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Case Report

Congenital Nasal Pyriform Aperture Stenosis Associated with a Solitary Median Maxillary Central Incisor Syndrome (A Case Report and Review of the Literature)

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Abstract

Introduction: Congenital nasal pyriform aperture stenosis is a rare and unusual cause of respiratory distress at birth, much rarer when it's associated with a solitary median maxillary central incisor syndrome.

Case Report: A newborn female patient presented a noisy oral respiration with cyanosis during the feeding, the nostrils were narrow but permeable to a thin probe. A facial CT scan was performed showing the presence of a nasal pyriform aperture stenosis with a single central incisor. She was placed on medical treatment with improvement of her clinical conditions without undergoing surgery.

Conclusion: CNPAS is an anterior nasal opening obstruction and should be included in the differential diagnosis of congenital nasal obstruction in neonatal and infancy period.

Some patients may be treated by conservative management, and severely affected ones may undergo surgery.

Keywords: *Congenital Nasal Pyriform Aperture Stenosis; Maxillary Central Incisors; Facial CT Scan*

Introduction

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare etiology of upper airway obstruction, clinically defined by early neonatal respiratory distress with nasofibroscope impossibilities of crossing the pyriform aperture, it can either be isolated or syndromic [1].

This malformation can be explained by maxillary nasal process overgrowth, leading to dyspnea and mortality. Therefore, immediate diagnosis and treatment are essential.

We report a case of a CNPAS with a solitary median maxillary central incisor.

Case Report

A 5-month-old female patient was born by vaginal delivery after 39 weeks of amenorrhea. The parents were not consanguineous. Her mother was poorly followed-up during her pregnancy.

At birth, the Apgar score was 10 in the 5th minute. The newborn was eutrophic with a birth weight of 3400 g. She presented a noisy oral respiration with cyanosis during the feeding. The nostrils were narrow but permeable to a thin probe. The rest of the clinical examination hadn't shown any abnormalities.

The newborn was hospitalized in neonatology department and placed on oxygen and nasal irrigation.

A facial CT scan was performed demonstrating a non-complete stenosis of the 2 nasal cavities, associated to a single central incisor. The choanal opening were normal (Figure 1 and 2).

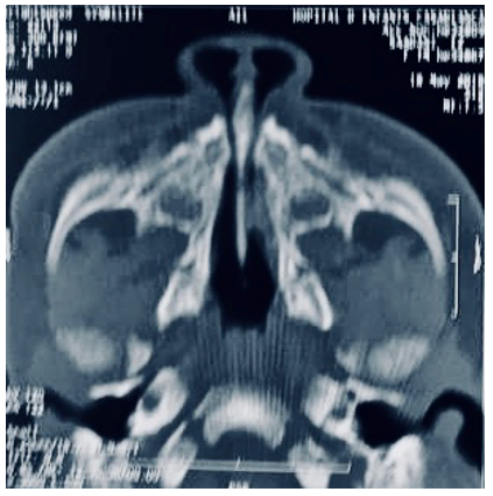

Figure 1: *The CT scan, non-complete stenosis of the nasal pyriform aperture. The choanal opening were normal.*

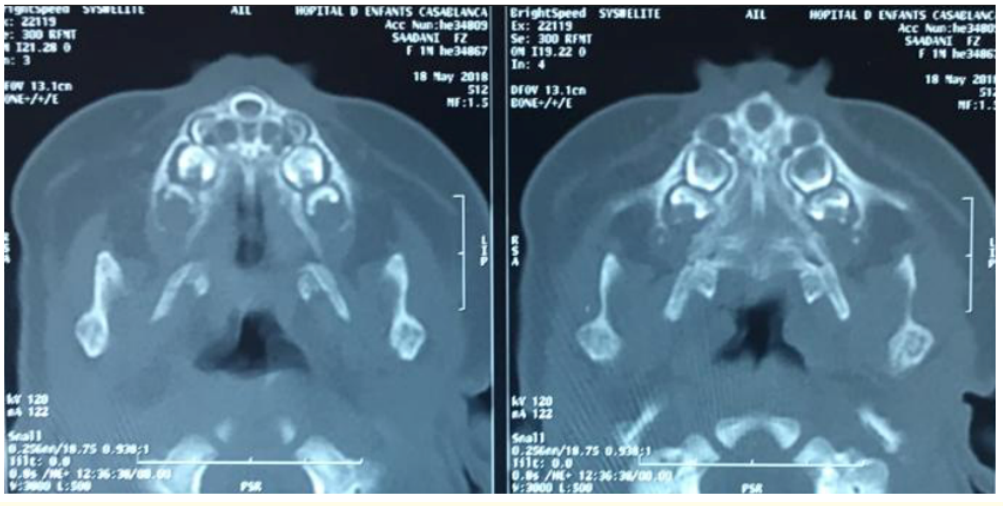

Figure 2: *The CT scan, single central incisor.*

The patient was placed on medical treatment (nasal lavage and corticosteroids) with improvement of her clinical conditions without undergoing surgery.

She kept a mild dyspnea with feeding difficulties until the age of 6 months.

Afterward, the follow-up was normal.

Discussion

CNPAS is a rare cause of congenital nasal obstruction. The association with the maxillary central incisors syndrome is exceptional. The obstructive origin of neonatal respiratory distress is suspected in the presence of worsening cyanosis improving with cries [2].

The three causes of congenital nasal obstruction are: choanal atresia, medial nasal stenosis, and Congenital nasal pyriform aperture stenosis [3].

Several studies demonstrated a relationship between CNPAS and craniofacial abnormalities such as holoprosencephaly, cleft palate, and the early presence of maxillary central incisors similar to our case [4].

CNPAS clinically presents with unspecific symptoms, such as episodes of cyanosis, inspiratory stridor, sternal retraction, thoracic asymmetry, hypoxemia, and acidosis [4].

The nasal pyriform aperture is bounded by the nasal bone superiorly, the nasal process of maxilla laterally, and the horizontal process inferiorly. CNPAS result from the abnormal overgrowth of the maxillary nasal process into the nasal cavity and leads to narrowing the anterior nasal airway [5].

The diagnosis is suspected in the presence of respiratory distress signs and the facial CT scan confirms the diagnosis. The measurements are made on an axial section passing through the internal nasal meatus [6]. They are standardized according to the age of the child. CT scan excludes differential diagnosis (mainly choanal atresia) and shows associated dental abnormalities, particularly a single median incisor presented in almost 50% of cases [5,6].

This association is a part of the maxillary central incisors syndrome, which includes abnormalities in the midline's development. Its incidence is estimated at 1/50,000 live births.

In the first days of life, the disease management is based on gavage feeding and nasal irrigation, then the medical care depends on the severity of the pathology:

- In mild CNPS, a nonsurgical approach with the use of local decongestants is preferable [8].
- In moderate or severe stenosis, surgical approaches are recommended and involved pyriform aperture enlargement through an endo-oral sub labial approach to reshape the stenotic area. This method is safe and enables good field exposure, prevents damage to the nasolabial soft tissues, and does not cause visible scarring [9].

Conclusion

CNPAS is an anterior nasal opening obstruction. It should be included in the differential diagnosis of congenital nasal obstruction in neonatal and infancy period.

It can either be an isolated disease or associated to other dysmorphology and it can be considered as a microform of holoprosencephaly.

Some patients may be treated by conservative management, and severely affected ones may undergo surgery.

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