Association between congenital nasolacrimal duct cyst and bilateral choanal atresia

José Faihles Lubicna Neto1, Gabriel Kuhl2, Mariana Magnus Smith3, Person Antunes de Souza4, Leonardo Radunz Vieira5

Keywords: nasal obstruction, choanal atresia, nasolacrimal duct.

INTRODUCTION

Choanal atresia (CA) is defined as a failure in the development of communication between the nasal cavity and the rhino pharynx, causing complete obstruction of the nasal air flow flow\(^2\). A congenital cyst of the nasolacrimal duct (CCnLd) happens through an obstruction of the Hasner valve, located in the nasal cavity and the rhino pharynx, causing complete obstruction of the nasal air flow flow\(^2\). The association of both anomalies seems to be extremely rare, and there is only one case reported in the literature\(^4\).

In the present study there are two cases described of the association between CCnLd and CA, although being a rare finding, the association found in the cases draws our attention to the need to consider other congenital nasal anomalies in individuals with CA.

CASE REPORT

CASE 1

Female neonate, at 37 weeks of gestation, presented with progressive respiratory failure immediately after birth. The maneuver of pushing a probe through the nasal cavities was not efficient, leading to the suspicion of complete CA. Since there was a progressive worsening of the patient’s respiratory status, we proceeded with orotracheal intubation to stabilize the patient. Facing the initial suspicion of CA, we ordered a skull and face CT scan, which showed bilateral CA, and also a mass in the left-side inferior meatus area. A Facial CT scan confirmed bilateral CA, with a meaningful cystic image in the left nasal cavity. A Facial CT scan confirmed bilateral CA, with a meaningful cystic image in the left nasal cavity. The patient evolved well and was successfully extubated after recovery from anesthesia. The patient was discharged under normal ventilation. After 1 year the patient remained well, without recurrences of the choanal stenosis or that of the cyst.

CASE 2

Female neonate with respiratory dysfunction immediately after birth, in whom it was also difficult to progress with the aspiration tube in both nasal cavities. She was then intubated and her condition stabilized. Upon NFE done in the ICU, we noticed a bilateral CA and a cystic image in the left nasal cavity. A Facial CT scan confirmed bilateral CA, with a meaningful posterior thickening of the vomer. The patient was also seen by the genetics team, who ruled out other malformations. Two days later the CA was corrected by endoscopic surgery and suffered cyst marsupialization.

DISCUSSION

Choanal atresia is more common among females\(^1,2\), which we also found in our sample. They can be uni or bilateral, and 60-70% are unilateral\(^3,4\). Both described cases were bilateral, as it happened to most of the patients who required early intervention. The incidence is 1 for every 5-7 thousand live births. The diagnosis is ideally established in an urgent basis immediately after birth through NFE, which is not always available. That is why neonatologists usually are the first to raise the hypothesis of CA by noticing the failure in pushing the nasal aspiration tube farther through the nasal cavity into the pharynx.

Paranasal sinus CT scan and NFE are the gold standard tests\(^5\), since they set the diagnosis and enable the examiner to identify the type of atresia. Historically, CA were described as bony in 90% of the cases and membranous in 10%. However, recent studies suggest that the mixed form is the most common (70%) present in the two cases described here. Such differentiation is important when one selects the surgical treatment.

There are basically three surgical techniques for the treatment of CA: transnasal puncture (TN), transpalatine correction (TP) and the endonasal endoscopic technique (EE). Today, the EE is preferred for it does not have high recurrence rates such as TN, not so many complications as the TP\(^5\).

Associated congenital anomalies can be found in approximately 50% of the cases, and the CHARGE syndrome (coloboma, heart disease, mental retardation, genital and ear anomalies) is the most frequently described condition. Nasal malformations are not frequently seen with CA. In both cases here reported the patients did not have systemic anomalies; notwithstanding, they both had CCnLd.

REFERENCES


1 MD - Federal University of Rio Grande do Sul - UFRGS (1993); ENT Residency at Hospital de Clinicas de Porto Alegre (1992-1993), MSc (1997) and PhD (2000) in Medicine: Medical Sciences - UFRGS. Fellowship at the Pediatric ENT Division at the Massachusetts Eye & Ear Infirmary, Harvard Medical School, Boston, USA (1997-1998) Adjunct Professor IV Of The Federal University of Health Sciences - Porto Alegre, Professor at the Graduate Program in Medical Sciences - UFRGS, Director of the Pediatric ENT Division at Hospital da Criança Santo Antônio do Complexo Hospitalar Santa Casa de Porto Alegre, Scientific director of the Associação Gaúcha de Otorrinolaringologia; Chairman of the ENT committee of the Pediatric Otorhinolaryngology Society of RS and member of the Management Core of the ENT Department of the Brazil Association of Pediatricians; Board member at the Brazilian Association of Otorhinolaryngology and Neck and Facial Surgery, in the International Relations and Teaching, Residency and Training Departments.
2 MD - University of Colima (1978). MD - Universidade de Caxias do Sul (1977) Medical Resident at Hospital de Clinicas de Porto Alegre (1978); Assistant Professor II - Federal University of Rio Grande do Sul; Other - Founding Partner of the Brazilian Association of Laryngology and Voice and aide at the Society of Otorhinolaryngology of Rio Grande do Sul.
3 MD - Pontifícia Universidade Católica do Rio Grande do Sul (1994); MD - Universidade Federal do Rio Grande do Sul (2000) and Medical Residency at the Hospital de Clinicas de Porto Alegre (2004) (Effective Member of the Brazilian Association of Otorhinolaryngology and Special Gradate Student at the Hospital de Clinicas de Porto Alegre. Experienced in Medicine, with emphasis in Otorhinolaryngology).
4 MD - Universidade Federal de Pelotas (2006), Resident Physician at Santa Casa de Misericórdia of Porto Alegre.
5 MD - Universidade Federal de Ciências da Saúde de Porto Alegre (2009).