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OLGU BİLDİRİSİ / CASE REPORT

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

S. Aslan, C. Yılmazer, H. Yavuz, L.N. Özlüoğlu

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

Yenidoğanlar hayatlarının ilk birkaç ayında zorunlu olarak burundan nefes alırlar. Yenidoğanda bilateral konjenital atrezi; düzensiz nefes alma, intermittan siyanoz, ağlama ile semptomların düzelmesi, beslenme ile respiratuvar sıkıntının artması ve bazen neonatal asfiksiye bağlı ölümlere neden olması dolayısı ile acil bir durumdur. Fark edilmeden yetişkin döneme kadar ulaşabilen konjenital bilateral koanal atrezi literatürde çok nadir olarak belirtilmiştir. Biz bilateral burun tıkanıklığı ve anosmi şikayeti bulunan, literatürdeki ikinci en yaşlı olgu olan 49 yaşında bilateral koanal atrezi hastasını sunuyoruz.

Anahtar Sözcükler: Koanal atrezi, asfiksi, yetişkin, bilateral, burun tıkanıklığı.

Abstract

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, intermittant 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.

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Introduction

Choanal atresia is the congenital obstruction of posterior nasal cavity, and encountered at a rate of 1/5000-8000 live births.[1] Most of the atresias are unilateral (67%), right sided and, having bone component (90%).[2,3] However in the latest literature it is being advocated that most of the anomalies have a

Sündüs Aslan, MD; Cüneyt Yılmazer, MD; Haluk Yavuz, MD; Levent N. Özlüoğlu, MD
Department of Otolaryngology and Head&Neck Surgery, Faculty of Medicine, Başkent University, Ankara

mixed bony-membranous structure.[4] Twenty to fifty per cent of these cases are associated with other congenital anomalies.[5,6] 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.[7-10]

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 necessiate 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.

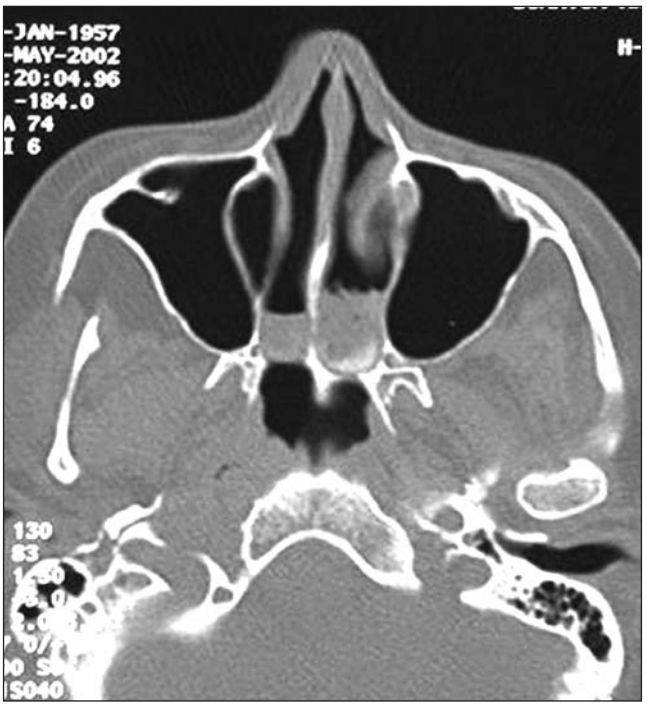

Figure 1. Axial CT scan depicting the bilateral choanal atresia.

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 pyterigoid 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

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.[6,11] Very rarely, neonates with complete BCA can gain the ability to breathe through the mouth.[12] In this case the neonate, if survives, has a constant mouth breathing and continuous nasal discharge.[11]

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.[13] The rest of the literature was reporting cases diagnosed at between 17 and 33-year-old.[14-18]

