Introduction

Congenital nasal pyriform aperture stenosis (CNPAS) is an uncommon cause of neonatal upper airways obstruction. It was first described by Douglas in 1952 and the first clinical case of CNPAS was described by Brown et al in 1989 [1,2]. The etiology is unknown but it appears that because of the overgrowth of the nasal process of maxilla in the fourth month of fetal development. Clinically, a child with a mild form of CNPAS often has noisy breathing but is still able to maintain his airway and feed appropriately. However, a newborn who has a severe form of CNPAS will often present with significant respiratory distress that needs oxygen support or even intubation. It can be manifested with episodic apnea, cyclic cyanosis, stridor, poor feeding, and failure to thrive.

Case report

A full-term female baby with weighed 2.27 kg was born at 38 weeks via spontaneous vertex delivery with a good Apgar score. The antenatal period was uneventful. The neonate developed respiratory distress at 11 hours of life when she developed noisy breathing, tachyypnea with a moderate intercostal recession, and soft monophasic stridor. She was then transferred to the neonatal intensive care unit for close observation and was put on nasal Duo positive airway pressure (DuoPAP). Initially, she was treated for congenital pneumonia. She was completed IV c-penicillin for 1 week and IV gentamycin for 3 days, however, she still unable to wean off oxygen. Thus, she was referred to ear, nose, and throat (ENT) to rule out upper airway abnormalities. Clinical examination revealed soft dysmorphic features with a small and receding chin. There was no obvious mass seen at the nostril and no evidence of cleft palate. A cold spatula test was performed, however, we noted reduced mist over bilateral nostril. Flexible nasoendoscopy was attempted, but the scope could not be passed through the bilateral nostril. A nasogastric tube size 6Fr also could only be passed up to 1.5cm in both nostrils. CT scan of...
paranasal sinus and neck was performed to look for any evidence of airway obstruction causing failure to wean off oxygen support for almost one month. CT scan revealed typical findings suggestive of CNPAS. There was a stenosis of the pyriform aperture, which measured 5.9 mm in width on an axial image, at the level of inferior turbinate (Figure 1), single central maxillary mega incisor (Figure 2), and triangular hard palate (Figure 3).

Given the persistent symptoms and failure to wean off oxygen support, the infant underwent surgery at 28 days of age. Through a sublabial approach (Figures 4 and 5), the stenotic pyriform area was widened by drilling the bone using a 1 mm diamond bar, taking care to avoid injury to the tooth socket and nasolacrimal duct (Figure 6). The inferior concha was dislocated laterally. Two soft nasopharyngeal airways were placed as a stent to maintain airway patency and to avoid restenosis (Figure 7). Oxygen support could be off after 10 days post-operative. The nasal stents were removed on postoperative day 12. She was discharged home with no complications. During her follow-up, she was doing well, no more noisy breathing and her weight is gaining.

Figure 1. Axial view of CT paranasal sinus on the bone window showing the nasal pyriform aperture stenosis at the level of the inferior meatus, which measured 5.9 mm in width.

Figures 2 A and B. CT image showing central maxillary mega incisor (yellow arrow)
Figure 3. Axial view CT of paranasal sinus on the bone window. Yellow arrow showed triangular hard palate

Figure 4. Gingivobuccal sulcus incision made through sublabial approach
Figure 5. Sublabial flap raised until exposed the anterior nasal spine (yellow arrow) and bilateral nasal process of the maxilla (blue arrow)

Figure 6. Bilateral nasal process of maxilla drilled using skeeter drill size 1.0 mm
Figure 7. Nasopharyngeal airway was placed in each nasal cavity as a stent

Discussion

Neonate is an obligate nasal breather till 6-8 weeks of life, thus any form of nasal obstruction will lead to respiratory distress with a spectrum of severity. Congenital nasal airway obstruction affects up to 1 in every 5,000 newborns, with choanal atresia being the most common cause. CNPAS is thought to be around one-fifth (1 in 25,000 live births) to one-third the incidence of choanal atresia [3]. CNPAS is a rare cause of nasal obstruction in neonates but should always be a differential cause in our minds. Some other causes that may lead to nasal obstruction are meningocele, meningoencephalocele, dermoid and epidermoid cysts, sinonasal tumors of any origin, septal dislocation or hematoma, choanal atresia, and dacryocystocele [4,5,6]. The most common among them is choanal atresia which is more posterior (approximately 3 cm from anterior nares) while the level of obstruction in CNPAS is more anterior (approximately 1 to 1.5 cm) and there will be difficulty in inserting a suction catheter size 5Fr [5,6,7].

