CONGENITAL CHOANAL ATRESIA

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ABSTRACT

Congenital choanal atresia (CCA) is the developmental failure of the nasal cavity to communicate with the nasopharynx. Bilateral choanal atresia is potentially life threatening in newborns. Most of these cases present early in life with cyclical phases of respiratory distress and apnoea, which gets relieved by crying. In fewer cases it goes undiagnosed to present in adult life with rhinorrhea and nasal obstruction. We report a rare case of bilateral choanal atresia presenting for the first time at 19 years of age with presenting with bilateral nasal obstruction, rhinorrhea and anosmia. His neonatal history was unremarkable. Endoscopy and CT scan confirmed atresia. The patient was successfully treated by transpalatal technique.

Key words: Choanal atresia, adult.

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INTRODUCTION

Congenital choanal atresia (CCA) is an uncommon malformation with an estimated incidence of 1:5000 to 1:10000 live births. Atresia may be bony, mixed bony-membranous or purely membranous. Most cases of choanal atresia are isolated malformation, but association with other congenital deformities has been reported in literature. Commonest among which is CHARGE Syndrome (coloboma, heart defects, atresia choanal, retarded growth, ear deformities). Unilateral atresia may go undiagnosed and unrecognized until later in life since respiratory distress is usually not encountered at birth. Bilateral choanal atresia is a life threatening condition in newborns. It is treated initially in newborn stage conservatively, then surgically later. However in rare cases like ours it may be diagnosed late in adult life with other symptoms.

CASE REPORT

A 19-year-old male patient presented with long standing bilateral nasal obstruction, mouth breathing, mucoid nasal discharge, and anosmia. There was no history suggestive of respiratory distress and his neonatal history was unremarkable. There was no symptom suggestive of any other anomalies. Local examination revealed the patient had elongated face, high arched palate and mucopurulent discharge in both nostrils with no airflow at the anterior nares. An attempt to pass a suction catheter into nasopharynx was unsuccessful. Diagnostic endoscopies revealed complete blockage of
both nostrils posteriorly. The diagnosis of bilateral choanal atresia was confirmed by axial CT scan that showed bilateral bony atretic plate with bony septal deviation posteriorly to left side (figure 1). A trans palatal approach was used to provide a nasal airway. Bilateral atretic plates were removed and the deviated bony spur was removed. The neo-choana was stented with no.5/6 portex endotracheal tube for 8 weeks (figure 2). Check endoscopies and local debridement was done every month for few months. On left side dilatation was required twice, which was done endoscopically. CT scan after 10 months of surgery revealed a patent choana on both sides (figure 3).

**DISCUSSION:**

Congenital choanal atresia was first reported in 1830\(^3\). Especially bilateral cases are usually part of various craniofacial syndromes\(^4\).

Bilateral choanal atresia causes acute life threatening respiratory distress in newborns within hours of parturition as neonates are obligate nasal breathers. The symptoms gets relieved by crying and worsened by feeding. The diagnosis is established easily in neonate age by the inability to pass a suction catheter into nasopharynx. However, CT scan of nose and skull base is confirmatory both for diagnosis and assessing the extent and thickness of atretic plate\(^5\).

Our case is unusual since he presented for the first time at 19 years of age with bilateral disease and with no significant history of postnatal respiratory distress or intubations or surgery in early childhood as reported by patient’s parents.

Baker et al have hypothesized that very rarely a newborn with bilateral choanal atresia may compensate by rapidly learning mouth breathing and therefore the

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**Fig 1:** Patient with stents in both nostrils.

**Fig 2:** C.T. scan of same patient.

**Fig 3:** C.T. scan showing bilateral choanal atresia.
restenosis remains an important problem. But, these have been attributed to local infection, local granuloma, foreign body reaction to stent and synechia. Although, transpalatal repair is a 'bigger operation', has some 'bigger' advantages like wider exposure, creation of larger neo-choana initially and decreased incidences of restenosis. In our case, since the patient was adult with thick bony atretic plate and posterior bony septal deviation, we preferred the trans-palatal method.

DISCLOSURES

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REFERENCE:
