Case Report

Bilateral choanal atresia – An unusual case of severe respiratory distress in a newborn without nasal flaring

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ABSTRACT

Respiratory distress in a newborn can be due to various causes and some need active intervention. Choanal atresia (CA) is a rare congenital anomaly with its incidence estimated to be 1 case/5,000–8,000 births. It is characterized by narrowing or blockage of the nasal passages. It is important to make an early clinical diagnosis with emphasis on timely management as it can be life-threatening. The pediatrician may be not able to pass a feeding tube through the neonate's nostril even on repeated attempts. Detailed evaluation should be performed for the CHARGE association. High-resolution computed tomography can aid the diagnosis and transnasal endoscopic surgery is the preferred treatment modality. Here is a case report of a term neonate born with severe respiratory distress who was diagnosed to have bilateral CA on evaluation and managed with nasal endoscopic surgery.

Key words: Bilateral choanal atresia, CHARGE association, McGovern nipple, Paradoxical cyanosis, Transnasal endoscopic surgery

Respiratory distress is one of the common clinical presentations in newborns. Some newborns can present with clinically significant respiratory distress, in which active interventions are needed. Choanal atresia (CA) is an uncommon anomaly in newborns causing respiratory distress associated with failure of communication of the nasal cavity with the nasopharynx. This clinical entity was first reported by Roederer, in 1755, and anatomically described by Otto, in 1829, as the abnormal presence of an obstructive tissue that prevents the communication between the nasal cavity and nasopharynx [1]. Females are more affected than males (2:1) and 40% of cases are bilateral. It is thought to be caused by the persistence of the oronasal membrane [2]. Bilateral CA is a life-threatening emergency. Hence, early diagnosis and management are crucial to prevent respiratory distress and hypoxia.

CASE REPORT

A term female infant born by normal vaginal delivery (birth weight 2.21 Kg) to a second gravida mother without significant antenatal history was noted to have tachypnea and severe respiratory distress soon after birth. The antenatal period was uneventful. There was no significant medical or family history. The APGAR scores were 8/10 and 9/10 at 1 and 5 min, respectively.

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Soon after birth, the baby developed tachypnea and severe respiratory distress in the form of suprasternal and subcostal retraction without nasal flaring. The baby had a respiratory rate of 72/minute with the oxygen saturation ranging between 88% and 92%. However, the baby was noted to have an improvement in the saturation on crying. The chest X-ray was normal.

It was impossible to pass the feeding tube through both nostrils on repeated attempts. Hence, a possibility of CA was made. Imaging with high-resolution computed tomography (HRCT) (Fig. 1a) with contrast revealed bilateral membranous type CA. Nasal endoscopy confirmed the same (Fig. 2a). Routine blood investigations were within normal limits.

The baby was evaluated for CHARGE association (Coloboma iris, Heart defect, Atresia choanae, Retarded growth and development, Genital hypoplasia and Ear abnormalities and hearing defect) which showed congenital heart diseases such as atrial septal defect, multiple small ventricular septal defects, and tiny patent ductus arteriosus. Ophthalmology evaluation and ultrasound abdomen and pelvis were done to rule out coloboma and genitourinary abnormalities, respectively.

ENT opinion was sought and transnasal endoscopic surgical repair was done successfully with the bilateral stent in situ (Fig. 1b). The newborn was treated with IV antibiotics and other supportive measures. Post-surgery period was uneventful and the baby was discharged on day 14 of life. The stent was removed after 4 weeks and follow-up nasal endoscopy showed a patent nasal cavity (Fig. 2b).

