception together with concordance in monozygotic twins and the existence of rare familial cases support the role of genetic factors such as de novo mutation of a mutant gene or subtle submicroscopic chromosome rearrangement[10]. Chromosomal abnormalities were found in 6% infants with choanal atresia[3]. Wouters[24] described partial triosomy 4q and monosomy 9p resulting from a familial translocation in a child with CA.

Meer[25] suggested the possibility of an undefined teratogen as an aetiologic factor for development of CA. Wilson[26] reported a case of bilateral choanal atresia who had been exposed to carbimazole in utero because of maternal Grave's disease. The mother of our case was healthy and did not have any history of taking any medicine other than vitamin.

The anatomic classification of CA is commonly quoted as 90% bony and 10% membranous[27]. Some of the authors found that it is unilateral and complete in majority of cases[27,28] but other reported bilateral atresia more frequently[24] which is consistent with the present case.

Choarial atresia can occur as an isolated anomaly as in the present case but more commonly associated with one or more concomitant congenital anomalies. The anomalies may include tall forehead, maxillary hypoplasia, prognathism, microphthalmia, hypertelorism, arhinia, mandibular hypoplasia, complete absence of tongue, limb anomaly, ectodermal dysplasia, polysyndactyly and ileal atresia[4,15,21,22,29,30].

The CT plays a significant role in the diagnostic and therapeutic approach to congenital choanal atresia and should be a method of choice to evaluate neonate with nasal obstruction.

Many authors described different techniques for management of posterior choanal atresia. Joshephson[7], Panwar[30] and Uri [31] suggested that the repair of congenital choanal atresia using the transnasal endoscopic approach is the best method. This route offers excellent visualization of the posterior choana and hence the ability to open the defect widely with a high surgical success.

On the other hand Sadek[23] and Reddy[32] were in favour of another method of treatment of bilateral CA which consisted of endonasal perforation with a curved trocar. A stenting technique was followed, the aim of which was to provide good fixation, stability and easy post-operative nursing care.

Friedman[29] informed that the correction can be done by a transnasal approach under endoscopic control. Straight urethral sound was used to perforate the plate. Endotracheal tube was subsequently inserted as nasal stent. Another author also supported this technique[33] as it is minimally invasive and less traumatic which was applied to the present case for correction of CA.

Conclusion

Bilateral congenital choanal atresia is a paediatric emergency and must be treated surgically. The newborn having respiratory difficulty and cyanosis at birth which is releived by crying should be suspected as a case of bilateral congenital choanal atresia. The doctors and rurses attending the newborns should be aware of this condition so that they can ensure the detection of the cases at birth and can give immediate relieve to those babies.

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