Abstract: Bilateral choanal atresia is extremely rare in adults and only 4 cases have been reported in English literature. We report a case of 18 year old girl with bilateral complete choanal atresia who underwent endoscopic trans septal repair of the same.

Keyword: Bilateral choanal atresia in adult, choanal atresia

Bilateral choanal atresia in an 18 year old girl and endoscopic trans septal repair- a case report

Introduction

Choanal atresia is rare congenital anomaly in which there is occlusion of posterior nasal orifice and absence of communication between nose and nasopharynx. It can be unilateral or bilateral. Incidence is 1 out of 5000 to 7000 live births (2). 50% of cases are associated with other congenital anomalies (3). Choanal atresia can be incomplete or complete. Since newborns are obligate nasal breathers complete bilateral choanal atresia is a potentially life threatening condition unless diagnosed and managed immediately after birth. Bilateral choanal atresia is extremely rare in adults (3,5,6) and only 4 cases have been reported in English literature. We report a case of 18 year old girl with bilateral complete choanal atresia and endoscopic trans septal repair of the same. Case report An 18 year old girl presented to the Out Patient department with history of bilateral nasal obstruction and nasal discharge since childhood. She also had history of difficulty in feeding after birth, mouth breathing and snoring and hyponasal voice since childhood. There was no history of any other congenital anomalies. There was no history of birth asphyxia or cyanotic spells in infancy. There were no other remarkable complaints during childhood and adolescence. She had no relevant family history. Picture of the patient General examination revealed hyponasal voice or rhinolalia clausa and had all typical features of a chronic mouth breather like elongated face, open mouth, crowded teeth, hitched up upper lip, pinched in nose and high arched palate. Anterior rhinoscopy revealed secretions in floor of both nasal cavities and atrophied small turbinates. Diagnostic nasal endoscopy showed atresia of posterior nasal aperture on both sides and atrophied turbinates. CT scan of the paranasal sinus showed bilateral complete bony choanal atresia. Detailed examinations and investigations by ophthalmologists, gynaecologist and nephrologist revealed no other congenital anomalies such as CHARGE syndrome.
Our patient underwent endoscopic trans-septal repair of choanal atresia under general anaesthesia. A septal incision was made and mucoperichondrium was elevated on both sides as in sub mucous resection of septum and mucosa over the atretic plates upto lateral wall is elevated on either side along with the septal perichondrium and septal periostium. Posterior bony septum was removed and atretic bone drilled out on both sides. Mucosa removed using debrider. A wide common posterior nasal aperture was created. Mucosa covered over the remnant bony parts. No stent or catheters were placed. Patient started nasal breathing immediately after the surgery. Patient was discharged after 2 days and daily saline nasal irrigation was recommended. Follow up evaluation after 3 weeks showed patent choana and the patient had become a complete nasal breather after 18 years of mouth breathing. Snoring had disappeared completely. The patient is on regular follow up for after 3 weeks of surgery.

Intra operative picture showing endoscopic trans-septal elevation of mucosa from vomer and atretic plates

CT Scan (coronal PNS) showing collected secretions in the floor of nasal cavity and atrophic turbinates

Picture of left nostril showing patent opening after 3 weeks of surgery

Discussion

Choanal atresia was first described by Johann Roderer in 1755(1). Choanal atresia can be described as failure to develop the communication between the nasal cavity and nasopharynx. The incidence is 1 in 5000 to 7000 live births(2). Females are twice commonly affected than males. Unilateral defects are more common than bilateral. The atretic plate can be bony (20%), membraneous (10%) or mixed (70%)(4). Choanal atresia may present as an isolated malformation or in 20% to 50% of cases(3), part of a group of congenital malformations, such as CHARGE syndrome comprising coloboma of iris, heart malformation, mental retardation and growth, genital anomalies and ear anomalies and deafness. Other anomalies associated with choanal atresia are cleft lip and palate, abnormality of tongue, facial palsy, laryngeal malformation, oesophageal atresia, tracheoesophageal fistula, renal and skeletal abnormalities. So a complete evaluation is needed by otorhinolaryngologist, paediatrician, ophthalmologist, gynaecologist and nephrologist. Various theories are postulated regarding the origin of this anomaly. The most accepted theories are Persistence of oral-pharyngeal membrane, Failure to break the physiological oronasal membrane of Hochstetter, Adherence of abnormal mesodermal tissue located in the nasal choana.

Medial growth of vertical and horizontal processes of palatine bone Unilateral congenital choanal atresia present as unilateral nasal obstruction and nasal discharge. In many cases it may go unnoticed and present in adult life. Bilateral congenital choanal atresia is considered as life threatening condition unless diagnosed and corrected immediately after birth. Newborns are obligate nasal breathers and bilateral choanal atresia present as asphyxia in the neonates. Two types of asphyxia in newborns are described one is cyclical type in which the child cries which facilitates mouth breathing; later the child get exhausted and goes to sleep which produce respiratory distress and cyanosis and again with crying, the symptoms are relieved. The second type is suckling type in which the child develops asphyxia and cyanosis when suckling. The child will be relatively normal other times. The diagnosis is based on high level of suspicion and diagnostic tests. The various tests used are probing with catheter, cold spatula test, contrast radiography, instillation of dyes into the nose and observe the nasopharynx etc. The gold standard tests are diagnostic nasal endoscopy and computerised tomography. Immediate surgical management is necessary in congenital bilateral choanal atresia. The various surgical approaches are trans palatal excision and stenting, trans nasal endoscopic excision with or without stenting and trans septal approach. Oral airway, endotracheal intubation or tracheostomy may be needed to maintain the airway till the definite surgery is performed. Presentation of congenital bilateral choanal atresia in adult life is extremely rare(3,5,6). Only four cases have been described in world literature(3,5,6). Panda et al(3) reported a 22-year-old patient with bilateral choanal atresia. The passage in this patient was established via a transnasal endoscopic approach. There were no other congenital abnormalities in this patient. Yasar and Ozkul(6), reported a 51-year-old patient with bilateral choanal atresia. Repair was done by transnasal endoscopic approach. El-Sawy(5) et al reported a 24-year-old patient with bilateral nasal obstruction and total loss of sense of smell and hypogammaglobulinaemia. Computerised tomography of this patient revealed aplasia of frontal and sphenoid
Adequate opening was achieved with transnasal endoscopic approach; however, restenosis requiring a second operation occurred. Fadalullah et al reported a 23 year old girl with history of nasal obstruction and nasal discharge. Endoscopic examination and paranasal sinus tomography revealed bilateral choanal atresia. She did not have any other congenital abnormalities. She underwent endoscopic trans nasal repair and mitomycin C without stent.

Our case is fifth in the literature, an 18 year old girl with history of bilateral nasal obstruction and nasal discharge. Paranasal sinus tomography and endoscopic examination revealed bilateral choanal atresia. She did not have other congenital abnormalities. She underwent endoscopic trans septal repair without stent. Conclusion Congenital bilateral choanal atresia is a rare life threatening anomaly in newborn. Immediate diagnosis and management is necessary. Presentation of congenital bilateral choanal atresia in adult is extremely rare. An 18 year old girl with history of nasal obstruction and mouth breathing since birth was found to have congenital bilateral choanal atresia and endoscopic trans septal repair was done. Follow up evaluation after 2months showed patent posterior nasal orifice.

References