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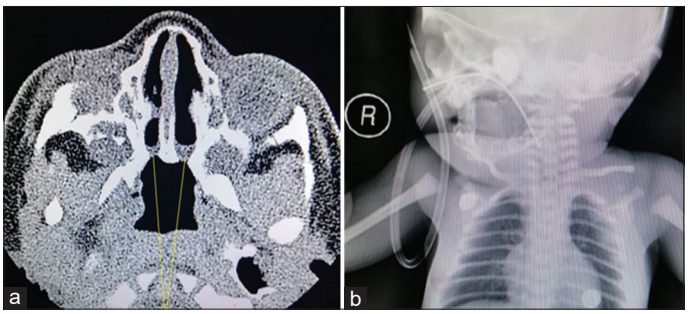

Figure 1: (a) High-resolution computed tomography with contrast revealed bilateral membranous type choanal atresia; (b) Follow-up X-ray showed successful placement of bilateral stent *in situ*

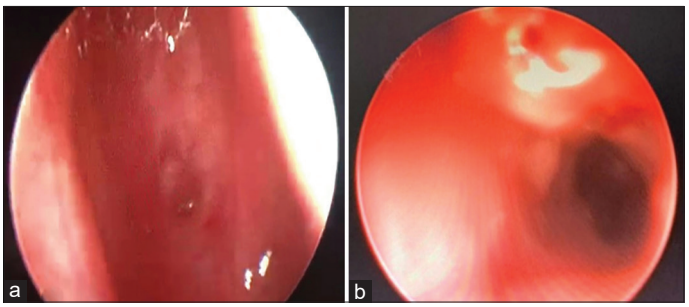

Figure 2: (a) Nasal endoscopic view before surgery; (b) Follow-up nasal endoscopy showed patent nasal cavity

DISCUSSION

Bilateral CA is a surgical emergency in newborns which occur as a part of the failure of communication of the nasal cavity with the nasopharynx. This congenital condition is caused by the failure of the migration of neural crest cells [2]. It may be bony or membranous or mixed [3,4]. Approximately, 1 in 5,000–8,000 births are associated with CA. Clear female predominance is found in many studies but have not been attributed to any etiopathological reason. Unilateral forms are more common and may present in later life. Bilateral cases account for about 36–40% of cases and are neonatal emergencies [1]. CA shows an interesting "2-1 rule" in many characteristics like the ratio of unilateral to bilateral CA, female-to-male ratio, and the incidence of the right side to the left side CA [5].

Four theories have been implicated in the pathogenesis of CA, which include (a) persistence of the buccopharyngeal membrane from the foregut; (b) abnormal persistence or location of mesoderm forming adhesions in the nasochoanal region; (c) abnormal persistence of nasobuccal membrane of Hochstetter; and (d) misdirection of neural crest cell migration [6]. Maternal intake of retinoic acid, thioamides such as methimazole, carbimazole, and propylthiouracil are associated with the development of CA [6].

The infant is an obligate nasal breather, except when the mouth is open. Bilateral CA can cause cyanosis and hypoxia which get relieved on crying or gasping since mouth opening can release the air obstruction (paradoxical cyanosis). Feeding difficulty, choking, or cyanosis may occur due to the inability to breathe and swallow together [3]. Nasal flaring is typically absent in bilateral CA as compared to other causes of respiratory distress in newborns. The baby may develop cyclical respiratory distress [4]. This asphyxia may, in turn, lead to cardiopulmonary arrest and death if not managed on an emergency basis. CA may be associated with many congenital anomalies, especially CHARGE syndrome. In the present case, growth retardation and acyanotic congenital heart disease were the associations found [5].

Diagnosis of CA can be clinical or imaging-based. Inability to pass the feeding tube or absence of misting in the postnasal mirror or spatula placed in front of the nose are diagnostic clues. HRCT with contrast is the diagnosis of choice. Transnasal endoscopy can also aid in the confirmation of the diagnosis. Differential diagnosis includes nasolacrimal duct cyst, pyriform aperture stenosis, turbinate hypertrophy, septal deviation, dermoid or pharyngeal bursa, encephalocele, or nasal neoplasm [2,3].

Emergency care in infants with bilateral CA includes endotracheal intubation or McGovern nipple to maintain the airway. Among the five different approaches proposed (transpalatal, transeptal, sublabial, transnasal, and transantral). Of which, transnasal endoscopic repair is most frequently performed. Stent placed *in situ* along with topical mitomycin C helps to prevent restenosis [3,7].

Rare cases of adults presenting with bilateral CA were also reported which can be managed with endoscopic choanoplasty. They may give a history of anosmia, nasal obstruction, mouth breathing, and nasal discharge since childhood [8]. Unilateral CA is not an emergency; hence, surgical correction can be delayed till 6 months of age or till 5 kg weight is attained [9].

