Case report

Isolated Congenital Bilateral Choanal Atresia and Nasopharyngeal Atresia- a case report.

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Abstract:

A term neonate developed respiratory distress, paradoxical cyanosis (relieved by crying) soon after birth. Inability to insert No.5 French infant feeding tube through the nose into the pharynx led to the diagnosis of Bilateral Choanal Atresia , which was confirmed by HRCT Scan of the nose. Insertion of an oropharyngeal tube reduced the respiratory distress. Extensive investigations did not reveal any other congenital anomaly. The baby was treated with Transnasal Surgery.

Keywords: Choanal atresia, oropharyngeal tube, transnasal surgery.

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Introduction:

Choanal atresia is a rare life threatening disorder in neonates, (incidence 1 in 7000 live births[1]), but it is the most common indication of surgical intervention of the nose in neonates[2]. Persistence of Bucconasal membrane of Hochstetter or Buccopharyngeal membrane of the foregut (ruptures by fifth or sixth weeks to form the choanae) results in choanal atresia[3]. Presently the theory of misdirection of neural crest migration and subsequent mesodermal flow is thought to offer the strongest evidence behind the development of choanal atresia[4]. Neonates are obligate nasal breathers due to the nasoraceptor reflex[5]. Neonates with bilateral (B/L) choanal atresia will most often not attempt to breathe through the mouth-resulting in asphyxia[6]. Some authors hypothesized that the disorder is of autosomal dominant inheritance with reduced penetrance and variable expressivity[7] while others support the role of denovo mutations[8]. The diagnosis and management of this rare emergency in a symptomatic neonate with bilateral choanal atresia requiring transnasal repair at a very early postnatal age is presented.

Case Report:

Following an uncomplicated pregnancy, a lady delivered a male infant at term by spontaneous vaginal delivery. The lady had no addiction and was not on any medication during pregnancy. Apgars at 1 and 5 minutes were 8 and 9. However the newborn started to have respiratory distress soon after birth. There was tachypnoea (respiratory rate 65-70/ min), suprasternal suction, stridor, nasal flaring, chest retractions and cyclical cyanosis. During crying, bilaterally equal vesicular breath sounds was heard and there was significant amelioration of cyanosis. There was no dysmorphism, no other external congenital anomalies; anthropometric parameters were normal. Presence of intermittent cyanosis- aggravating during sleep and resoluting during crying, prompted us to exclude the possibility of choanal atresia. Attempts to pass a No.5 French infant feeding tube and a red rubber catheter per naris was unsuccessful. On holding a metallic spatula below the infant's nasal aperture there was no misting– hence a diagnosis of bilateral choanal atresia was provisionally made.

A 0-0 size oropharyngeal tbe was inserted with subsequent resolution of respiratory distress and

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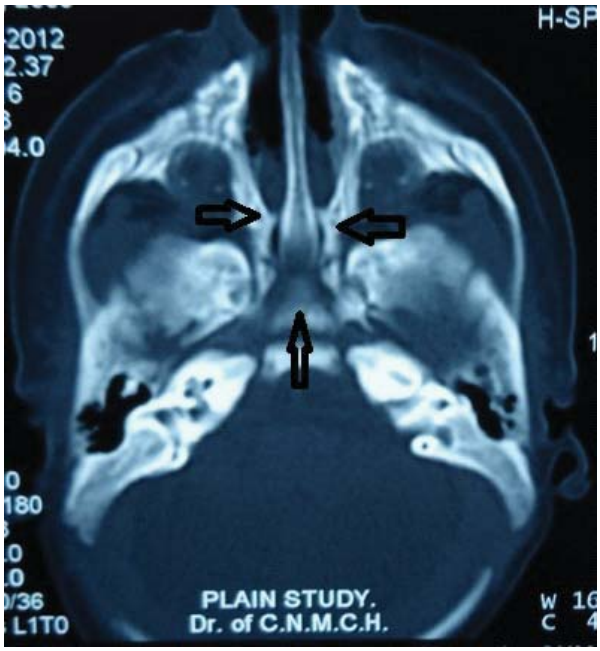

Figure-1: HRCT showing hypertrophy of the posterior end of the Vomer almost abutting with the pterygoid plates of the Sphenoid laterally (shown by horizontal arrows) and the body of the Sphenoid posteriorly (shown by vertical arrow).

cyanosis. Oxygen saturation (SpO2) ranged between 90-95 %. Initially intravenous fluids were given, then feeding was started with expressed breast milk via orogastric tube. High Resolution Computed Tomography Scan (HRCT) of the nose, paranasal sinuses and skull base (after suction clearance of the nose and application of decongestant drops)revealed marked hypertrophy of the posterior end of the Vomer almost fusing with the body of the Sphenoid with resultant choanal and nasopharyngeal atresia. Chest skiagram, Echocardiography, Renal ultrasound, Fundoscopy and Brainstem Auditory Evoked Response were normal.

