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Congenital stenosis of the piriform aperture: Two new cases

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Abstract

Introduction: Congenital piriform aperture stenosis is a rare malformation of the nasal passages that is classified as a craniofacial anomaly; it may occur in isolation or as part of a genetic syndrome. Imaging, primarily computed tomography (CT), is essential for confirming the diagnosis. Management must be multidisciplinary.

Material and methods:

Clinical Case 1: Male newborn, admitted at 11 days of age, for management of bilateral choanal atresia. A facial CT scan revealed: stenosis of the piriform aperture on the left, a narrowed appearance of the piriform aperture on the right with mixed choanal atresia on the right and bony choanal atresia on the left.

Clinical Case 2: A full-term female newborn admitted at 10 days of life for respiratory distress. A facial CT scan revealed: stenosis of the piriform aperture with bilateral stenosis of the anterior portions of the nasal fossae, predominantly on the right side.

Both patients underwent surgery involving enlargement of the piriform aperture, with satisfactory intraoperative endoscopic confirmation bilaterally. The postoperative course was uneventful.

Conclusion: Congenital piriform aperture stenosis remains a rare condition, but its severity necessitates early diagnosis and multidisciplinary management. To ensure long-term outcomes and improve quality of life, it is essential to standardize postoperative follow-up and gain a better understanding of the genetic and pathophysiological basis of the disease.

Keywords: Congenital piriform aperture stenosis; Craniofacial anomaly; Neonatal nasal obstruction; choanal atresia; Genetic syndrome; Facial bone structure

1. Introduction

Piriform aperture stenosis is one of a group of craniofacial anomalies characterized by a wide range of phenotypic variations, often associated with other malformations such as choanal atresia or genetic syndromes [1,2]. Congenital piriform aperture stenosis is a bony narrowing of the anterior opening of the nose, causing severe nasal obstruction from birth [3,4].

Respiratory distress is the primary symptom because newborns and infants are exclusively nasal breathers [4].

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The diagnosis is based on clinical findings and imaging (CT scan) to distinguish it from other causes of nasal obstruction, such as choanal atresia.

Treatment involves surgery through endoscopic correction.

Postoperative follow-up is a crucial step that relies primarily on clinical findings and endoscopic examination of the nose, and sometimes imaging, with restenosis being the main risk.

Improved management depends on the creation of a national registry for screening for these malformations [5].

2. Material and methods

2.1. Clinical Case 1

Male newborn, delivered vaginally, from an unmonitored pregnancy. Apgar score not specified; mother is 39 years old, G7P5 (P1 and P3 died at 6 and 7 months of age, respectively, from undetermined causes). The mother has no significant medical history and there is no history of consanguinity.

Admitted at 11 days of life, referred from Mohammed V Hospital in Tangier for management of bilateral choanal atresia.

Weight 3300 g, height 50 cm, head circumference 34 cm, pink, toned, and responsive with a Silverman score of 1-2/10 (Guedel airway in place), no facial dysmorphia.

Cardiac auscultation was normal with no difference in blood pressure between the four limbs, and transthoracic echocardiography was normal.

The malformation workup was negative.

A CT scan of the facial skeleton revealed: stenosis of the left piriform aperture, a narrowed appearance of the right piriform aperture with mixed choanal atresia on the right and bony atresia on the left, and a bifid appearance of the vomer bilaterally (Figure 1).

Endoscopic examination was consistent with a malformation of the piriform bone.

The surgical procedure involved an approach through the vestibular region using a sublabial route, with drilling and enlargement of the piriform foramen, resulting in satisfactory endoscopic visualization bilaterally.

Two No. 3 catheters were placed into the cavum and removed 15 days postoperatively.

The postoperative course was favorable, and the follow-up period is 4 years and 3 months with no recurrence.

