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Radiology in Focus

Bilateral choanal atresia in an adult: is it compatible with life?

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Abstract
Bilateral choanal atresia is potentially life-threatening. The condition almost always presents in the new born, with alternating phases of respiratory distress and apnoea relieved by crying. We report a rare case of bilateral choanal atresia presenting for the first time at 22 years of age. The patient had no features of cyanosis or apnoea, presenting only with bilateral nasal obstruction, rhinorrhoea and anosmia. His neonatal history was unremarkable. No syndromic association was noted excepting for telecanthus. The presence of bony atresia was confirmed on computed tomography (CT) scan and transnasal endoscopic surgery was used to obtain a patent airway. This case is a rare report of bilateral choanal atresia presenting for the first time in adult life.

Key words: Choanal Atresia; Adult

Introduction

Congenital choanal atresia is an uncommon malformation with an estimated incidence of 1 in 5000 to 1 in 8000 live births, with a slight female preponderance. Atresia may be bony (90 per cent), membranous (10 per cent), unilateral or bilateral and complete or incomplete.[1] Ten to 50 per cent of cases show associated anomalies.[2,3] The commonest of which is the CHARGE syndrome (coloboma, heart defects, atresia choanal, retarded growth and development, genital hypoplasia, ear deformities).[4] Bilateral choanal atresia almost always presents with respiratory distress and cyanosis at birth. We report a rare case of congenital bilateral choanal atresia, presenting for the first time in early adult life.

Case report

A 22-year-old male patient presented with bilateral, nasal obstruction, mouth breathing, mucoid rhinorrhoea and anosmia. There were no episodes of respiratory distress and his neonatal history was unremarkable. There were no symptoms suggestive of any other abnormality and, on examination, there was no evidence of physical dysmorphism. Local examination revealed telecanthus and depression of the nasal bridge, and mucopurulent discharge was seen in both nasal cavities with no airflow at the anterior nares. An attempt to pass an 8-French rubber catheter into the nasopharynx was unsuccessful and a methylene blue test was negative. The diagnosis of bilateral choanal atresia was confirmed by a high-resolution axial CT scan that showed complete, bilateral bony atresia that was thicker on the left (Figure 1). A trans-nasal endoscopic approach was used to provide a nasal airway. Using a 0 degree nasendoscope a cruciate incision was made in the choana and a mucosal flap elevated over the atretic plate which was then removed using a 2.5 mm diamond burr tip. The neo-choana was stented with a No. 6 portex endotracheal tubes for six weeks and a check endoscopy was performed

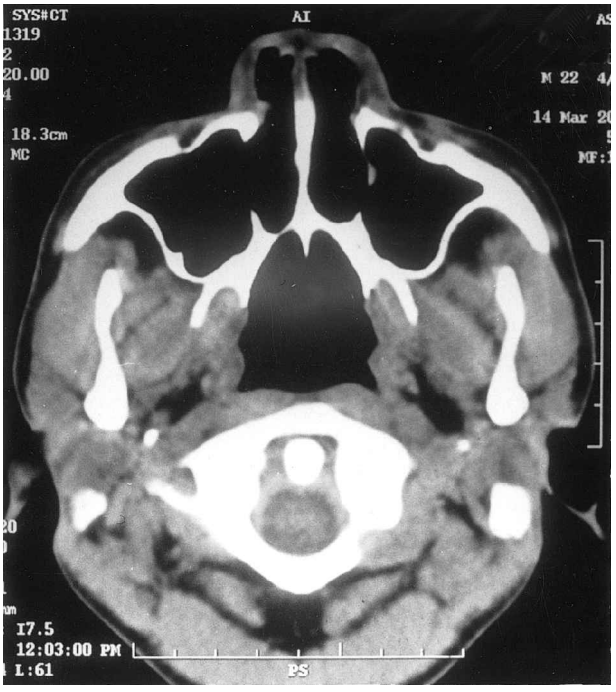

Fig. 1
CT scan nose and paranasal sinuses, axial sections, showing complete, bilateral, bony atresia at the level of posterior choana.

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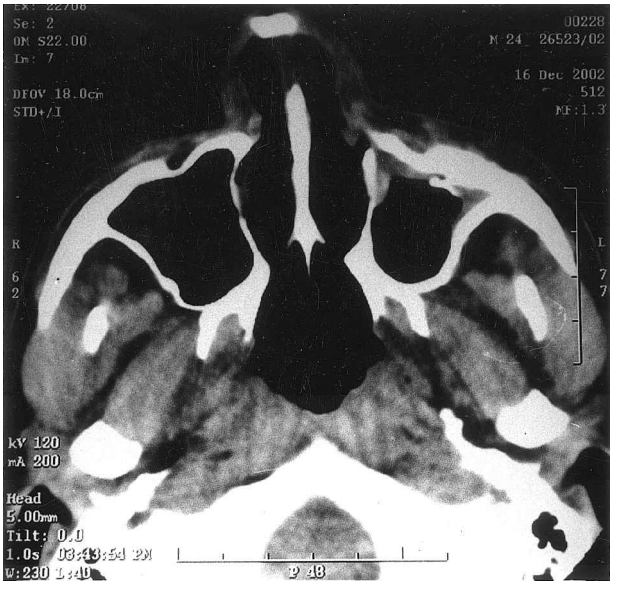

FIG. 2

Post-operative CT scan nose and paranasal sinuses showing patent posterior choanae after 12 months.

after the stent was removed. CT scan performed one year following the surgical procedure showed a patent and wide chona on both sides (Figure 2).

Discussion

Congenital choanal atresia was first described by Roderer[5] and the first reported case dates back to 1830 by Otto in Germany.[6] The atresia is bilateral in 60 per cent of cases when it is usually part of a generalized malformation of the central maxillofacial structures, with pyriform aperture stenosis, arched palate, encephalocele and a single central incisor. Craniofacial syndromes such as Down's, Treacher Collins, Crouzon, Apert, Pierre Robin and orofaciodigital syndromes have also been associated with choanal atresia.[7]

As neonates are obligate nasal breathers until four months of age the usual clinical presentation is of cyclical cyanosis and respiratory distress within a few hours of parturition. The symptoms are classically relieved by crying and worsened by suckling and the diagnosis is established by the inability to pass a 6/8 French rubber catheter into the nasopharynx. Confirmatory tests include the methylene blue test and CT scan of the skull base. Our case is unusual in its age and type of presentation since our subject presented for the first time at 22 years of age, having had no significant postnasal history of respiratory distress or intubation or surgery in infancy or early childhood. Moreover, no other congenital anomaly was noticed except for telecanthus and there was no similar condition in the family. Baker *et al.* have hypothesized that very rarely, a newborn with bilateral choanal atresia may compensate by rapidly learning mouth-breathing and diagnosis may therefore escape detection for months or years.[8] Reviewing the English literature, only one such case has been reported in a nine-year-old girl with bilateral choanal atresia of mixed type with Down's syndrome. She underwent transpalatal repair at one day of age but due to re-stenosis had to be subjected to endoscopic revision at the age of nine years.[9]

- **This paper is a case report of a 22-year-old male adult presenting with nasal symptoms who was found to have bilateral choanal atresia**
- **The patient had telecanthus but no syndromic association with atresia was noted**
- **Bilateral atresia in an adult has not been reported previously**

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