Congenital Nasal Piriform Aperture Stenosis: A Case Report

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Abstract

Congenital Nasal Piriform Aperture Stenosis (CNPAS) is a rare and an unusual cause of airway obstruction in newborns. Its diagnosis should be considered in the event of neonatal respiratory distress. The diagnosis is confirmed by Computerized Tomographic (CT) and should be made as early as possible for an optimal management. Associated syndromic abnormalities should be looked for by careful clinical examination, CT and MRI. We report this case in order to describe the radiological aspect and the scannographic contribution to the diagnosis of stenosis of the piriform aperture.

Keywords: Stenosis; Piriform aperture; Nasal cavity; Congenital abnormalities; Imaging

Introduction

Congenital Nasal Piriform Aperture Stenosis (CNPAS) is a very rare cause of neonatal respiratory distress, which can be life-threatening to the newborn. It is consecutive to an enlargement of the nasal processes of the jawbone which makes bony nasal permeability precocious [1]. Identified for the first time in 1952 by Douglas, then radiologically in 1988 by Ey et al. [2], the first clinical description was published by Brown et al. [3], in 1989. It is clinically suspected in front of early neonatal respiratory distress with the impossibility of crossing the piriform orifice with a fiberscope or a probe and is confirmed by Computerized Tomography (CT) revealing stenosis at the bony nasal inlet. This clinical definition of CNPAS seems to suit the majority of teams, but the radiological definition poses more problems with innumerable threshold values proposed by the authors [1,4,5]. This stenosis can be isolated or be part of the spectrum of holoprosencephaly, including the Solitary Median Maxillary Central Incisor Syndrome (SMMCIS) and other midline abnormalities. We report this case of isolated CNPAS in order to illustrate the appearance and value of the craniofacial CT scan in the diagnostic workup of this pathology.

Patient and Observation

We report a case of a female newborn from a full term pregnancy with a vaginal birth. At birth, she presented with neonatal nasal obstruction with rapid onset of respiratory distress. On examination, the nostrils were very narrow, almost impermeable to a thin probe. The remainder of the body exam was normal. On facial CT, bilateral stenosis of the anterior nasal orifices was observed, measuring 1 mm on the right and 1.4 mm on the left, surrounded by an outgrowth of the nasal processes of the jawbone and the choanae were normal in appearance (Figures 1, 2).

Discussion

CNPAS is a rare case of congenital nasal obstruction, most cases being related to choanal atresia. Since newborns and infants breathe exclusively through the nose, mouth breathing appearing between three and six months; this condition can be life-threatening by the rapid onset of severe respiratory distress when the piriform apertures are very narrow. The obstructive origin of neonatal respiratory distress is suspected in the presence of cyanosis which worsens during feedings and improves when crying [6]. Faced with this type of distress, it is important to locate the site of the obstruction, making it possible to differentiate the three main causes of congenital nasal obstruction: choanal atresia, medio-nasal stenosis and congenital stenosis of the piriform aperture [7,8]. The piriform aperture is the anterior or bony orifice and the choana is the posterior one of the nasal fossae. The location of the narrowing is immediately at the entrance to the nasal cavity for CNPAS (1 cm) and more posterior for choanal atresia (about 3 cm).

CNPAS is usually bilateral and symmetrical and is caused by enlargement of the nasal processes of the jawbone as well as their medial displacement. It can be isolated or integrated into the Solitary Median Maxillary Central Incisor Syndrome (SMMCIS) comprising developmental abnormalities of the midline, associating in a variable way: short stature, holoprosencephaly, hypopituitarism, microcephaly, mental retardation, a cardiac malformation, a cleft lip or palate, atresia of the esophagus, renal agenesis, a micro-penis and endocrine abnormalities such as diabetes insipidus, whether or not associated with a anterior pituitary gland insufficiency [7,8].

Few publications have reported clinical and radiological features with anatomical measurements of the piriforms orifice in patients with CNPAS. Belden et al. [5], estimated that
the minimum width of a normal piriform orifice is about 11 mm on a CT scan of a full-
term newborn, which corresponded to the smallest piriform opening width obtained from a
control group of asymptomatic newborns. Although this value is not consensual, all patients
with CNPAS published in the literature have piriform orifice width below this threshold.

Clinically, in case of moderate stenosis, respiratory discomfort appears later, usually oc-
curs during breastfeeding. For severe stenosis, nasal airway obstruction and respiratory dis-
tress set in quickly at birth. Clinical examination of the nose, often using a small fiberscope,
reveals a narrowing of the anterior part of the nasal cavity [9]. The diagnosis is then suspected
in front of narrow nostrils with difficulty in passing a nasogastric tube or a fiberscope at the
entrance to the nasal cavity. A complete physical examination of the body should be per-
formed to look for other frequently associated malformative abnormalities.

The tomodesitometry of the facial mass is the reference radiological imagery in front
of any suspicion of CNPAS, making it possible to confirm the diagnosis. Measurements are
made on an axial section passing through the lower nasal meatus, in a plane parallel to the
Frankfort plane, using sizers across the greater width of the piriform orifice. They are stan-
dardized according to the child’s age: in the neonatal period, a width of less than 11 mm indi-
cates a stenosis [5]. CT also makes the differential diagnosis mainly with choanic atresia and
explores associated dental abnormalities, in particular a solitary median maxillary incisor
present in nearly 60% of cases of CNPAS in the literature [7,9]. The computed tomography
criteria in favor of a CNPAS include [5]: a piriform orifice less than 11 mm in a term new-
born; a triangular shape of the palate; excess nasal process growth of the jawbone; and dental
abnormalities such as a single median maxillary incisor.

The management varies depending on the severity of the symptoms, the dysmorphosis
and essentially how the newborn or infant tolerates their nasal obstruction. Medical treat-
ment includes topical decongestants with or without nasal steroids. When sufficient, medical
therapy should be continued until craniofacial growth allows normal ventilation. Other-
wise, the treatment can be surgical at any age, the narrower the stenosis, the earlier the sur-
gery. Usually, the course is progressively favorable as the bones of the nose grow and the two
bone processes that cause the stenosis are spaced apart [7]. The postoperative anatomical
results are evaluated by facial CT.

Conclusion

CNPAS is a very rare congenital nasal obstruction with a clinical presentation similar
to that of choanal atresia. In the event of any neonatal respiratory distress, CNPAS must be
considered among the causes and prompt diagnosis workup and management are necessary.
The diagnosis is confirmed by the scanner. Associated syndromic abnormalities should be
looked for by careful clinical examination, CT and MRI. Treatment is variable, conservative,
or surgical if the stenosis is severe. This offers a good long-term prognosis in the absence
of severe associated malformations.

Conflicts of Interest

We declare not to have any conflicts.

Author’s Contribution

Germaine NIYITANGA, MD: Conception of the work, drafting the work, Author corre-
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Sakina NAIMI, MD: Acquisition of images, Rereading
Ibtissam ZOUITA, Professor: Critically for important intellectual, Approval
Dounia BASRAOUI, Professor: Revising, Approval
Hicham JALAL, Professor: Revising, Approval

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