Rare case of choanal atresia in a 54-years-old female

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Abstract

Introduction: Choanal atresia is a rare disorder presenting with complete separation between the nasal cavity and the nasopharynx. There are two types of this abnormal blockage of the nasal airway – congenital and acquired.

Material and Methods: This paper presents the case of a 54-years-old woman administered to the clinics for septoplasty. During the preoperative diagnostic process via CT scan and endoscope examination a coincidental finding was diagnosed – unilateral congenital choanal atresia. The medical history of the patient revealed difficult nasal breathing which is absent in the right nasal airway, also frequent rhinosinusitis, moreover orthodontic problems with her dentition and malocclusion; regarding her post-birth period she was on feeding tube and with oxygen mask in a neonatal clinic for several months; no facial traumas reported during the rest of her life.

Results: Septoplasty was managed and endoscopic assisted fenestration in the choanal regional was performed viatransseptal access. Seven days postoperatively before the removal of the septal splintsa CT scan was performed for evaluation of the surgical treatment and three months later – endoscopy which displayed well-formed without postoperative stenosis neochoana.

Conclusion: The congenital choanal atresia is a rare abnormality which determines the insufficient clinical experience with this disorder. It is possible that many cases could have been missed or misdiagnosed because of the low incidence of the condition. The conducted literature review reveals that the case report we are presenting is one of the oldest patients diagnosed with unilateral, congenital, bony choanal atresia.

Introduction

Choanal atresia is a rare abnormality presenting in the complete separation of the nasal cavity and the nasopharynx. This disorder was first described by Roederer in 1755 (Niloofar, 2015). There are two types – congenital or acquired. More frequent is the unilateral localization, more often among females (Ahmad, 2010). According to Schwartz (1986), 20% to 50% of the cases referred to a combination with other congenital abnormalities.

Materials and Methods

This paper presents the case of a 54-years-old woman administered to the clinics for septoplasty. During the preoperative diagnostic process via CT scan and endoscope examination a coincidental finding was diagnosed – unilateral congenital choanal atresia. The medical history of the patient revealed difficult nasal breathing which is absent in the right nasal airway, also frequent rhinosinusitis, moreover orthodontic problems with her dentition and malocclusion; regarding her post-birth period she was on feeding tube and with oxygen mask in a neonatal clinic for several months; no facial traumas reported during the rest of her life. Septoplasty was managed and endoscopic assisted fenestration in the choanal regional was performed via transseptal access. Under general anaesthesia a submucosal electrocoagulation on the inferior nasal concha was performed bilaterally, directed at the posterior regions in order to improve the surgical field. Further was proceeded on hydraulic dissection of mucoperiost and mucoperihondrium both sides along the entire length of the nasal septum. After mobilization of the cartilaginous part of the septum the vomer was dissected towards its cranial end as the dissection was managed under endoscopic control. With chisel and hammer the fracture lines were made, then with a bone clamp the part causing blockage of the choana was fully removed. The septoplasty was completed in full capacity. Further the transmucoseptal sutures were placed limiting the possibility of formation of postoperative membranous atresia and septal hematoma. Nasal splints were set for a week and loose nasal tamponade was placed for 24 hours. On the 7th day postoperatively a control CT scan was performed for evaluation of the surgical treatment and three months later – endoscopy. Fig 1. Fig 2.

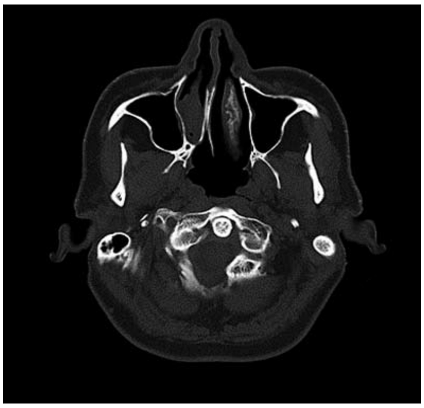

Fig 1. Before surgery (left) Seven days postoperatively before the removal of the splints (right)

Fig 2. Three months after the surgical treatment – an endoscopic examination displaying a well-formed neochoana.

Discussion

Choanal atresia is a discont in uance between the nasal cavity and epipharynx. Classified by structure the condition is divided into three types – bone, membranous and mixed type. Concerning the extend of the affect in 60% ofthe casesit is unilateral andin 40% ofthe cases – bilateral (Riepl, 2014). According to Ahmad (2010) also the more common form is one-sided, with about twofold higher prevalence in females. The same author indicates as more frequent thelocation of unilateral atresia in the right choana. About the incidence of the abnormality different authors publish similar epidemiologica lresults. Xue-zhong (2011) quotes Vanzieleghem (2001), according to which the choanal atresia presents with a frequency of 8.2 per 100,000. According toAhmad (2010) and Niloofar (2015) the disorder can be diagnosed in one of 5000-8000 newborns. According to Riepl (2014) the statistics are similar – inoneof 5000-7000 newborns.

The etiology of choanal atresia is still not fully elucidated at this time but basically it can be divided into congenital and acquired form considering the time and way of formation. Acquired choanalatresia is rare. As a reason for this type rhino-pharyngeal damage is usually reported, for example, after adenoectomy or radiotherapy for treating nasopharyngeal carcinoma, also due to specific granulomatous inflammation of nasopharynx such as tuberculosis or syphilis (Riepl, 2014), but according to Assansen (2009), sometimes the etiology remains unknown.Overtheyears, several theories have been considered about the occurrence of congenital choanal atresia. The most common are: buccopharyngeal membrane persistence, mesoderm persistence with adhesions in the choanal region, persistence of the Hochstetter membrane, cellular migration of spinal cord cells (Hengerer, 1982, Schwartz, 1986, Stankiewicz, 1990, Holland, 2001). Riepl (2014) quoted a Chiastudy (2002), which found a significant association between low thyroxine levels and bilateral choanal atresia in infants.

Unilateral choanal atresia is not a life-threatening condition, except in rare cases of severe airway obstruction. Therefore, diagnosis is usually managed in the later stages of life, not immediately after birth. It causes unilateral nose blockage. Diagnosis is usually performed by nasal endoscopy and / or computer tomography scanning, often as a random finding. This also determines the late planned operative treatment, sometimes in the elderliness (Ahmad, 2010).

In newborns with congenital choanal atresia, the clinical presence is more expressive as the newborn babies breathe almost enxcusively through the nose, especially during feeding. The typical manifestation is presented with intermittent cyanosis, progressing apnea, inability to eat and interrupted crying due to the need of breathing through the mouth. All this requires oral intubation, feeding tube, even urgent tracheotomy may be necessary (Ahmad, 2010; Xue-zhong, 2011; Riepl, 2014).

According to Schwartz (1986) 20% to 50% of the cases referred to a combination with other congenital abnormalities. Between 7-29% of the patients with choanal atresia suffers also from CHARGE syndrome (Harris, 1997) – autosomal dominant disease combining heart defects, choanal atresia, decelerated growth, genital hypoplasia, ear anomalies and deafness (Corrales, 2009). Another syndrome characterized by choanal atresia is the Treacher Collins syndrome, which also combines hypoplasia of the facial bones, deformities of the ear, characteristic eyes and eyelids, incomplete palate.

Conclusion

The congenital choanal atresia is a rare abnormality which determines thein sufficient clinical experience with this disorder. It is possible that many cases could have been missed or misdiagnosed because of the low incidence of the condition. The optional treatment is surgical consisting of various surgical techniques. The conducted literature review reveals that the case report we are presenting is one of the oldest patients diagnosed with unilateral, congenital, bony choanal atresia.

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