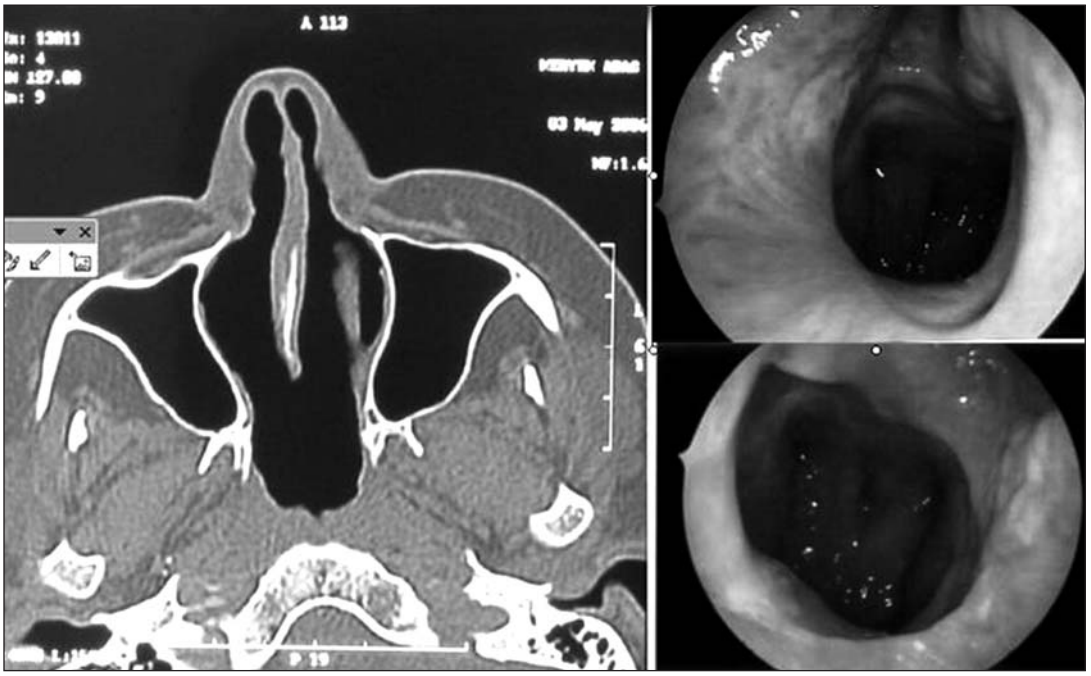

Figure 2. It is being demonstrated that conchae are bilaterally patent 4 years after the surgery with CT scanning (**A**) and endoscopic examination (**B** and **C**).

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.[15] 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.[19-22] However, these techniques have some drawbacks. Transpalatal technique is disadvantageous because of bleeding, risk of palatin fistula, disruption of palatin development, and orthodontic problems.[23] 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.[24] Transseptal approach is advisable for the children with unilateral choanal atresia.[25,26] Stankiewicz has first described the transnasal endoscopic technique using a Hopkins rod telescope.[27] 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.[28,29]

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 mytomycin 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.[30,31]

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

1. **Krespi YP, Husain S, Levine TM, Reede DL.** Sublabial transseptal repair of choanal atresia or stenosis. *Laryngoscope* 1987;97:1402-6.
2. **Gujrathi CS, Daniel SJ, James AL, Forte V.** Management of bilateral choanal atresia in the neonate: an institutional review. *Int J Pediatr Otorhinolaryngol* 2004;68:399-407.
3. **Uri N, Greenberg E.** Endoscopic repair of choanal atresia: practical operative technique. *Am J Otolaryngol* 2001;22:321-3.
4. **Josephson GD, Vickery CL, Giles WC, Gross CW.** Transnasal endoscopic repair of congenital choanal atresia: long-term results. *Arch Otolaryngol Head Neck Surg* 1998;124:537-40.
5. **Lazar RH, Younis RT.** Transnasal repair of choanal atresia using telescopes. *Arch Otolaryngol Head Neck Surg* 1995;121:517-20.

