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CASE STUDY

Congenital nasal pyriform aperture stenosis and solitary maxillary central incisor: Case report

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Abstract
Congenital nasal pyriform aperture stenosis has recently been described in the literature. It is caused by an upward overgrowth of the maxillary bone apophysis and may cause breathing and feeding problems from the neonatal period on. We present the case of a newborn girl diagnosed with this pathology associated with a solitary maxillary central incisor. Observation with conservative measures was the attitude chosen. One year after diagnosis the patient shows adequate height and weight development.

PALABRAS CLAVE
Estenosis orificio piriforme;
Cavidad nasal;
Malformaciones;
Embriología;
Cirugía

Estenosis congénita del orificio piriforme y megaincisivo central único

Resumen
La estenosis congénita del orificio piriforme es una anomalía descrita recientemente en la literatura, provocada por un crecimiento excesivo de la apófisis ascendente del hueso maxilar, pudiendo ocasionar problemas respiratorios y alimenticios desde el periodo neonatal. Presentamos el caso de una recién nacida diagnosticada de esta patología asociada a un megaincisivo central único. La actitud elegida fue la observación, junto con la aplicación de medidas conservadoras. Un año después del diagnóstico, la paciente tiene un adecuado desarrollo ponderoestatural.

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Introduction

Congenital nasal pyriform aperture stenosis (CNPAS) was described in 1988 by Ey[1] and defined, as such, by Brown[2] as a rare cause of neonatal nasal obstruction. It is a rare embryonic developmental anomaly and presents as a mild form of holoprosencephaly resulting from abnormal forebrain development in the midline facial structures.[3] It originates secondary to bilateral overgrowth of the maxillary nasal process resulting in a reduction in the calibre of the nasal pyriform aperture and the subsequent appearance of signs and symptoms of obstruction. It may be associated with a mega-incisor or solitary median maxillary central incisor[2-4]; associated endocrine changes have also been described, such as growth hormone deficit,[4] thyroid dysgenesis,[3] hypothyroidism, hypoglycaemic episodes, and absence of the anterior pituitary with panhypopituitarism.[5,6] The karyotype of patients with CNPAS or mega-incisor may be abnormal, showing alterations in chromosomes 18 (18p or r18) and 13 (13q).[7] Depending on the degree of stenosis, the clinical picture ranges from signs of mild nasal respiratory insufficiency to severe cases constituting a true life-threatening emergency and requiring orotracheal intubation. Treatment also varies from using conservative measures in a watch-and-wait approach to surgical intervention.

Case study

Patient was a female term neonate (Caesarean) weighing 3,630 g and measuring 49 cm admitted to the Neonatology Unit for generalised cyanosis, recurrent desaturation episodes, and signs of respiratory distress. On ENT examination, craniofacial, oral cavity, and oropharyngeal morphology were normal. The neck was completely normal to palpation. On anterior rhinoscopy, both nasal vestibules were noted to be reduced in diameter. Passing a nasal probe was impossible in the left nasal fossa and difficult in the right fossa, with the right choana apparently patent. Given the suspicion of a septopyramidal malformation, it was decided to perform a craniofacial CT scan, which yielded the definitive diagnosis (Figure 1 and Figure 2). No changes were found on assessment of the hypothalamic-pituitary axis. Given that oxygen saturation levels stabilised with free-field oxygen administration, the approach was to watch and wait, with monitoring over a period of four weeks in the Neonatology Unit; strict nasal fossa hygiene through administration of topical decongestant drops; aspiration of secretions; and room humidification; together with forced feeding via an orogastric tube. Subsequent care on the regular hospital floor was satisfactory, with an isolated desaturation episode related to accumulated secretions in the nasal fossa. Patient was discharged at six weeks from birth with outpatient follow-up—the last at one year of age, with no height-weight issues noted. On anterior rhinoscopy, pyriform aperture air space was still reduced but, in light of the patient's state of health, sufficient for her to continue developing normally.

