A Rare Case Report: Bilateral Choanal Atresia in an Adult Patient

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Abstract

Posterior choanal atresia is rare but is the most common congenital malformation of the nasal cavity. It is characterized by a completely closed choana. Generally, this malformation is observed in newborns as increased respiratory distress during feeding, cyanotic attacks, and relaxation with crying. In contrast to unilateral atresia, bilateral disease is a life threatening condition and is extremely rare in adults. Herein, we presented a case of bilateral membranous choanal atresia detected in an 18-year-old patient. An 18-year-old female patient was admitted to our clinic after complaining of nasal obstruction, nasal discharge, snoring, anosmia, and mouth breathing since she was a child. On endoscopic examination, a right-deviated nasal septum and bilaterally closed choanae were observed. Membranous bilateral choanal atresia was observed on computed tomography (CT). The patient underwent transnasal endoscopic surgery under general anesthesia. Both septoplasty and bilateral coanoplasty surgeries were performed. No complications were observed postoperatively or at one year of follow up. Bilateral choanal atresia is a rare anomaly in adults. The diagnosis can easily be made by endoscopic examination or CT. The transnasal endoscopic surgical technique is an easy, reliable, and effective treatment method.

Keywords: Adult, bilateral choanal atresia, nasal obstruction, stent

INTRODUCTION

Posterior choanal atresia is rare but is the most common congenital anomaly of the nasal cavity. It is characterized by a single or double-sided completely closed choana with bone or soft tissue (1-4). In contrast to unilateral disease, bilateral disease is a life threatening condition (1). Choanal atresia is 90% osseous and osseomembranous and 10% membranous. Unilateral choanal atresia is usually diagnosed after complaints of unilateral nasal obstruction, unilateral mucoid rhinorrhea at later ages because it rarely causes respiratory stress (2). However, bilateral choanal atresia patients who are not diagnosed until adulthood are quite rare and interesting (2-5). Because bilateral atresia causes respiratory stress, which especially increases during feeding and then relaxes with crying, and cyanotic attacks in newborns, it is often noticed and treated at birth (3-5). Herein, we present a case of bilateral membranous choanal atresia detected in an 18-year-old patient. To our knowledge, this case is the eleventh adult with bilateral choanal atresia reported in the literature and the first case of restenosis not developing among cases without stents.

CASE PRESENTATION

An 18-year-old female patient was admitted to our clinic after complaining of bilateral nasal obstruction, bilateral nasal discharge, snoring, anosmia, and mouth breathing since she was a child. The patient stated that the physicians she had seen before told her that she had a bony and cartilaginous curvature in the nose and had to undergo surgery when she was 18 years old. The patient had no other congenital anomaly or history of trauma. The patient had apparent mouth breathing and a speech disorder. On endoscopic examination, mucoid discharge was observed in both nasal cavities. The nasal septum was deviated to the right, and both choanae were closed (Figure 1). Membranous bilateral choanal atresia was observed on computed tomography (CT) (Figure 2). Consent was obtained to be ethically appropriate for use in the scientific article without disclosing the patient's identity. The patient underwent transnasal endoscopic surgery under general anesthesia. Both septoplasty and bilateral coanoplasty surgeries were performed. The posterior part of the vomer was removed. The stent was not inserted. No complications or restenosis were observed postoperatively or at one year of follow up (Figure 3).

Cite this article as: Kars A, Bingöl F, Atalay F. A Rare Case Report: Bilateral Choanal Atresia in an Adult Patient. Eur J Rhinol Allergy 2020; 3(1): 26-8.

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Received: 20.07.2019
Accepted: 06.12.2019
DOI: 10.5152/ejra.2020.179