Complications of surgery with stenting include hemorrhage, infection, choanal closure, choanal narrowing and stenosis, excoriation and erosion of nares, granulation tissue formation, stent blockage, stent displacement, and premature extrusion [10]. Some studies among newborns revealed that choanoplasty without stenting also had good outcomes with minimal blood loss, high rate of success, and minimum complications [11]. According to Brihaye *et al*., restenosis following surgery can be minimized by (a) construction of a large unineochoanae by removing the posterior part of the vomer and by drilling away the medial pterygoid; (b) choanoplasty without stenting; (c) rhinopharyngeal stenosis can be managed by resecting a part of the endochondral clivus bone; (d) covering raw surfaces with fibrin glue; and (e) appropriate post-operative care [12]. A study by Ramsden *et al*. showed that stent placed in situ along with topical mitomycin C helps to prevent restenosis [7]. Restenosis can be managed with carbon dioxide or Potassium titanyl phosphate laser or revision surgery [4].

CONCLUSION

CA is a rare congenital defect that can be either unilateral or bilateral. Bilateral CA is a surgical emergency in newborns and can present as paradoxical cyanosis and cyclical respiratory distress. CHARGE association and other congenital anomalies should be evaluated. Acute management includes airway maintenance and emergency surgical repair. Transnasal endoscopic repair is the preferred surgical method, though complication like restenosis is not uncommon.

RESEARCH ETHICS

Relevant Institutional Review Board and Ethics Committee provided approval for the case report dated 25-2-2022, Ethics committee, Medical Trust Hospital, Ernakulam.

PATIENT CONSENT

Written informed consent for patient information and images to be published was provided by the parents of the patient.

REFERENCES

1. Hadid F, Hakimi M, Benhommad O, Rochdi Y, Raji A. Neonatal choanal atresia: A series of 13 cases. Eur J Med Health Sci 2021;3:32-4.
2. Hassan AA, Bumuzah ZS, Alomar SA, Alwabari GA, AlAli ZA, Al Abbas ZY, *et al*. An unusual form of choanal atresia in a full-term newborn. Cureus 2020;12:11437.
3. Andaloro C, La Mantia I. Choanal atresia. In: StatPearls. Treasure Island, FL: StatPearls Publishing; 2022. Available from: https://www.ncbi.nlm.nih.gov/books/NBK507724 [Last accessed on 2022 Jan 21].
4. Roy S, Vivero R. Bilateral choanal atresia in a newborn. Ear Nose Throat J 2007;86:608-9.
5. Kwong KM. Current updates on choanal atresia. Front Pediatr 2015;3:52.
6. Pagon RA, Graham JM Jr., Zonana J, Yong SL. Coloboma, congenital heart disease, and choanal atresia with multiple anomalies: CHARGE association. J Pediatr 1981;99:223-7.
7. Ramsden JD, Campisi P, Forte V. Choanal atresia and choanal stenosis. Otolaryngol Clin North Am 2009;42:339-52.
8. Anajar S, Hassnaoui J, Rouadi S, Abada R, Roubal M, Mahtar M. A rare case report of bilateral choanal atresia in an adult. Int J Surg Case Rep 2017;37:127-9.
9. Moreddu E, Rossi ME, Nicollas R, Triglia JM. Prognostic factors and management of patients with choanal atresia. J Pediatr 2019;204:234-9.
10. Saafan ME. Endoscopic management of congenital bilateral posterior choanal atresia: Value of using stents. Eur Arch Otorhinolaryngol 2013;270:129-34.
11. Ibrahim AA, Magdy EA, Hassab MH. Endoscopic choanoplasty without stenting for congenital choanal atresia repair. Int J Pediatr Otorhinolaryngol 2010;74:144-50.
12. Brihaye P, Delpierre I, De Villé A, Johansson AB, Biarent D, Mansbach AL. Comprehensive management of congenital choanal atresia. Int J Pediat Otorhinolaryngol 2017;98:9-18.

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