Case Report

Congenital nasal pyriform aperture stenosis: A case with accompanying short lingual frenulum

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Abstract

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of nasal obstruction. We presented a case of CNPAS with accompanying short lingual frenulum. Surgical dilatation without osteotomy was used, and the infant had normal growth and development. In these cases, the less invasive surgical methods can be effective.

Key Words: Congenital, infant, nasal obstruction

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INTRODUCTION

Congenital nasal pyriform aperture stenosis (CNPAS) is a potentially life-threatening form of neonatal nasal obstruction. It is a rare condition due to bony overgrowth of the medial nasal process of the maxilla which is most often bilateral. Before 6–8 weeks of age, the infants preferred breathing is nasal, so the common clinical manifestations include dyspnea, feeding difficulties, and apnea, which resemble posterior choanal atresia.[1] This condition presents as an isolated feature or is associated with other malformations including holoprosencephaly, urogenital or cardiac anomalies, and endocrine abnormalities.[2] Because of leading lethal conditions, prompt and early diagnosis is essential for the appropriate management which could be either conservative or surgical.[2,3] In this case, we reported an infant with CNPAS with accompanied short lingual frenulum.

CASE REPORT

A full-term male infant who born at 39 weeks by cesarean section was 3.5 kg in weight, 49 cm in height, and 35 cm in cranial circumference. The Apgar scores were 6, 7, 9 at 1, 5, 10 min, respectively. The newborn presented with multiple episodes of apnea, cyanosis, and feeding difficulties. He was admitted to the Neonatal Intensive Care Unit because of severe respiratory distress, but intubation was not indicated. Ultrasonic scans and clinical examination showed no other dysmorphic feature including midline abnormalities, except short lingual frenulum. There were no cardiovascular or urogenital abnormalities. All of biochemistry tests including hormonal assay were normal. At first, he was suspected to epileptic disorder because of frequent cyanotic episodes, but further evaluation revealed that a five French catheter (outer diameter 1.67 mm) did not pass through the nostrils and using an oral airway lead to improvement of symptoms. Considering these signs and symptoms, choanal atresia was suggested, but a cranial computed tomography (CT) scan revealed stenosis of the pyriform aperture with diameter of 6.3 mm at maximum while the posterior nasal choana had normal caliber [Figure 1].

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The infant initially fed with infant formula by orogastric tube feeding. Conservative treatment with nasal decongestant and nasal conformers was not effective, and surgical management was considered at 15 days of age. In the surgical process, the aperture was dilated with boogies without drill-out of bony margins. A 3.5 mm endotracheal tube was used bilaterally and maintaining as the stent. The tracheal tubes were fixed translabial, and topical saline drop was used to improve extension of the nasal airway. After 2 weeks, nasal tubes were removed and the infant was discharged without further respiratory distress and no complication at 30 days of age. After 8 months of follow-up, the child presented normal development with physical growth within the normal range.

DISCUSSION

CNPAS may lead to asphyxia and respiratory failure, so early diagnosis and treatment is very important.[3] The diagnosis is based on obstructive symptoms, so other causes of nasal obstruction with traumatic, inflammatory, or congenital nature should be differentiated. The most important one is choanal atresia which may be clinically indistinguishable.[4] Clinical symptoms of CNPAS may be delayed until several weeks, but bilateral choanal atresia is manifested in the 1st s of life.[3] The pyriform aperture is at the former part of the nasal fossa. Axial CT scan of the nose and paranasal sinuses with 1.5–3.0 mm contiguous sections and in a parallel plane to the anterior hard palate is used for accurate diagnosing of this anomaly.[1] The narrowest part of the nasal airway is the bony inlet of pyriform aperture and bony overgrowth of medial maxillary nasal processes during the fetal development, may lead to prominent increase in nasal airway resistance. In a term infant, a whole pyriform aperture width <8 mm is indicator of CNPAS.[2]

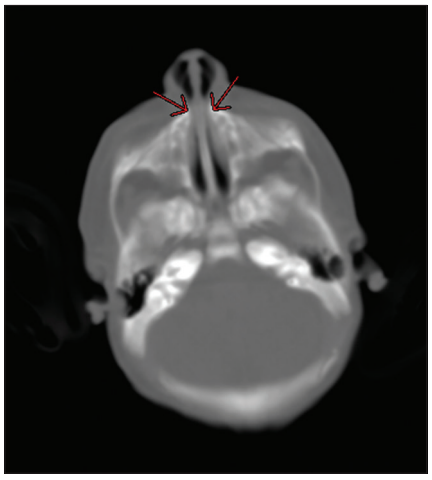

Figure 1: Axial computed tomography scan showing pyriform aperture stenosis

CNPAS may be manifested as an isolated form or in association with other abnormalities including holoprosencephaly or solitary median maxillary central incisor (SMMCI) syndrome which presents in about 60% of infants with CNPAS.[5] SMMCI syndrome is a developmental abnormality, including midline structures of the head which involve the brain (holoprosencephaly), cranial bones and maxilla, and nasal airways (choanal atresia or CNPAS).[6] In our case, there was no other abnormality, except short lingual frenulum which is also a midline structure of the head. Developmentally, the medial nasal processes approach each other during the 6th week of gestation; after that, a single globular process is formed that finally gives rise to the frenulum of the upper lip, nasal tip, prolabium, and primary palate.[7] Short lingual frenulum may be asymptomatic but may have some complications such as difficulties in speech or breastfeeding and dental problems.[8] In the case of short lingual frenulum, a history of feeding, speech, or social difficulties indicates surgical intervention such as frenotomy, frenectomy, or frenuloplasty.[8] In our case, there was no feeding problem until 8 months of growth.

Treatment of CNPAS is initially conservative with nonsurgical approach. Sometimes with growth of the infant, CNPAS would be problematic again after several months, so the surgery should be considered as the last method.[2] Oral airway and silastic stents with accompanying decongestive nasal drops and saline solutions may lead to improvement.[5,9] In moderate to severe cases, infants may not respond to conservative treatment, and this method should not be applied for more than 10–15 days to avoid serious consequences such as mucosal ulcerations or inflammation and metabolic side effects.[5,9] We used surgical treatment after 15 days and no restenosis was observed until 8 months. Surgical treatment typically includes widening the bony inlet with bone resection in anterior nasal aperture, by sublabial approach.[10] Drilling out the nasal process may lead to septal perforation, septal ulcerations, and damage to the tooth buds and granulations. Another way is to dilation of pyriform without drilling because, in neonates, the nasal bone is soft and can be dilated.[2,5] We used boogies for gradual dilatation of nasal pyriform, and the result was acceptable after 8 months. Finally, nasal stents are used for 7–10 days to limit scar-related stenosis and recurrence. Appropriate dilatation is when a 3.5-mm endotracheal tube can be passed.[2,10] Postoperative

care includes treatment of gastroesophageal reflux, using decongestants and nasal humidification for several weeks.[2] Supplemental feeding is needed because feeding problems are common.[9]

CONCLUSION

This case reminds that CNPAS is an important differential diagnosis of neonatal nasal obstruction. Evaluation with CT scan differentiates this anomaly accurately. This condition may be treated conservatively but respiratory distress and necessitate surgical treatment in severe cases. The less invasive surgical methods such as mere nasal dilatation without osteotomy can be effective with minimum complications.

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Conflicts of interest

There are no conflicts of interest.

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