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CLINICAL RECORD

Bilateral congenital choanal atresia encountered in late adulthood

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Abstract

Objective: We describe a case of bilateral congenital choanal atresia in the oldest patient reported with this condition in the recent English language literature.

Method: Case report and a review of the relevant English language literature, presenting the embryopathogenesis, diagnostic methods and treatment options for this condition.

Results: A 53-year-old woman having difficulty with nasal breathing, and with a continuous nasal discharge, was admitted to our clinic. Bilateral congenital choanal atresia was diagnosed by endoscopic examination and paranasal sinus computed tomography. Surgical treatment used an endoscopic transnasal approach. The follow-up examination a year later revealed adequate choanal openings bilaterally.

Conclusion: To our knowledge, this is the oldest patient with bilateral congenital choanal atresia to be reported in the recent literature. This condition is rarely encountered in adulthood but should be considered as a possible differential diagnosis of persistent nasal obstruction.

Key words: Choanal Atresia; Adult; Nasal Obstructions; Diagnosis

Introduction

Congenital choanal atresia is a rare fetal developmental anomaly characterised by unilateral or bilateral obliteration of the posterior nasal opening by bone or soft tissue.[1] Approximately 60 per cent of choanal atresia cases are unilateral.[2] The choanal obliterating plaque consists of bone in 30 per cent of cases and a bony-membranous mixture in 70 per cent. It is possible to encounter complete or incomplete obliteration.[3,4] Respiratory distress is seen in almost all bilateral congenital choanal atresia cases directly after birth.[5] The conventional view is that urgent surgery is life-saving in neonates because they are obligatory nasal breathers. Bilateral congenital choanal atresia is very rare in adulthood, and there are only a few cases reported in the literature.

Case report

A 53-year-old woman was referred to our clinic with difficulty in nasal breathing and continuous nasal discharge. Bilateral congenital choanal atresia was diagnosed by endoscopic examination (Figure 1) and paranasal sinus computed tomography (CT) (Figure 2). There were no other anomalies. The patient's past medical history revealed only chronic sinonasal problems with predominant nasal obstruction.

The patient was operated upon under general anaesthesia. Endoscopic examination along with palpation gave the impression that the bilateral plaques were of the mixed type. The overlying mucosa was elevated with a blade and elevator, and flaps were prepared. The membranous part of the atresia was opened by resection, and then the pterygoid process of the sphenoid bone and vomer were drilled out. Adequate bilateral choanal openings were created (Figure 3). Stents prepared from a size six silicon endotracheal tube were placed in the openings and fixed to the membranous septum with 3/0 silk sutures.

The post-operative period was uneventful, and the stents were removed six weeks later. Follow-up examination a year later revealed adequate choanal openings bilaterally.

Discussion

Bilateral congenital choanal atresia was first described by Roederer in 1755. The first reported case was from Germany, presented by Otto in 1830.[1,3]

There are various theories about the embryopathogenesis of congenital choanal atresia. One states that the condition results from a persistent nasobuccal or buccopharyngeal membrane. Another theory proposes the abnormal migration of neural crest cells.[1,5,6] Most

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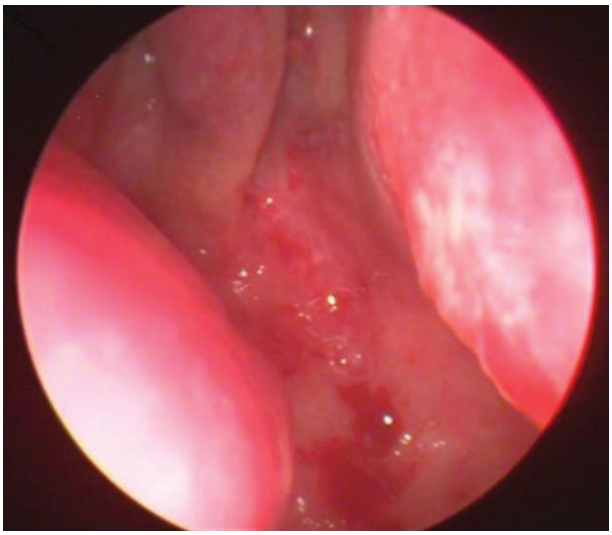

FIG. 1
Endoscopic, posterior view of the right nasal passage.

bilateral congenital choanal atresia cases are associated with other anomalies such as the 'CHARGE' syndrome, comprising coloboma, heart anomalies, growth retardation, and genital and ear anomalies.[1,3,5]

Bilateral congenital choanal atresia is characterised clinically by respiratory distress and cyanosis associated with feeding, which disappear when crying. This is because infants are compulsory nasal breathers in the first four months after birth, due to the high cervical localisation of the larynx at this age.[3] For this reason, newborn bilateral congenital choanal atresia is an emergency condition for which early diagnosis and treatment are vital. Unilateral atresia, however, may often remain unrecognised for many years as its manifestations are non-specific, for example, rhinorrhoea and obstruction.