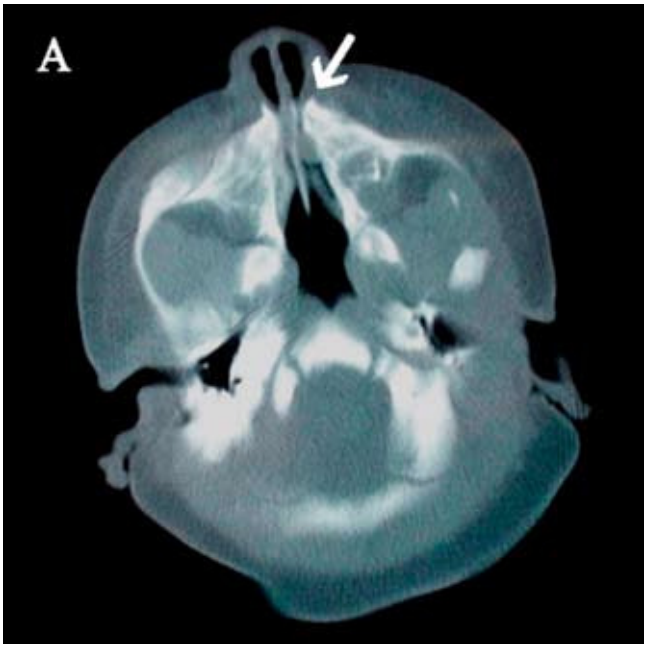

Figure 1 Reduction in size of the nasal pyriform aperture, showing a maximum diameter of 6 mm (transverse measurement of its entirety); patent choanae, where the airway is of normal calibre.

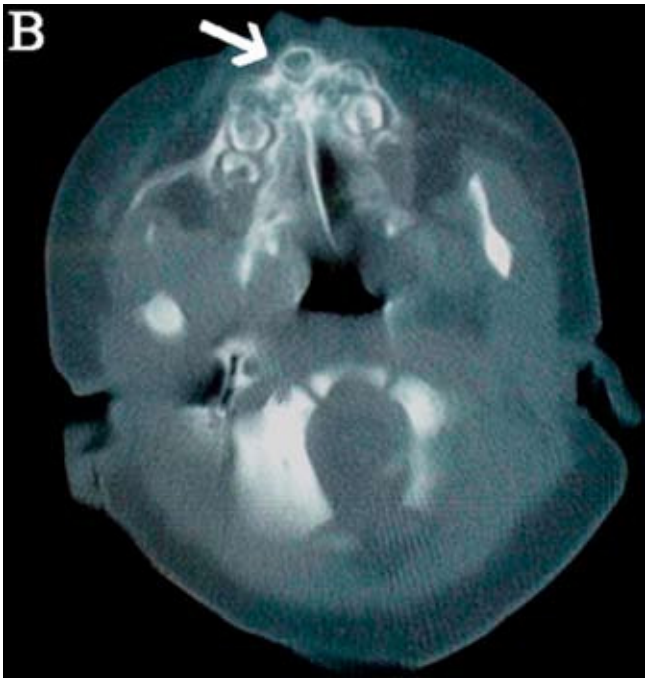

Figure 2 As an associated anomaly, one central upper incisor is seen instead of two.

Discussion

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of nasal obstruction in the neonatal period. Its incidence is not reflected in the literature reviewed except insofar as it occurs with mega-incisor, estimated at

1/50,000 live births, with choanal atresia at 1/5,000, and with holoprosencephaly at 1/16,000 live births or 1/250 for spontaneous abortions.[8] The exact pathogenesis of this malformation is still unknown, but it is associated with an array of defects, the two most common being the single central mega-incisor and pituitary agenesis or insufficiency of the central endocrine gland.[9] The first of these is harmless to the patient's subsequent development–if it does not occur with the second, which may be life-threatening if not diagnosed early on. This is a pathology of major importance because, in and of itself and with some of its associated conditions, it can endanger the patient's life. In the case presented, the patient had recurrent episodes of dyspnoea with cyanosis, especially in connection with accumulation of secretions in the nasal fossa, and an episode of upper airway infection. Hormone assays were within normal limits. Given that neonates are obligate nasal breathers, any type of severe nasal obstruction can cause significant impairment of ventilatory function; early diagnosis may prevent asphyxia.[2,10,11] The pyriform aperture represents the narrowest portion of the nasal cavity; therefore, slight variations in its calibre will result in a major increase in nasal airway resistance. Symptoms may appear at any time from the first hours of life until months after birth in connection with an infectious process of the upper airways; symptoms will also vary depending on the degree of stenosis–with the possibility of a life-threatening emergency developing. Signs and symptoms of upper airway obstruction appear: apnoeic episodes, cyclic cyanosis (worse with feeding and better with crying),[10] difficulty breathing during ingestion, nasal flaring together with supraclavicular or intercostal retraction; these may be associated with varied dysmorphic features.