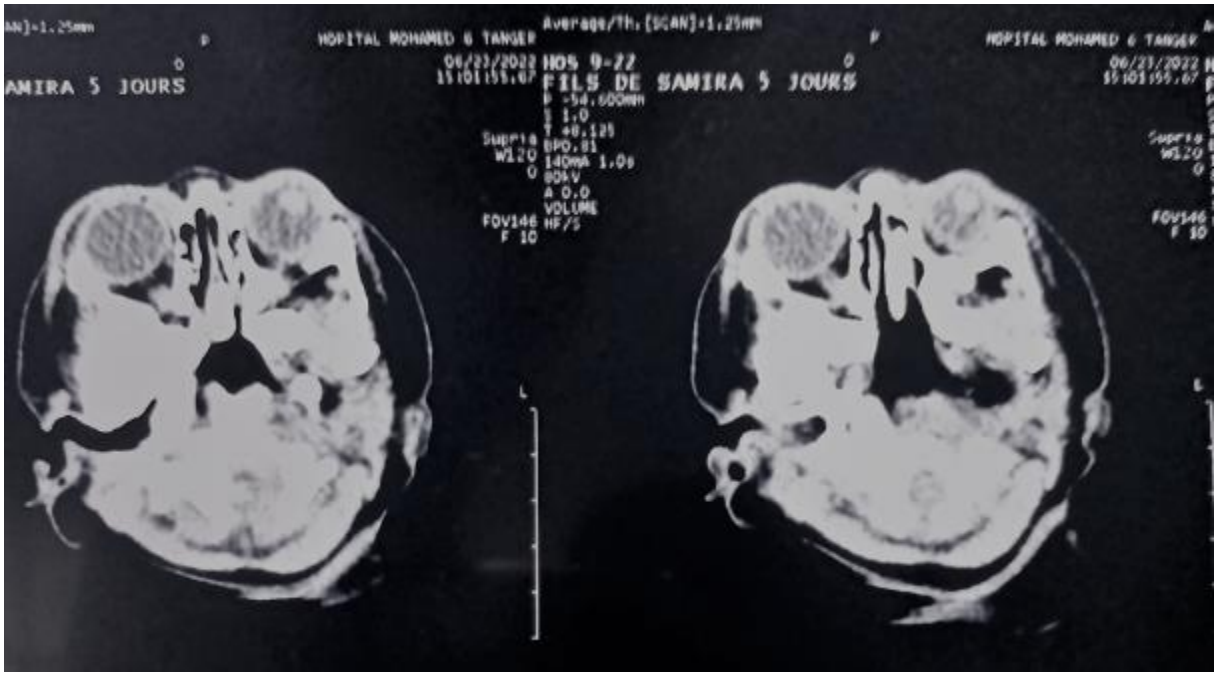

Figure 1 stenosis of the left piriform aperture, a narrowed appearance of the right piriform aperture

2.2. Clinical Case 2

Full-term female newborn, admitted at 10 days of life; scheduled cesarean section on a scarred uterus; Apgar score 10/10; birth weight 3500 g; no history of infection; born to a 32-year-old mother, G2P2, with no significant medical history.

Admitted immediately after birth for respiratory distress due to bilateral choanal atresia at a private clinic.

Examination on admission: respiratory distress rated 1/10, consisting of mild subcostal retraction, with bilateral choanal obstruction; SaO_2 72% on room air, 98% on oxygen mask, and placement of a Guedel airway.

Hemodynamically and neurologically stable. Malformation screening was negative.

A CT scan of the facial skeleton revealed: a 4.5 mm stenosis of the piriform aperture with bilateral stenosis of the anterior portions of the nasal fossae, predominantly on the right, extending 9 mm on the left and 23 mm on the right. The vomer measures less than 3 mm

(Figure 2).

The surgical procedure involved an approach through the vestibular region using a sublabial route, with drilling and enlargement of the piriform opening, and satisfactory endoscopic visualization on both sides.

Two No. 3 probes were placed in the cavity and removed 15 days postoperatively.

The follow-up period is 47 months with no recurrence.