Anatomically, the nasal pyriform aperture is the most anterior and narrowest opening of the bony nasal airway. It is pear-like in shape opening that bounded superiorly by nasal bone, inferiorly by the palatine process of the maxilla, and laterally by the nasal process of the maxilla. The cause of the pathology is unknown but it appears that because of the overgrowth of the nasal process of the maxilla in the fourth month of fetal development [8]. It may present an isolated condition or in association with other congenital abnormalities such as holoprosencephaly, cleft palate, and single median maxillary central incisor (SMMCI) with the associated central nervous system and endocrine abnormalities [9,10]. Several case studies have indicated that between 14-66 % of children with CNPAS also have SMMCI [7]. SMMCI occurs during the fourth month of fetal development. It appears in the midline of the maxillary dental arch in both primary and secondary dentition. SMMCI is linked to chromosomes 7 and 8 deletions; these are the chromosomes that carry the holoprosencephaly genes, hence SMMCI is seen as a marker or microform of holoprosencephaly. As a result, the presence of SMMCI in conjunction with CNPAS necessitates the use of magnetic resonance imaging (MRI) of the brain to rule out the presence of holoprosencephaly sequence and pituitary abnormalities [11].

Presentation of CNPAS depends on the severity of the obstruction. If the nasal obstruction is mild, the child usually presents with noisy breathing but is still able to maintain an adequate airway and can feed without difficulty. As for severe obstruction, the child will present early with respiratory distress, episode of cyclical cyanosis that relieved
with crying, episodic apnea that may require oxygen support, or even intubation. As in our case, the child developed respiratory distress after delivery requiring oxygen support. When the pediatric team failed to wean the child off from oxygen for almost one month, diagnosis of CNPAS was considered although it is a very rare cause of neonatal airways obstruction. Hence, because of its very rare occurrence, the diagnosis of CNPAS in this newborn was slightly delayed.

CNPAS diagnosis is usually based on clinical evaluation, nasal endoscopy as well as imaging. Failure to pass suction catheter size 5Fr or slim nasoendoscopy within the first 1cm of the nostrils is considered diagnostic [7,8]. In this child, failure to insert suction tubing of 6Fr beyond 1 cm from the anterior nares prompted our team to evaluate a few possible causes of neonatal nasal obstruction. To confirm the diagnosis of CNPAS, axial CT scan sections in a plane parallel to the hard palate can be performed, demonstrating narrowing of anterior nasal and overgrowth of maxillary process. The normal range of the width of pyriform sinus between the age group of 0-6 months is 8.8-17.2 mm. In term neonate, each pyriform aperture width less than 3 mm, or an entire pyriform aperture width less than 8 mm confirms the diagnosis [11]. CT scan also gives good information to exclude other pathology like mid nasal stenosis, deviated nasal septum, choanal atresia, or congenital mass originating in the nasal cavity itself or mass which extends down from the cranium.

Once CNPAS has been diagnosed, initial medical treatment such as topical nasal decongestants, intranasal steroids, humidification, insertion of oral airways, and lavage feeding can be tried for about 15 days [6,11]. If medical treatment fails, surgical treatment should be considered. It aims for widening the bony inlet. It can be performed via a sublabial approach and reshaping the stenotic area with burs. However, surgery carries several risks such as injury to the nasal mucosa, tooth buds, nasolacrimal duct, septum, and nasal ala. Other than this approach, treatment modalities of CNPAS are nasal dilatation using balloon dilatation or Hegar cervical dilatation as reported by a few authors with good outcomes [12]. Adequate dilatation is achieved when the appropriate size of the nasopharyngeal airway could be placed easily at the end of dilatation or drilling. Nasopharyngeal airway size always follows the ETT size of the child. Nasal stenting is recommended to be used to provide a patent nasal airway, immediate relief of the nasal obstruction while the operative site is healing, and also to prevent recurrence and scar-related stenosis. The length of the nasal stent should be based on the length of the nasal stenosis where concomitant pathology should be taken into account. In isolated CNPAS, a shorter stent should be applied while a longer nasal stent should be used in patients with choanal atresia to prevent obstructive scarring in the posterior nasal area [8]. Duration of placement of the stent is variable, ranging from 5 days to 28 days depending on the initial degree of stenosis and mucosal dissection during surgery. Postoperatively, regular nasal saline and suctioning are very important to prevent any crusting or stent obstruction that might be potentially life-threatening [13]. Regular suction following saline drops must be emphasized to parents if the child is to be discharged with a stent in situ in certain circumstances.

CNPAS is a condition that improves with age and has a very good prognosis. No recurrence has been observed in the literature, even when the first surgery was insufficient [15]. However, the presence of comorbidities such as craniofacial dysmorphisms, neurologic malformations, and airway anomalies have been shown to lower surgical success rates [16]. Several follow-ups should be considered post-operatively especially to look for airway and feeding difficulties, particularly in those requiring hospital readmission. If signs of restenosis do occur, then re-expansion may be required [17].

Conclusion

Although CNPAS is a rare disease, pediatricians and otolaryngologists should consider CNPAS as a differential diagnosis in a child with airways obstruction. Early recognition of the diagnosis and early intervention is important for a good outcome.

References


