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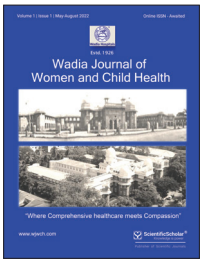

Case Series

Rare causes of congenital neonatal nasal obstruction leading to respiratory distress at birth and its management: A case series

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ABSTRACT

Neonatal nasal obstruction may present as an acute respiratory emergency. Neonates primarily are obligate nasal breathers. Hence, any nasal obstruction whether arising due to skeletal or anatomical anomalies or due to nasal mass or cyst can present with cyanosis, stridor, feeding problems, and/or failure to thrive. Although choanal atresia remains one of the most diagnosed nasal anatomical anomalies leading to this problem; other rare causes of neonatal nasal obstruction, namely, midnasal stenosis, pyriform aperture stenosis, congenital nasal mass, or cyst should also be considered. Hence, a thorough evaluation with diagnostic nasal endoscopy along with an imaging modality (computed tomography/magnetic resonance imaging) is warranted. Management whether conservative or surgical intervention is guided by the severity and nature of the symptoms. This series highlights our experience of four different case scenarios of respiratory distress arising out of nasal anatomical anomalies and its subsequent management and outcome.

Keywords: Congenital nasal obstruction, Midnasal stenosis, Pyriform aperture stenosis, Nasopharyngeal teratoma, Choanal atresia

INTRODUCTION

Respiratory distress in the newborn is an emergency and requires prompt attention. A thorough evaluation is essential in identifying the cause of the distress. Variable factors and causes may lead to respiratory distress in newborn patients and neonatal nasal pathologies remain one of the rarest and most often underdiagnosed conditions.[1] Midnasal stenosis is a rare entity encountered secondary to excessive growth of the nasal bones halfway along the nasal cavity. It is commonly associated with syndromes characterized by midfacial hypoplasia, namely, Apert syndrome, but sporadic cases have also been reported.[2] Pyriform aperture stenosis, on the other hand, arises due to bony overgrowth of the nasal process of the maxilla. It is most often associated with epiphora, a "mega incisor" or prosencephaly.[3] A nasopharyngeal teratoma often presents as a firm mass and is commonly associated stillbirth, prematurity, and polyhydramnios. Maternal serum alpha-fetoprotein levels and beta-human chorionic gonadotropin levels may be raised. Choanal atresia

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may be of bony or membranous type; however, in 70% of the cases, a mixed picture is seen. A newborn with bilateral nasal obstruction may present immediately after birth with respiratory distress, noisy breathing (stertor), and cyanosis during feeds and failure to thrive. Diagnosis can be confirmed with nasal endoscopy along with an imaging modality like computed tomography (CT scan). Treatment is guided by the severity of symptoms. This article presents four different rare cases of neonatal nasal obstruction encountered by us and its surgical approach and outcome.

CASE 1

A full-term male neonate with dysmorphic facial features, weighing 3.1 kg at birth, second child born through lower segment cesarean section presented at 5 hours of life with respiratory distress. The patient was initially put on oxygen support through nasal prongs but could not maintain oxygen saturation. A provisional clinical diagnosis of choanal atresia was made due to the inability to pass an infant feeding tube through both nostrils and a cold spatula test showing no mist formation below both nostrils. The neonate was intubated with a size 3.5 endotracheal tube and stabilized. Dysmorphic features in the form of protruding forehead, prominent and protruding eyeball, broad nasal bridge, micrognathia with retrognathia, and syndactyly of all 4 limbs were seen. CT scan of the brain and paranasal sinuses revealed mid-face hypoplasia with bilateral posterior choanal and mid-nasal stenosis along with midline cleft in the soft palate [Figure 1]. There was also a premature fusion of bilateral coronal sutures with harlequin deformity of bilateral orbit, exophthalmos, and brachycephaly. These findings raised the possibility of Apert syndrome. Under general anesthesia, a diagnostic nasal endoscopy (DNE) was undertaken using a 2 mm 0° endoscope as 4 mm scope was non-negotiable. DNE showed bilaterally hypertrophied inferior and middle turbinate and a bony narrowing of both sides about midway inside nasal cavities. The bilateral inferior turbinate was fractured and redundant mucosa over the inferior and middle turbinate cauterized to create midnasal space. Stenosed choanae was dilated using urethral sound and posterior septectomy was done to create adequate postnasal space. A size 3 endotracheal tube was refashioned and inserted as a nasal stent and left for 6 weeks [Figure 2]. Stent was kept patent with instillation of topical saline drops and frequent suctioning. After 6 weeks, the stent was removed. The child started having respiratory distress after stent removal and a DNE showed prolapsing redundant nasal mucosa. Hence, a new nasal stent was reinserted and kept for another 6 weeks. The patient was on regular follow up and after 3 months, his nose remains patent.