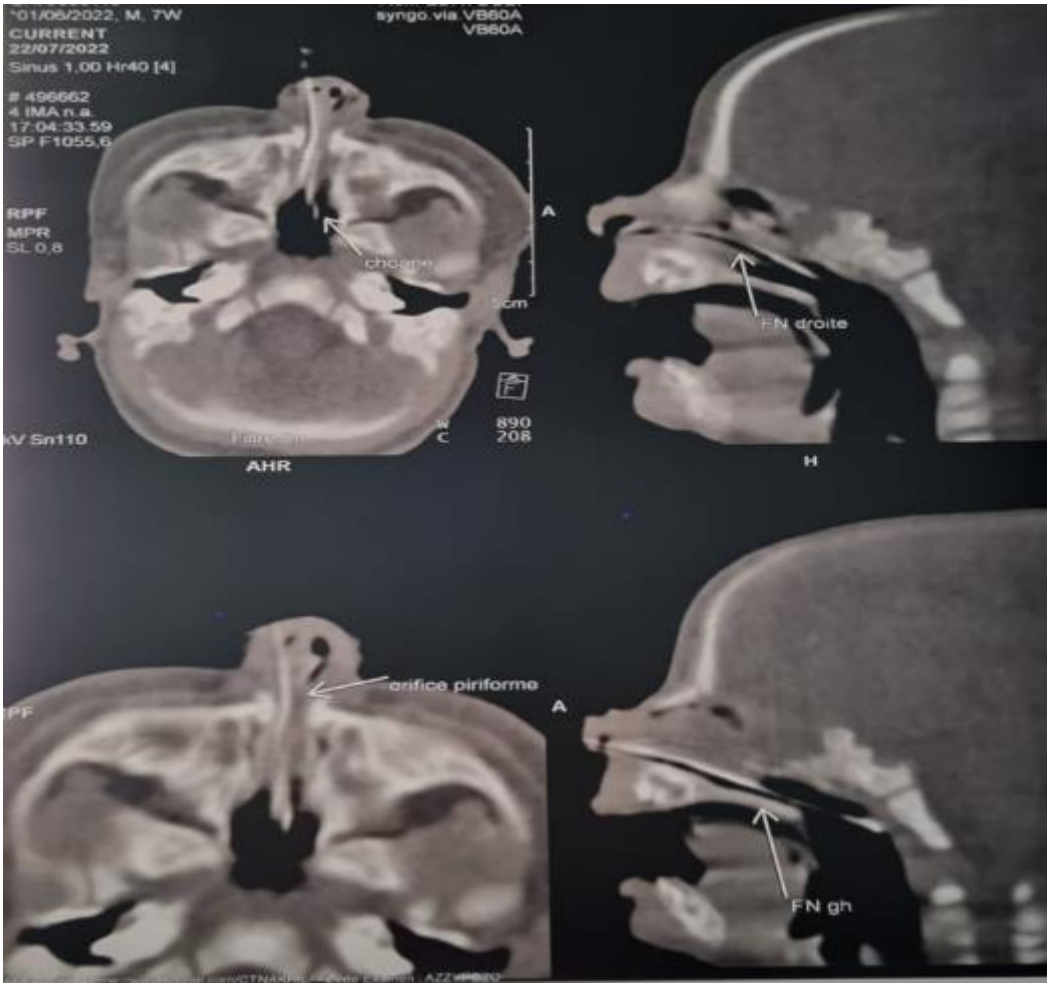

Figure 2 stenosis of the piriform aperture with bilateral stenosis of the anterior portions of the nasal cavities

3. Results and discussion

Craniofacial anomalies, including piriform aperture stenosis, present a wide range of phenotypes, which are associated either with other malformations such as choanal atresia or with genetic syndromes such as Kabuki syndrome and mediator kinase complex syndrome (MED13L) [6].

As well as other syndromes such as Loeys-Dietz syndrome, Treacher Collins syndrome, CHARGE syndrome, and 22q11.2 deletion syndrome, these conditions share common features such as nasal abnormalities or congenital stenosis of the piriform aperture [2,7] .

Neither of our two patients presented with a specific syndrome, and the evaluation for congenital malformations was negative, with a geneticist's opinion that was inconclusive regarding dysmorphism.

The diagnosis is primarily based on clinical findings: respiratory distress, nasal obstruction, difficulty feeding with cyanosis, noisy breathing, and the presence of facial dysmorphism may be indicative. Imaging (CT, MRI) can confirm the stenosis of the piriform aperture (a reduction in the diameter of the piriform aperture to less than 11 mm in the neonatal period) and associated lesions while ruling out other causes of nasal obstruction such as choanal atresia, nasal masses, or the presence of a supernumerary nostril [4,8,9]. Some studies note variability in presentation based on sex, genetic type, and severity, with a female predominance in some cases [2,7].

The initial evaluation should be multidisciplinary, involving otolaryngology, genetics, pediatrics, and sometimes maxillofacial surgery [3,6].

The primary goal of care is to ensure that the newborn's airways remain open [4,10]. Initial management may include symptomatic measures such as nasal decongestion and oxygen therapy while awaiting surgical treatment.

Surgical treatment involves endoscopic correction, with stents placed as needed to prevent restenosis; the emergence of new techniques (such as mucosal flaps and stenting) reduces the long-term risk of restenosis [4,7,11].

Postoperative follow-up is essential for detecting complications such as restenosis or infections[11]. This monitoring is clinical and endoscopic, sometimes involving the use of imaging to assess the condition of the airways and the quality of healing [7,11].

Restenosis can occur in the weeks or months following the procedure[7]. This risk is closely linked to the surgical technique; the placement of nasal stents or the use of mucosal flaps significantly reduces this risk compared to conventional techniques [4,7]. However, even stents can cause local infections or the formation of synechiae [4].

In both cases, the follow-up period was 51 and 47 months, respectively, with no restenosis.

The long-term prognosis will involve facial growth, breathing, and quality of life, with a risk of late-onset recurrence of the obstruction [11]. Close monitoring and careful follow-up help prevent these complications.

4. Conclusion

Congenital piriform aperture stenosis, although rare, is part of a broad spectrum of craniofacial anomalies, often occurring within the context of a syndrome. Diagnosis and treatment have benefited from advances in imaging and surgical techniques, which have significantly reduced recurrence rates and long-term complications.

Care is multidisciplinary, and improving it would require, on the one hand, prenatal screening which remains limited and dependent on more sensitive imaging protocols, and, on the other hand, the creation of national registries.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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