ABSTRACT

Bilateral choanal atresia is an otorhinolaryngology emergency. It is a potentially life-threatening disorder because the affected newborn is an obligate nose breather. We report clinical and radiological evaluation of a patient with congenital bilateral choanal atresia, who presented at the 20-year of age with other congenital anomalies. Patient was diagnosed as congenital bilateral choanal atresia, right accessory nasal deformity, high arched palate with congenital ptosis bulbi of his left eye. He was posted for transnasal endoscopic surgery under general anesthesia. Unique feature of this case is left congenital ptosis bulbi along with bilateral congenital choanal atresia probably the first case report having both anomalies in a single patient. While performing surgery, we should carefully study the anatomy of the basisphenoid, as in our case we did not attempt to correct atresia on right side.

Keywords: Congenital choanal atresia, Congenital ptosis bulbi, Transnasal endoscopic sinus surgery.


Source of support: Nil

Conflict of interest: None

INTRODUCTION

Bilateral congenital choanal atresia is considered a lethal congenital malformation as newborns are generally obligate nasal breathers for the first few months of life so, it will present as an acute respiratory emergency at birth with classical picture of cyclical cyanosis (blue spells relieved by crying). We present a case of 20-year aged male with bilateral congenital choanal atresia with congenital right accessory nasal deformity, high arched palate and left congenital ptosis bulbi.
The stent was fashioned from portex endotracheal tube cut to length. The stent were secured by a circumseptal 0′ prolene suture and were left in situ for 6 weeks. After 6 weeks, portex tube was removed and on nasal endoscopy well epithelialized posterior choanae were noted on left side. Rhinolalia clausa and sleep snoring was all markedly relieved. Patient is on regular monthly follow-up presently (Fig. 5).

DISCUSSION

The etiology of congenital atresia is unknown. Three embryologic theories have been advanced the most logical being that a medial overgrowth of the vertical and horizontal process of the palatal bones results in choanal atresia. This theory, however, fails to explain membranous choanal atresia. It is of interest that associated congenital anomalies occur with congenital choanal atresia in 43% of the case reports. Overall, incidence of choanal atresia being 1 in 7000 live births, 90% of atresia are bony while 10% being membranous.

Bilateral choanal atresia is an otolaryngology emergency. It is a potentially life-threatening disorder because the affected infant is an obligate nose breather. Symptoms range from intermittent to severe respiratory distress with cyanosis that is aggravated by feeding and alleviated by crying. Approximately, 75% of children with
bilateral choanal atresia have other congenital abnormalities, as exemplified by Charge syndrome (coloboma of eye or microphthalmia, heart malformation, choanal atresia, retarded growth, genital hypoplasia, and ear abnormalities, typically external).\textsuperscript{4} In our case, patient did not had respiratory difficulty after the birth because of other congenital anomaly of palate provided the compensatory mechanism facilitating the oropharyngeal respiration. Surgical correction of bilateral choanal atresia is performed as soon as possible after the diagnosis has been made; however, there is little consensus regarding the optimal surgical approach.\textsuperscript{5} Transpalatal, transnasal, transeptal, and sublabial approaches have been utilized with varying degrees of success.\textsuperscript{5} Recently, the transnasal endoscopic procedure has been advocated as a safe and efficacious method with the best possibility for long-term nasal patency.\textsuperscript{5,6}

Congenital accessory nasal deformity is a supernumerary malformation in the external nose. Supernumerary malformation may appear in different ways. It may replace the normal structure or co-exist with it. Removal of the congenital accessory is the optimal treatment method.\textsuperscript{7} The patient in our report bore three anterior nares, however, this should be differentiated from three-nostril malformation, a type of cleft nose occurring when the median line of the nasal ridge presents as a major groove. In our case, only the soft tissue formed the malformation in one side of the anterior nares, therefore, this case was diagnosed as ‘congenital accessory nasal deformity’.

**CONGENITAL PHTHISIS BULBI**

Phthisis bulbi as a result of some suppurative traumatic or non-traumatic uveal inflammation is not a very rare condition, but shrinking of the globe as a result of uveal inflammation \textit{in utero} is very rare indeed, and deserves to be recorded in medical literature.\textsuperscript{8} Congenital microphthalmia should be distinguished from phthisis bulbi, which usually represents an acquired shrinkage of the eye following trauma, inflammation, and so forth rather than a primary development defect. The standardized A-scan and B-scan echography help us to distinguish the phthisis bulbi from atrophy of the eyeball without shrinkage and atrophy of the eyeball with shrinkage.

**CONCLUSION**

Congenital bilateral choanal atresia is a life-threatening disease in newborns. However, it can also be found in adults, who may present with mouth breathing, sleep snoring and symptoms of nasal obstruction. Choanal atresia is often accompanied by other congenital abnormalities. In our case, we have a 20-year aged patient with congenital bilateral choanal atresia with right supernumerary accessory nostril with right external nose deformity along with high arch palate. But unique feature of our case is left congenital phthisis bulbi along with bilateral congenital choanal atresia probably the first case report having both anomalies in a single patient. While performing surgery, we should carefully study the anatomy of the basisphenoid, as in our case we did not attempt to correct atresia on right side.

**REFERENCES**