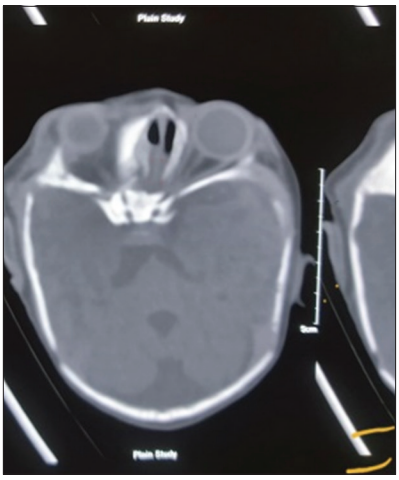

Figure 1: CT axial section showing midnasal and posterior choanal stenosis.

Figure 2: A clinical photograph of an infant with Apert's syndrome after repair of midnasal stenosis with nasal stent *in situ*.

CASE 2

A preterm female neonate was brought to the emergency room on the 2nd day of life with stridor since birth. The patient was intubated but a nasogastric tube could not be negotiated. Magnetic resonance imaging (MRI) revealed a mass lesion obstructing the naso-oropharyngeal lumen [Figure 3]. The mass was excised in toto with a transnasal endoscopic approach [Figure 4]. No nasal stents were used as the nasal and postnasal space was adequate. To avoid nasal crusting during the postoperative period, we advised frequent instillation of saline nasal drops along with gentle suctioning through the nasal stent. Histopathological examination showed a nasopharyngeal teratoma. DNE done 6 months after surgery showed a patent nasal cavity and nasopharyngeal space.

CASE 3

A full-term female neonate was admitted to the neonatal intensive care unit (NICU) with complaints of

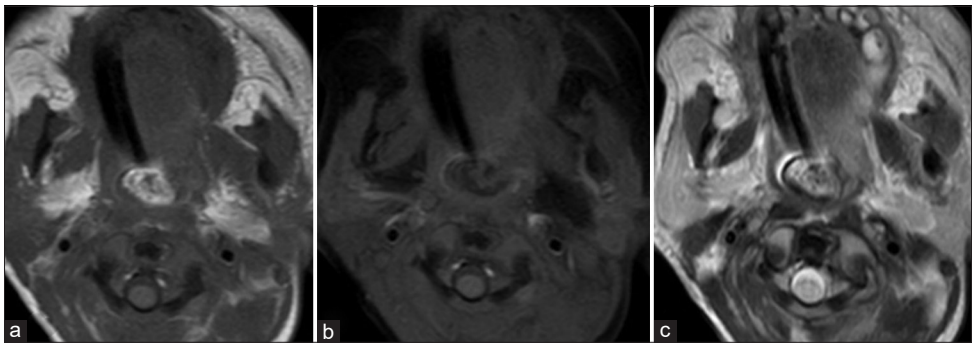

Figure 3: Axial T1-wieghted (a), T1 fat-saturated (b) and T2-wieghted (T2W) (c), images showing a well-defined T1 hyperintense lesion which is suppressed on T1 fat-saturated image suggesting fat content and has mixed hypo and hyperintense signal on T2W images. The lesion is abutting the free edge of soft palate anteriorly and posterior wall of naso-oropharynx, also note it causes obstruction to naso-oropharyngeal lumen.

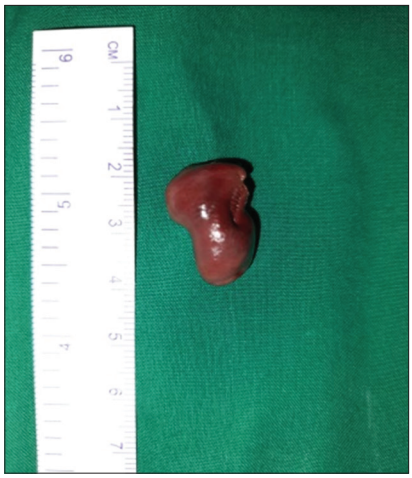

Figure 4: Gross specimen of nasopharyngeal teratoma.

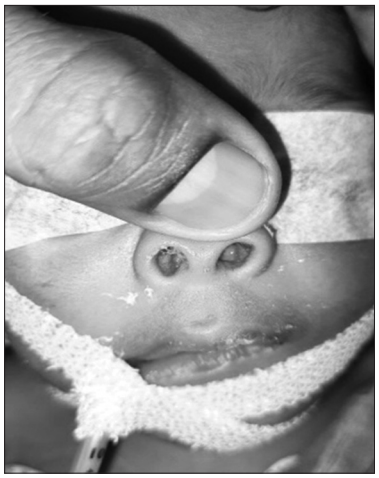

Figure 5: A clinical photograph of a neonate with pyriform aperture stenosis.

feeding difficulty and cyanosis during feeds that were relieved by crying. The inability to pass a nasogastric tube through the nasal passageway raised the suspicion of choanal atresia. Anterior rhinoscopy revealed a stenosis of the pyriform aperture [Figure 5], and the diagnosis was confirmed with a CT scan which showed an aperture width of <5 mm at the level of the inferior meatus. There were no other associated abnormalities. Surgical intervention was undertaken with a sublabial approach and the mucoperiosteal flap was elevated. The overgrown nasal process of maxilla was drilled away and the mucoperiosteal flap was reposited. Postoperatively, nasal stents were used for 6 weeks [Figure 6]. The patient was extubated 48 hours after surgery and did not require further respiratory support. Follow-up DNE done after 6 weeks showed synechiae formation between the inferior turbinate and septum but the child was asymptomatic.

