Adult congenital bilateral choanal atresia at 49-year-old: a case report of a rare entity

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49 yaşında bilateral konjenital yetişkin koanal atrezi: Nadir bir oğlu sunumu

Newborns are obligatory nose-breathers in the first few months of their births. Bilateral congenital choanal atresia in neonates is an emergency situation, manifesting by irregular breathing, intermittent cyanosis, relief of symptoms by crying, worsening of respiratory distress by feeding, and sometimes, death due to neonatal asphyxia. Congenital bilateral choanal atresia is very rarely reported in the literature to be reaching to adulthood without being noticed. We present a case of bilateral choanal atresia with the complaints of bilateral nasal blockage and anosmia at 49-years-old who is the second oldest ever reported case.

Key Words: Choanal atresia, asphyxia, adult, bilateral, nasal obstruction.

Introduction

Choanal atresia is the congenital obstruction of posterior nasal cavity, and encountered at a rate of 1/5000-8000 live births. Most of the atresias are unilateral (67%), right sided and, having bone component (90%). However in the latest literature it is being advocated that most of the anomalies have a
mixed bony-membranous structure. Twenty to fifty per cent of these cases are associated with other congenital anomalies. Cardiac and craniofacial anomalies, hypopituitarism, cleft palate, ear anomalies, growth retardation, and CHARGE syndrome (coloboma, hearing deficits, choanal atresia, growth retardation, genital defects, and endocardial cushion defects) are some of the commonly encountered associated anomalies.

Unilateral choanal atresias might not be detected until childhood or adulthood, despite causing nasal obstruction, mucoid nasal discharge, anosmia, sleep disturbances, and day time fatigue. Bilateral choanal atresias (BCA), on the other hand, may necessitate emergency intervention because of respiratory difficulty at birth. Moreover, undiagnosed BCA frequently lead to death due to neonatal asphyxia. Consequently, BCA cases surviving until adulthood are rare.

In this paper, we present a case of bilateral choanal atresia patient at 49-year-old who is the second oldest ever reported.

**Case Report**

A female patient of 49-year-old presented to our clinic with the complaints of nasal blockage since birth, and anosmia. On nasal examination, any nasal deviation sufficient enough to cause nasal blockage was not detected, and it was also noted that her inferior and middle conchae were atrophic. BCA was diagnosed with endoscopic examination. The patient had neither any associated congenital anomaly nor any history of respiratory difficulty, cyanosis, asphyxia during neonatal period, nor surgical intervention, nor trauma. Computerized tomography (CT) revealed that atretic plate was having both bone and fibrous components (Figure 1). The procedure of transnasal opening of choanal atresia with endoscopic method was planned.

Under general hypotensive anesthesia, mucoperiosteal flaps were dissected so as to reveal the atretic plate utilising 0-degree, 4 mm nasal endoscope. The bony part of the atretic plate was drilled starting from its inferomedial border using a long shaft diamond burr. Neochoana was widened until the perpendicular plate of the palatine bone and medial pterygoid lamina of the sphenoid bone laterally, the nasal septum medially, palatine bone inferiorly and the sphenoid sinus superiorly. The diamond burr was also used to remove the posterior aspect of the vomer, preserving the mucosa at the other side. Exposed osseous parts were covered by mucosa. Gelatin sponge soaked with mitomycin with the dosage of 0.4 mg/ml was applied to the newly opened choanal area for 3 minutes. In order to provide a stent, a Nelaton catheter (internal diameter 6 mm, outer diameter 1 cm) was fenestrated in the middle and bent; one end of it was then placed into

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*Figure 1. Axial CT scan depicting the bilateral choanal atresia.*
the nasal cavity and the other end was introduced into the other nasal cavity as to reach to the nasopharynx. Afterwards, both ends of the Nelaton catheter was tied in the nasopharynx to form a loop shape, and the catheter loop rotated as to re-place the tied ends in front of nose and fenestrated portion in the nasopharynx. Finally, the tied ends were opened to aid nasal breathing.