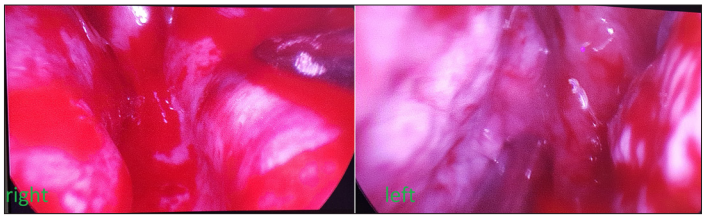

Figure 1. Endoscopic appearance of preoperative left and right choanal atresia

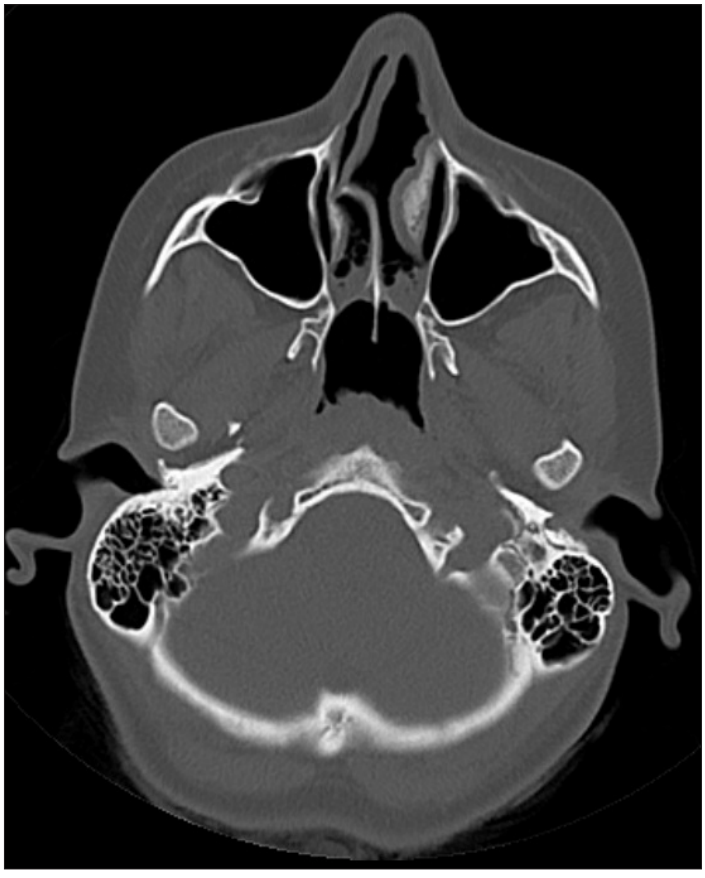

Figure 2. Axial section computed tomography (CT) view of bilateral choanal atresia

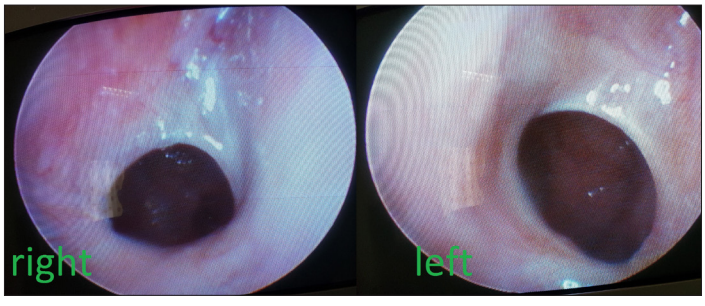

Figure 3. Endoscopic view of the right and left choana opening at one year postoperatively

DISCUSSION

Choanal atresia was first described by Johann Roderer in 1755 (4, 6). Posterior choanal atresia is rare but is the most common congenital anomaly of the nasal cavity; it is characterized by a single or double-sided complete closure of the choanae with bone or soft tissue (1-4). Choanal atresia is 90% osseous and osseomembranous and 10% membranous (2). Membranous atresia was detected in our patient. This anomaly is observed in 1 in 5000-7000 live births and is associated with other congenital anomalies in 50% of patients (1). Diseases such as microcephaly, micrognathia, Down syndrome, humeroradial synostosis, palatal defects, Treacher Collins syndrome, Crouzon syndrome, Apert syndrome, Pfeiffer syndrome, nasopharyngeal anomalies, branchial anomalies, mandibulofacial synostosis, and nasal ethmoidal encephalocele may accompany choanal atresia, but the most common accompanying anomaly is CHARGE syndrome (2, 4). Therefore, it is necessary to evaluate the cases detected in detail (3).

The etiology and pathogenesis of choanal atresia remains incompletely understood. In one study, a mutation in retinol dehydrogenase 10 related to A vitamin metabolism was shown to play a role in its etiology (7). According to one theory, choanal atresia occurs as a result of an abnormality in the migration and development of neural crest cells (3, 4). According to another theory, it is caused by the remnants of the nasobuccal and buccopharyngeal mucosa (4). Atresic plaque is located in front of the posterior vomer and the palate, and the plaque thickness is 1-12 mm (8).

Nasal endoscopic examination is important in the diagnosis of choanal atresia but does not provide information on the type of atresia. Therefore, the most used and most ideal method for diagnosis of choanal atresia and identifying the type of choanal atresia is CT (2). Membranous atresia was demonstrated with CT in our patient.

Bilateral choanal atresia is a condition that must be urgently treated in newborns (1). Because newborns learn mouth breathing at 4-6 weeks of age, they must initially breath through their nose (2, 3). In bilateral atresia, this situation presents respiratory stress, which especially increases during feeding and then relaxes with crying, and cyanotic attacks immediately after birth (2-4). Death is frequent in cases in which early diagnosis is not possible. Therefore, bilateral choanal atresia is rarely encountered in older patients (2). Only ten cases of bilateral choanal atresia were reported in the literature before our case.

In the treatment of choanal atresia, the open transpalatal technique has been replaced by a minimally invasive transnasal endoscopic technique (9). The transnasal endoscopic method is a simple, safe, and effective approach to treat this disease (2). The most common postoperative complication is restenosis. A stent can be used to prevent this complication, but the use of a stent is controversial (1). Some researchers advocate the superiority of stent use (10). We used the transnasal endoscopic method similar to that used in the cases in the literature in our patient. Due to the risk of a foreign body reaction, the need to use long-term antibiotics, and the risk of necrosis in the columella region, we decided not to use a stent. We aimed to prevent re-narrowing by removing the posterior portion of the vomer. No restenosis was observed in our patient at one year. Only 1 of 10 adult patients with bilateral atresia reported in the literature required revision surgery due to restenosis (5).

CONCLUSION

Bilateral choanal atresia is an extremely rare condition in adults. It can be easily diagnosed by nasal endoscopic examination or CT. Transnasal endoscopic choanaplasty is the current treatment approach. This treatment should be carefully considered in patients with complaints of continuous bilateral nasal obstruction, bilateral nasal discharge, snoring, anosmia, and mouth breathing.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - A.K.; Design - F.B. Supervision - F.A.; Fundings - F.A.; Materials - F.B.; Data Collection and/or Processing - A.K.; Analysis and/or Interpretation - F.A.; Literature Search - A.K.; Writing Manuscript - A.K.; Critical Review - F.B.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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