Unilateral Choanal Atresia in Adult: Case Report

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Abstract: Unilateral choanal atresia usually presents later in life than bilateral and may present in adults. Here is a case report of a 19 year old male who presented with complains of unilateral nasal obstruction and discharge not clearly pointing to the diagnosis. The canalization was done endoscopically. This article focus the role of CT scan and nasal endoscopy in the diagnosis and treatment of the same.

Keywords: Choanal atresia, CHARGE, nasobuccal membrane, nasal obstruction, Endoscopy

1. Introduction

Choanal atresia is defined as a developmental failure of the posterior nasal cavity to communicate with the nasopharynx. Choanal atresia has an incidence of 1 in 5000 to 7000 births¹, occurs more commonly in females than in males and is more often unilateral than bilateral². Bilateral choanal atresia requires rapid management at birth and prompt diagnosis because neonates are predominantly obligate nasal breathers³. Unilateral choanal atresia is often diagnosed after several months of life. Unilateral choanal atresia is usually not associated with other facial anomaly or with syndromic malformations although most common associated congenital anomaly is CHARGE association (C=coloboma, H = heart disease, A = atresia of choanae, R = retarded growth and development, G =genital hypoplasia, E = ear deformities or deafness)⁴. Persistent unilateral rhinorrhea or failure to pass a catheter through the obstructed nasal fossa (during a general anaesthesia for example) makes the diagnosis of choanal atresia probable. Choanal atresia is an uncommon and rarely recognized cause of unilateral nasal obstruction. This case report documents the case of a patient with unilateral choanal atresia who remained undiagnosed for many years.

2. Case Report

A 19 year old man presented with a prolonged history of complete nasal obstruction and discharge on left side. He also complained of loss of sense of smell on left side since childhood with associated features of mouth breathing and snoring. On examination he had slight facial asymmetry with the dorsum slightly deviated to right.

On speculum examination initially it was confused as a case of chronic rhinosinusitus as thick vicid secretion could be seen at the floor of nose. On x-ray PNS only hazy bilateral maxillary sinuses could be noted.

Patient was taken for endoscopic examination, which on suction of the left nasal cavity on first pass nasal endoscopy showed absence of posterior choana, which was confirmed on Rhinography and CT-scan. It was finally diagnosed a case unilateral bony choanal atresia. This patient was treated multiple times before at many general practitioners as a case of allergic chronic rhinosinusitis. Computed tomography coronal cuts elicited bony atresia with prolapsed mucosal thickening at the left choana with polypoidal mass at posterior choanae region. Rest of the investigations where within normal limits.

Patient was posted for transnasal surgery. Endoscopic transnasal route was preferred, with initial use of burr and later enlarged using dilators. A pack was placed pre operatively in nasopharynx to protect the structures in the area. Silastic tube was inserted for six weeks to maintain the patency. Antibiotics, anti-inflammatory, saline nasal spray and proper nasal toileting was advised and the patient made an uneventful post-operative recovery.

3. Discussion

Choanal atresia is a congenital obstruction of posterior nasal aperture or choana, usually thought to be secondary to persistence of either the nasobuccal membrane of Hochstetter or the bucco-pharyngeal membrane from the foregut. This membrane normally ruptures between the fifth and sixth weeks of gestation to produce choanae. Failure of this membrane to rupture causes atresia of choanae and may be bony, membranous or mixed⁵. Previously reports suggested a 90% bony stenosis and 10% membranous, but more recent analysis suggests a mixed bony/membranous in 70% and pure bony in 30%⁶.

Unilateral or bilateral choanal atresia was first described by Roederer in 1751 and was first reported in Britain in 1881 by Ronaldson .Carl Emmert in Bern operated successfully on a patient of choanal atresia in 1851.He perforated the bilateral choanal atresia via the transnasal approach using a curved trocar after having practiced the perforating force on the hard palate of child's corpse. Bilateral choanal atresia presents at birth as a respiratory emergency. Occasionally unilateral choanal atresia may present in young with feeding difficulties especially when the non-affected side of the face is occluded. Unilateral cases do not present until late childhood or adulthood.

Anomalies associated with choanal atresia include polydactyly, nasal-auricular and palatal deformities, Crouzon's syndrome, Down syndrome, Treacher-Collinssyndrome, DiGeorge syndrome, craniosynostosis, microencephaly, meningocele, meningoencephalocele, facial asymmetry, hypoplasia of the orbit and midface, cleft palate, and hypertelorism⁷. Retrospective review of medical

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comorbid conditions of 78 children with choanal atresia found that common medical problems were otitis media with effusion (32%), upper and lower airway diseases (32% and 23%, respectively), cardiac anomalies (19%), and gastrointestinal tract disorders $(18\%)^8$. Statistically significant correlations were found for bilateral choanalatresia and cardiac disorders, CHARGE syndrome, obstructive sleep apnea, hematological problems, and prematurity or failure to thrive. Generally, 65% to 75% of patients with choanalatresia are unilateral, whereas the rest are bilateral⁹. About 30% are pure bony, whereas 70% are mixed bony-membranous¹⁰. Theatretic plate is usually sited in front of the posterior end of the nasal septum. The anatomic deformities include a narrow nasal cavity, lateral bony obstruction by the lateral pterygoidplate, medial obstruction caused by thickening of the vomer, and membranous obstruction¹¹. Acquired posterior choanal atresia rarely occurs. It is usually caused by rhinopharyngeal injury e.g. after adenoidectomy; radiotherapy for nasopharyngeal carcinoma¹²; tuberculosis or syphilis of epipharynx¹³, or sometimes by unknown causes.

4. Conclusion

Choanal atresia can be an isolated anomaly, however 60% cases of congenital defect has found to be associated with Down's and Treacher Collin's syndrome but may be found with other isolated defects such as micrognathia, trache-oesophageal fistula, cleft and high arched palate, missing teeth and facial cleft. In addition to these random associations choanal atresia has recently been linked with a number of specific defects the so called CHARGE association. Our reported case highlighted the fact that choanal atresia especially unilateral is a rarely recognised cause of nasal obstruction and is often diagnosed late. Hence endoscopic/radiological investigations are mandatory to make a diagnosis for complaints of persistent nasal obstruction for so many years.

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Figure 1: Rhinography



Figure 2: CT-Scan showing bony atresia



Figure 3: endoscopic view of the atresia



Figure 4: Endoscopic view of the atresia with inferior turbinate