Diagnosis was based on the symptoms, the physical examination, and especially the imaging tests. We believe that a CT scan should be done on all patients in whom this pathology is suspected. By consensus, complete stenosis of the pyriform aperture exists when the maximum transverse diameter is 3 mm or the total diameter is <8 mm. The differential diagnosis includes other forms of neonatal nasal obstruction, whether acquired pathologies, such as mucosal oedema secondary to rhinitis[12] (the most common) or hypertrophy of the concha, or congenital pathologies such as choanal atresia (the most common type of congenital nasal obstruction), stenosis of the nasal cavity associated with craniofacial anomalies, midnasal stenosis, or lacrimal sac mucocele. If endocrine dysfunction is suspected on the basis of symptoms and laboratory findings, cranial magnetic resonance imaging will be required.[5] In choosing one or the other therapeutic option–conservative management or surgical intervention–we take into consideration the intensity of the symptoms and their impact on the patient's general condition. Thus, if symptoms are not severe, we incline toward conservative management with a watch-and-wait approach, recommending meticulous cleansing of the nasal fossae (aspiration of secretions, instillation of normal saline nasal drops, and room humidification); application of topical nasal decongestants[2,10,11]; McGovern nipple or insertion of a Guedel cannula to maintain an oral airway; and forced feeding (gavage) via orogastric tube. We take a somewhat sceptical view of using a home apnoea monitor and of training the parents in cardiopulmonary resuscitation.[12] If there are ongoing desaturation episodes, cyanosis, severe or persistent apnea with pulmonary hypertension, and failure to thrive,[13] then it becomes necessary to perform orotracheal intubation and transnasal or sublabial surgical intervention (the sublabial approach being easier because it provides a larger field)–at all times bearing in mind preservation of the nasolacrimal ducts and the tooth buds as well as the nasal mucosa–with reaming of the excess bone under microscopic visualisation and posterior endonasal packing with stents for the first week under antibiotic coverage.[2,14,15] In our opinion, surgery should be performed only in those cases of severe stenosis where the conservative measures described above would not be sufficient. From a review of the literature, it appears that, based on post-operative check-ups over the course of one year, the surgery has no harmful side effects on nasal or facial development. In our patient's case, given her lack of symptomatology using the hygienic measures recommended, we believe that follow-up imaging studies are not necessary–despite the persistence of a certain degree of pyriform aperture stenosis on anterior rhinoscopy.

Conclusions

Congenital nasal pyriform aperture stenosis (CNPAS) has been presented as a rare but important cause of neonatal nasal obstruction. It originates with developmental anomalies of the midline craniofacial structures of unknown aetiology; therefore, there may also be a mega-incisor or solitary median maxillary central incisor (occasioning only an aesthetic problem), or there may be endocrine changes, which makes it crucial that the hypothalamic-pituitary axis be assessed. Due to overgrowth of the maxillary nasal process bilaterally, the nasal pyriform aperture is reduced in calibre. The subsequent appearance of signs and symptoms of obstructed breathing, with intensity directly proportional to the degree of stenosis, ranges from signs of mild nasal respiratory insufficiency to severe cases of obstruction constituting a life-threatening emergency. Diagnosis may be based on symptoms and examination of the upper airway, but a CT scan provides the definitive information and supports the decision to intervene surgically, if necessary. Depending on the severity of symptoms, treatment will vary from a watch-and-wait approach, with implementation of conservative measures, to surgical intervention as a last resort.

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