Bilateral Congenital Choanal Atresia Presenting at Age Twenty Two years : An Unusual Case Report

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Abstract

Choanal atresia is a congenital absence of communication between the nasal cavity and nasopharynx. Bilateral Choanal Atresia usually present immediately after birth and in the neonatal period. We report an unusual case who presented at the age of twenty two years. This case was successfully managed by a transnasal approach. Choanal atresia should be considered as a rare diagnostic possibility in any patient who presents with total nasal obstruction and persistent mouth breathing.

Choanal atresia is a congenital failure of communication of the nasal cavity and nasopharynx. The atresia may be unilateral or bilateral, and it may be membranous or bony in nature. Incidence is quoted at 5 in 7000 live births (Lund et al, 1996). This congenital anomaly results in breathing difficulty and is associated with nasal discharge and impairment in sense of smell (Yamagishi et al, 1988). Bilateral complete choanal atresia is an acute respiratory emergency in newborns. Unilateral atresia is more commonly seen but usually not associated with respiratory distress. Unilateral atresia is more common than bilateral atresia in proportion 3 : 2. Rarely, neonates and infants compensate by breathing through the mouth and this may lead to delay in diagnosis for months or years (Candan et al, 1991). In this paper we have presented a young lady aged twenty two years with bilateral congenital choanal atresia which is a late and rare presentation.

Case Report

A twenty two years old female patient was referred to the Ear, Nose and Throat department for further evaluation and management of bilateral complete nasal obstruction with anosmia since birth. ENT examination revealed mucopurulent discharge in both nasal cavities. Cavities were cleaned and endoscopic transnasal visualisation of nasopharynx was not possible. Nasal catheter test suggested complete occlusion of the posterior choanae. Plain X-ray of the

paranasal sinuses showed haziness of maxillary sinuses. The contrast was held up at posterior choana on contrast nasography (Fig. 1). Axial CT Scan at the level of the posterior choana showed a thin bony ridge separating the anterior nasal cavities from the

Fig. 1

nasopharynx (Fig. 2). Thus the diagnosis of bilateral bony choanal atresia was confirmed. Patient had no other congenital abnormality, specially CHARGE association was looked for. (C-Coloboma of the eye; H-Heart disease; A-Atresia of the choana; G-Genital hypoplasia; and E-Ear anomalies or deafness).

The surgical repair of the atresia was carried out by a transnasal approach. After decongesting the nasal

Fig. 2

mucosa, the plate was visualised using storz 4.5mm 0 telescope. After the initial puncture of the atretic plate. It was then widened with sphenoidal punch forceps approximately 10mm in circumference dilated up to 12 mm with progressively larger dilators. Nasal silicon stent was placed for ten days post operatively. Follow up nasogram showed that posterior choanal opening was patent. On subsequent follow up at six months, patient had been symptom free from her nasal obstruction but her anosmia still persists.

Discussion

Choanal atresia is a congenital failure of communication between nasal cavity and

nasopharynx. Choanal atresia is more often unilateral with preponderance 3 : 2. Overall incidence of bilateral choanal atresia being 5/7000 live births. 90% of atresia are bony and 10% membranous (Lund et al, 1996). Occasionally there can be CHARGE* (Morgan et al, 1993) association, 56% of patient suffered with posterior choanal abnormalities in a study of 50 patients with CHARGE syndrome. Anosmia or impairment of smell presents with bilateral atresia is reported to be due to disuse atrophy of olfactory mucosa (Yamagishi et al, 1988). This condition is best diagnosed by high resolution axial CT scan after decongesting the nasal mucosa. CT scan also provides accurate differentiation to whether atresia is bony or membranous (Brown et al, 1986). Our patient came from a remote area where neonatal and paediatric services were not available. Diagnosis of choanal atresia was unfortunately not made over the subsequent years and she was treated conservatively for nasal obstruction. Although choanal atresia is ture medical emergency our patient was usually able to mouth breathe from neonatal period. Since the first description of choanal atresia by Roderer in 1755, many methods of surgical correction have been described. Among them four main approaches are well known.i.e 1) Transnasal 2) Transpalatal 3) Transseptal and 4) Transantral. The first two approaches are generally used for repairs. Transnasal approach is quicker and with minimal blood loss and does not interferes with palatal growth (Feuerstien et al, 1980).

In summary, choanal atresia should be considered as a rare diagnostic possibility in any patient who presents with total nasal obstruction and persistent mouth breathing.

Acknowledgement

The authors would like to thank Dr. Abir K. Bhattacharya, Consultant Otolaryngologist, and Senior Lecturer in Otolaryngology, University of London, U.K., for his assistance and Mrs. Kauthar Fajardo for meticulously typing the manuscript.

* (C-Coloboma of eye, H-Heart disease, A-Atresia of choana, G-Genital hypoplasia, E-Ear anomalies or deafness).

References

1. Bhattacharya, A. K, Lund V.J. (1996) · Unilateral choanal atresia in siblings-a rare occurrence *The Journal of Laryngology and Otology* 110 : 665-667.
2. Brown, O.E, Smith, T, Armstrong, E, Grundfast, K (1986) : The evaluation of choanal atresia by computed tomography. *International Journal of Paediatric Otorhinolaryngology* 12(1) · 85-98.
3. Candan, S, Mizrak, S. J, Karagoz, M, Muhtar, H, Gumele, H. R. (1991) : Bilateral congenital choanal atresia at age 16 . An interesting case. *Journal of Otolaryngology* 20 (6) 433-4.
4. Feuerstein, S. S, Krepsi, Y. P, Sachdev, R. K. (1980) : Transnasal correction of choanal atresia. *Head and Neck surgery* 3 . 97-104
5. Morgan, D, Bailey, N, Phelps, P, Bellman, S, Grace, A, Wyse, R. (1993) : Ear-Nose-Throat abnormalities in the CHARGE association. *Archives of Otorhinolaryngology Head and Neck Surgery.* 119 · 49-54.
6. Yamagishi, M, Hasegawa, S. and Nakano, Y. (1988) · Examination and classification of human olfactory mucosa in patients with clinical olfactory disturbances. *Archives of Otorhinolaryngology* 245 316-320.
7. Canovas-Llorens, D. L, Carbonell-Casasus, J. (1994) · Bilateral bony choanal atresia in an adult *Annals of Otorhinolaryngology. IBero-Am.* 21 (5) : 487-96.

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