CASE 4

A preterm male neonate required oral airway right after birth in view of respiratory distress and cyanosis and was subsequently intubated and shifted to NICU. NICU staff was unable to pass a nasogastric tube through both nostrils. The cotton wool test and cold spatula test were inconclusive. A diagnostic nasal endoscopy pointed toward bilateral choanal atresia, which was further confirmed with a CT scan as a bilateral bony type of choanal atresia. Echocardiography, ultrasound kidneys, ureters and bladder, and MRI brain ruled out other associated anomalies. Surgical intervention was undertaken with a transnasal approach. The nasal cavity was decongested with adrenaline-soaked cotton patties. The posterior part of septum and atretic choanae was infiltrated with 2% xylocaine-adrenaline. As the nasal cavity was narrow, bilateral inferior turbinate was fractured and reduced

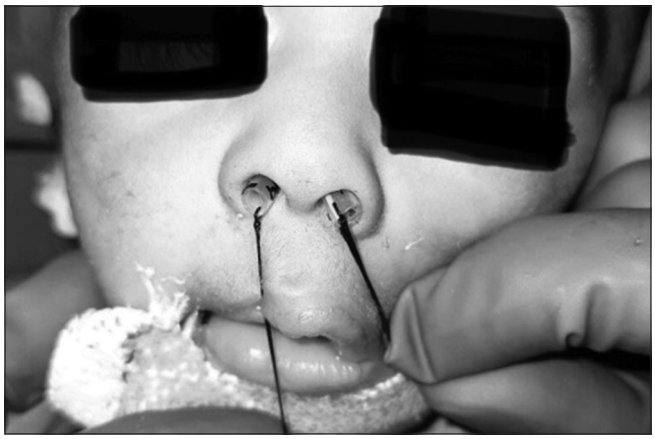

Figure 6: Clinical photograph of an infant highlighting the use of nasal stents after surgical repair of pyriform aperture stenosis.

to create space for the introduction of instruments. Urethral sound was used to poke into the atretic plate inferomedially. Posterior septectomy was performed. Postoperatively, nasal stents were used for 6 weeks, along with regular topical instillation of sterile saline nasal drops. Follow-up DNE done after 6 weeks showed a patent postnasal space.

DISCUSSION

A newborn with complete bilateral nasal obstruction may present with respiratory distress at birth as all children are obligate nasal breathers for the first few months of life. Choanal atresia has been reported as the most common cause of congenital nasal obstruction followed by pyriform aperture stenosis, midnasal stenosis, nasal agenesis, and nasal cyst.[4] A provisional clinical diagnosis is made after an inability to pass an infant feeding tube through the nostril. The cotton wool or cold spatula test are other modalities to demonstrate nasal obstruction. Confirmation of clinical diagnosis requires an imaging modality preferably CT (nose and paranasal sinuses) to determine the site, degree, extent of the obstruction, and the nature of obstruction (bony/membranous/mixed). Initial management to relieve respiratory distress includes use of McGovern nipple or Guedels airway to keep the mouth open. In case of severe distress and failure of above measures to relieve symptoms, the patient might require endotracheal intubation until a surgical plan is executed.

Treatment of midnasal stenosis and pyriform aperture stenosis is usually conservative, allowing the child's midface to grow, such that by the age of 6 months the obstruction is relieved. Surgery is indicated if there is failure of conservative management, feeding difficulties with cyanosis, repeated extubation failure, and failure to pass a size 5 French Gauge catheter through the nose.[5,6] Choanal stenosis can be repaired through the transnasal and transpalatal approaches, but the sublabial, transantral, and transseptal approaches have also been described.[7] To keep the nasal passage patent and to prevent restenosis; a nasal stent is inserted in the immediate post operative period. However, the duration of nasal stenting varies, and the role of nasal stent post-surgery is also debated.[8,9] During the post-operative period to avoid nasal crusting frequent, instillation of saline nasal drops was advised along with gentle suctioning through the nasal stent.

CONCLUSION

Respiratory distress resulting from neonatal nasal obstruction is a grave emergency and a correct diagnosis by means of imaging and nasal endoscopy is essential to identify the exact site of obstruction. For children with significant respiratory difficulty, failure to thrive, failure to wean off oxygen, or failure of conservative management, surgical intervention with or without stent placement can be considered.

Ethical approval

All procedures involving human participants were in accordance with the ethical standards of the institution.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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