Figure-2: HRCT showing absence of air lucencies in nasopharynx- suggesting nasopharyngeal atresia. 'nc' signifies nasal cavity, 'oc' signifies oral cavity

After stabilization, the baby was taken up for surgery on the tenth day of life. In Transnasal endoscopic surgery,patency of the choana was established by drilling the atretic plate. The choanal diameter was increased with a bone nibbler and debridger. Temporary nasal stents of Portex tube were inserted and secured by circumseptal '0' Prolene suture.

Meticulous stent care by regular saline irrigation and suction clearance was done in the post operative period to prevent stent blockage. At the time of discharge on the tenth postoperative day, the baby was breastfeeding well. He was examined weekly on outpatient basis and advised antibiotics during this period to prevent infections. The stents were removed six weeks after surgery. During follow up over six months, the baby normally gained weight and achieved age appropriate developmental milestones. Fortunately the anticipated post-operative complications like sleep apnoea, persistent nasal stuffiness and rhinorrhoea did not occur. The baby did not require any revision surgery.

Discussion:

In 90% of cases, the obstruction has a bony component, whereas in 10% cases it is purely membranous[9]. Among those patients with a bony component of choanal atresia, only 30% has purely bony obstruction, whereas 70% have both bony and membranous obstruction[10, 11]. In our case, the purely bony bilateral obstruction was caused by excessive hypertrophy of the posterior end of the Vomer, its fusion with the Sphenoidal body resulted in nasopharyngeal atresia. Bilateral atresia is rarer than unilateral varieties[12] (65-75% of cases are unilateral[3]).

Neonates with choanal atresia should be evaluated for other congenital anomalies like CHARGE syndrome (Coloboma, Heart abnormalities, Atresia choanae, Growth and developmental retardation, Genital and

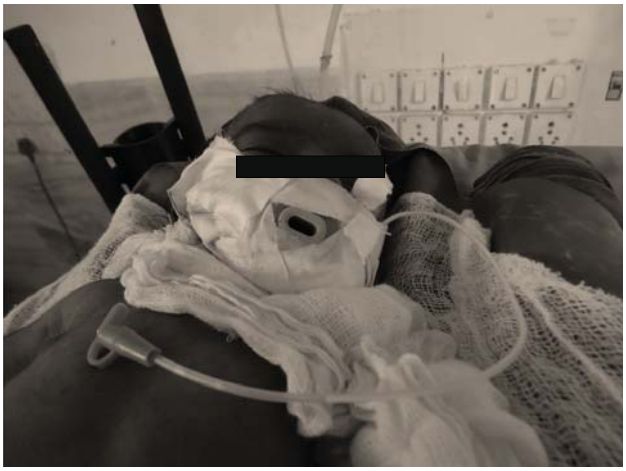

Figure-3: Picture of the baby with oropharyngeal tube and orogastric tube in situ.

Ear abnormalities), cerebral abnormalities, trachoemalacia, subglottic stenosis, nasal cavity stenosis and high arch palate. Associated multiple abnormalities are found more commonly in cases of bilateral atresia[13] - incidence being as high as 50-60% [2, 3,14, 15,16]. However in our case with bilateral atresia, extensive evaluation did not reveal any other congenital anomaly. Some delay may occur to allow a complete workup. Establishment of an adequate airway is of utmost importance to prevent intermittent cyanosis and hypoxia till further management with corrective surgery is accomplished[17]. Such babies can be made to learn mouth breathing using Mc Govern nipple [18]. Tracheotomy should be considered in cases of bilateral atresia in which the child has other potential life threatening problems and early surgical repair is not feasible[13].

The surgical management of choanal atresia is a challenging endeavour in the domain of Pediatric Otorhinolaryngology. Apart from transnasal procedure, transpalatal procedure can also be done. Transnasal surgery is associated with lesser chances of bleeding and palatal deformation. However limited field of vision, risk of injury to the Sphenopalatine artery, inability to adequately remove vomerine septal bone and displacement of indwelling stent are its limitations[1,3,19]. Occassional nasal dilatation and revision surgery is needed after stent removal[3]. Use of antineoplastic agents (Mitomycin C) to inhibit granulation tissue formation has been described[1]. However no such endeavours were necessary for the reported case. Addiction to cigarette and caffeine, use of medications like Methimazole, steroids, Antiepileptic drugs, Non Steroidal Anti Inflammatory Drugs, Folate antagonists during pregnancy are implicated as causes of choanal atresia [20,21]. However no such history could be elicited in our case.

Conclusion:

the outstanding aspects, which need to be emphasized, are that the baby was born to a lady without any risk factors, with purely bony variant of congenital choanal and nasopharyngeal atresia, without any other birth defects and had excellent outcome following surgery.

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