The procedure was uncomplicated postoperatively. Nelaton stent was withdrawn after 6 weeks, and follow-ups at one and three months showed no granulation tissue at operation site and the choanae were patent. The clinical and radiographical examination at 4th year confirmed that the choanae were still patent and wide enough, and also the patient was not experiencing nasal blockage complaint (Figure 2). Unfortunately, there was no improvement in the smell sensation of the patient.

**Discussion**

Neonates are obligatory nose-breathers in the first few months of their lives. For this reason the presence of congenital BCA is an emergency situation. On the examination of such a neonate, one may expect to find irregular breathing, intermittent cyanosis, relief of respiratory difficulty with crying, and worsening with feeding. Especially undiagnosed BCA can cause death with neonatal asphyxia. Very rarely, neonates with complete BCA can gain the ability to breathe through the mouth. In this case the neonate, if survives, has a constant mouth breathing and continuous nasal discharge.

When we searched the literature for BCA diagnosed at adulthood, we found that the case of Yaşar and Özkul at 51-year-old, was the oldest ever reported case. The rest of the literature was reporting cases diagnosed at between 17 and 33-year-old.
Meanwhile, our patient was 49-year-old and, as to our knowledge, was the second oldest ever reported case. On the other hand, the duration of the postoperative follow up of our case was 4 years. This was substantially longer than the one Yaşar and Özkul reported which was followed up 18 months only.

Despite the nasal blockage was improved the patient still continued having anosmia. Gross-Isseroff et al. have studied the postoperative olfactory function at their case series of four of which 3 of them were BCA, and concluded that bilateral cases sustained olfactory deficit while unilateral case had the normal olfactory function. Although the number of their cases was very limited, they further speculated that early sensorial stimulation is necessary for the olfactory function to develop normally in a similar fashion to visual development. Likewise, we also think that there has been a permanent olfactory function loss in our case due to deprivation of any stimuli for long years.

The treatment of choanal atresia is surgical. The ideal procedure should be a short and safe operation involving complete removal of atretic plaque, and maintenance of a longstanding patency without damaging the important structures for the facial development. For these purposes, apart from puncture and dilatation, four different surgical techniques have been defined. The transpalatal and transnasal approaches are the two main approaches for repair of choanal atresia. However, these techniques have some drawbacks. Transpalatal technique is disadvantageous because of bleeding, risk of palatin fistula, disruption of palatin development, and orthodontic problems. Transantral sphenopalatin technique, even though there is less risk of damaging vascular structures, is also disadvantageous because of risk of damaging developmental structures. These approaches are not advised for children less than 6 years of age because of the risk of adversely influencing facial development. Transseptal approach is advisable for the children with unilateral choanal atresia. Stankiewicz has first described the transnasal endoscopic technique using a Hopkins rod telescope. Endoscopic transnasal approach provides a high resolution direct field of vision. With minimal damage to normal anatomy, the exact borders of atretic plaque, and posterior aspect of vomer is very clearly visualized. With these aspects, the restenosis rates can be reduced and success rates can be increased.

The failure due to granulation tissue and restenosis represents the main problems for choanal atresia surgery. Mitomycin is an antitumor agent known to inhibit fibroblast proliferation and migration and allows for healing with decreased scar formation. From this perspective, the application of mitomycin to the neochoanae during choanal atresia surgery seems to improve the outcome of surgery by eliminating postoperative granulation tissue and reducing the rate of restenosis.

As a conclusion, bilateral choanal atresias may necessitate emergency intervention. Moreover, undiagnosed BCA frequently lead to neonatal death due to asphyxia. For that reason, BCA diagnosed at adulthood is quite rare and, can be successfully treated by the endoscopic transnasal approach in all age groups.

References


**Conflict of interest statement:**
No conflicts declared.

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