Congenital choanal atresia can be diagnosed by several methods. The easiest is to try to pass a small

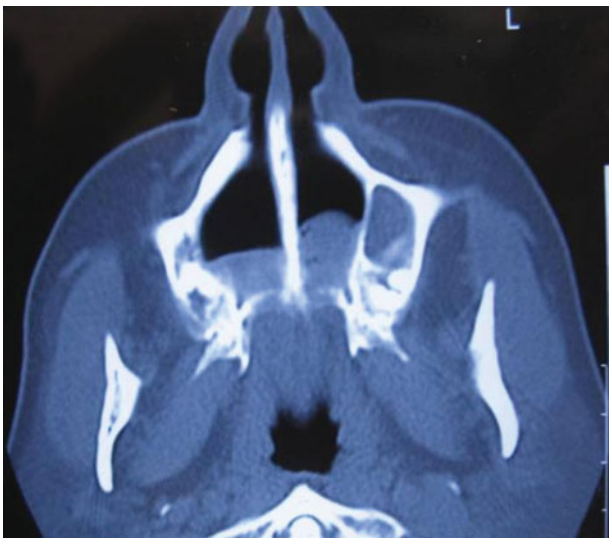

FIG. 2
Axial computed tomography head scan showing the patient's choanal atresia.

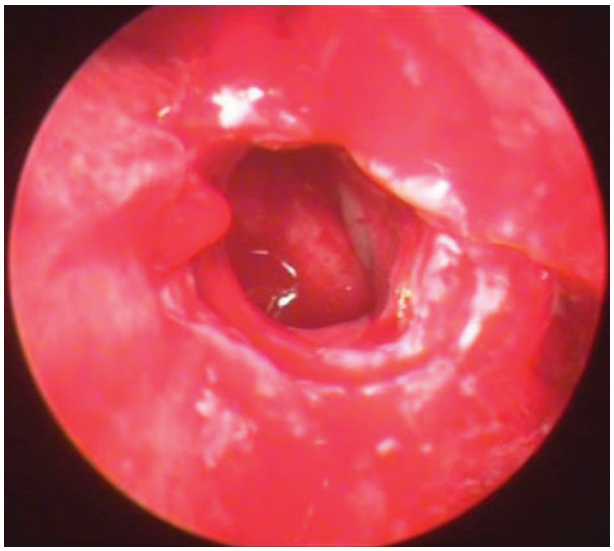

FIG. 3
Endoscopic view showing an adequate choanal opening following endoscopic surgery.

Nelaton catheter from the nose into the nasopharynx. It should be noted that the distance from the nares to the posterior nasopharyngeal wall is at least 2.5 cm. Rigid or flexible endoscopy, operational microscopy, a nasopharyngeal mirror or finger examination can also be used. Radiography, performed in the supine position with a radiopaque agent given through the nasal cavity, is another, rarely used option. Paranasal sinus CT is the standard evaluation method in diagnostic radiology.[1,3,5] Computed tomography scanning enables the detection of bilateral or unilateral atresia, bony and membranous components, and other associated nasal abnormalities. Quantitative measurements made from the CT scan can also be used to guide surgical treatment of the condition.

Many techniques have been described for the management of congenital choanal atresia. Currently, the most commonly used method is an endoscopic transnasal approach. Transpalatal and trans-septal approaches are not often used. Endoscopic transnasal interventions should be preferred because they provide better visualisation and lower complication rates.[7] Transpalatal and trans-septal approaches are not advised for newborns. Spinal canal injury, cerebrospinal fluid leakage and cavernous sinus injury are the most worrisome surgical complications. These complications are minimised by advanced endoscopic applications which provide a direct view, and by efficient bleeding control.

- **Bilateral choanal atresia is rare in adults**
- **It should be considered in persistent nasal obstruction cases**

We used a transnasal endoscopic approach in our case, and encountered no complications. In our patient, the atretic plaques were of the mixed type formed by

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both bone and membrane. Therefore, we used both soft tissue resection and bone drilling during surgical treatment.

Congenital choanal atresia is a rare entity in adulthood and few such cases have been reported. Yasar and Ozkul's description of a 51-year-old patient with congenital choanal atresia is one such report.[8] Our patient is notable in that she is to our best knowledge the oldest such patient reported in the recent English language literature.

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