6. **Deutsch E, Kaufman M, Eilon A.** Transnasal endoscopic management of choanal atresia. *Int J Pediatr Otorhinolaryngol* 1997;40:19-26.
7. **Dunham ME, Miller RP.** Bilateral choanal atresia associated with malformation of the anterior skull base: embryogenesis and clinical implications. *Ann Otol Rhinol Laryngol* 1992;101:916-9.
8. **Brown OE, Burns DK, Smith TH, Rutledge JC.** Bilateral posterior choanal atresia: a morphologic and histologic study, and computed tomographic correlation. *Int J Pediatr Otorhinolaryngol* 1987;13: 125-42.
9. **Ferguson JL, Neel HB.** Choanal atresia: treatment trends in 47 patients over 33 years. *Ann Otol Rhinol Laryngol* 1989;98:110-2.
10. **Morgan DW, Bailey CM.** Current management of choanal atresia. *Int J Pediatr Otorhinolaryngol* 1990;19:1-13.
11. **Caldarelli DD, Friedberg SA.** Transnasal microsurgical correction of choanal atresia. *Laryngoscope* 1977;87:2023-30.
12. **Baker DC Jr, Waltner JG, Novick W.** Posterior choanal atresia. *Ann Otol Rhinol Laryngol* 1960;69:805-9.
13. **Yaşar H, Özkul MH.** Bilateral congenital choanal atresia in a 51-year-old woman. *Am J Rhinol* 2007;21:716-8.
14. **Carpenter RJ, Neel HB 3rd.** Correction of congenital choanal atresia in children and adults *Laryngoscope* 1977;87:1304-11.
15. **Gross-Isseroff R, Ophir D, Marshak G, Ganchrow JR, Beizer M, Lancet D.** Olfactory function following late repair of choanal atresia. *Laryngoscope* 1989;99:1165-6.
16. **Panda NK, Simhadri S, Ghosh S.** Bilateral choanal atresia in an adult: is it compatible with life? *J Laryngol Otol* 2004;118:244-5.
17. **Nakata S, Noda A, Misawa H, Yanagi E, Yagi H, Nakashima T.** Obstructive sleep apnoea associated with congenital choanal atresia. *J Laryngol Otol* 2005;119:209-11.
18. **Uslu H, Uslu C, Varoğlu E, et al.** Bilateral choanal atresia; evaluation with scintigraphy: case report. *Int J Pediatr Otorhinolaryngol* 2006;70:171-3.
19. **Vickery CL, Gross CW.** Advanced drill technology in treatment of congenital choanal atresia. *Otolaryngol Clin North Am* 1997;30:457-65.
20. **Pirsig W.** Surgery of choanal atresia in infants and children: historical notes and updated review. *Int J Pediatr Otorhinolaryngol* 1986; 11:153-70.
21. **Schweinfurth JM.** Image guidance–assisted repair of bilateral choanal atresia. *Laryngoscope* 2002;112:2096-8.
22. **Pasquini E, Sciarretta V, Saggese D, Cantaroni C, Macrì G, Farneti G.** Endoscopic treatment of congenital choanal atresia. *Int J Pediatr Otorhinolaryngol* 2003;67:271-6.
23. **Park AH, Brockenbrough J, Stankiewicz J.** Endoscopic versus traditional approaches to choanal atresia. *Otolaryngol Clin North Am* 2000;33:77-90.
24. **Osguthorpe JD, Singleton GT, Adkins WY.** The surgical approach to bilateral choanal atresia. Analysis of 14 cases. *Arch Otolaryngol* 1982;108:366-9.
25. **Hall WJ, Watanabe T, Kenan PD, Baylin G.** Transseptal repair of unilateral choanal atresia. *Arch Otolarnygol* 1982;108:659-61.
26. **Mcintosh WA.** Transseptal approach to unilateral posterior choanal atresia. *J Laryngol Otol* 1986;100:1133-7.
27. **Stankiewicz JA.** The endoscopic repair of choanal atresia. *Otolaryngol Head Neck Surg* 1990;103:931-7.
28. **Khafagy YW.** Endoscopic repair of bilateral congenital choanal atresia. *Laryngoscope* 2002;112:316-9.
29. **Cedin AC, Peixoto Rocha JF Jr, Deppermann MB, Moraes Manzano PA, Murao M, Shimuta AS.** Transnasal endoscopic surgery of choanal atresia without the use of stents. *Laryngoscope* 2002; 112:750-2.
30. **Holland BW, McGuirt WF Jr.** Surgical management of choanal atresia: improved outcome using mitomycin. *Arch Otolaryngol Head Neck Surg* 2001;127:1375-80.
31. **Prasad M, Ward RF, April MM, Bent JP, Froehlich P.** Topical mitomycin as an adjunct to choanal atresia repair. *Arch Otolaryngol Head Neck Surg* 2002:128:398-400.

Conflict of interest statement:
No conflicts declared.

Correspondence: Sündüs Aslan, MD
Ürgen Paşa Mah. 75. Yıl Bulvarı
Ceren Apt. No:6 Daire:19
Antakya HATAY
Gsm: (0506) 770 92 91
e-posta: drsundus@gmail.com

İLAN
BAŞ YÜZ Ağrıları
KİTABI
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İLAN
KBB KONGRE