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CHOANAL ATRESIA;
DIAGNOSIS AND SIMPLE ENDOSCOPIC REPAIR AT REMOTE PRIMARY CARE CENTERS.

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ABSTRACT... Choanal atresia (CA) is a rare, congenital malformation resulting as a failure in communication between the posterior nasal cavity and the nasopharynx. The clinical course is often asymptomatic in unilateral CA leading to higher rates of misdiagnosis, in comparison to bilateral CA, which manifests itself as a surgical emergency at birth. Most cases present as isolated malformations, but it may also be associated with other congenital anomalies in 20-50% of cases. Currently, the most important diagnostic tool for CA is computerized tomography (CT) and confirmatory diagnosis is usually achieved with the help of nasoendoscopic examination. Although, different surgical approaches have been used in the past, transnasal endoscopic repair is currently preferred over others. Herein, we describe our experience of three cases and share our simple stentless endoscopic technique, to facilitate physicians working in low facility units for a timely diagnosis and prompt treatment.

Key words: Choanal atresia, Endoscopic treatment, surgical approaches, mitomycin C, restenosis

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INTRODUCTION

Choanal atresia (CA) is a rare, congenital malformation defined as a failure in the development of communication between the posterior nasal cavity and the nasopharynx. It seems to be more frequent unilaterally (55%) and affects one in every 7000-8000 newborns, with a Female-to-male ratio of 2:1[1]. The clinical course is often asymptomatic in unilateral CA leading to misdiagnosis, whereas bilateral CA is a surgical emergency at birth and presents with symptoms of intense dyspnea with cyanosis, that is aggravated by feeding and improves on crying[2]. Currently, the most important diagnostic tool for CA is computerized tomography (CT) and confirmatory diagnosis is usually achieved with the help of nasoendoscopic examination[3].Herein, we describe our successful experience of three cases, two of which were surgically treated with simple transnasal endoscopic cauterization without specialized post-operative stenting or use of mitomycin C[4]. This article includes our experience and useful information on choanal atresia, which we believe isn't very rare in this part of the world and often misdiagnosed due to lack of awareness among physicians. We have also shared our improvised and simpler endoscopic technique for surgical correction of the atresia. A discussion about the diagnostic modalities and implications of different treatment options is undertaken.

CASE 1

Our First case was a 5-year-old female, who presented with complain of left-sided mucoid nasal discharge since 2 years. The patient had received multiple treatments including Nasal decongestants and anti-allergic medications. There was no history of trauma, allergies and no associated symptoms related to nose, ear and throat. Based on the chronicity and history, a Rigid nasal endoscopic examination was done which revealed a posterior obstruction, followed by a CT scan which showed left-sided membranous choanal atresia (Image 1).Rest of systemic examination was unremarkable and no associated

congenital anomaly was seen. Family history was positive for asthma, but not significant for any congenital anomalies.

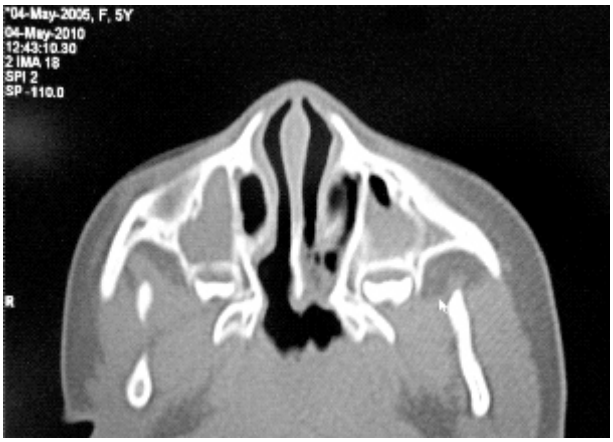

Image 1: Axial CT scan nose and paranasal sinuses of case 1, showing left-sided membranous Choanal atresia.

Case 2

The second case was a 13-year-old female who presented with left sided nasal discharge since 6 weeks. The patient also complaint of frequent nasal blockage and further history didn't reveal any allergic or other associated symptoms. There was no significant family history of congenital anomalies. Systemic examination was unremarkable. Endoscopy revealed unilateral Choanal atresia which was subsequently confirmed by CT scan, which showed left-sided mixed choanal atresia (Image 2).

Both patients underwent transnasal endoscopic cauterization under general anesthesia. Surgical technique included endoscopic cauterization of the membranous portion in the first case. In the second case, mucosa over the atretic bone was first perforated, followed by cauterization. Bony portion was then nibbled with the help of back biting forceps. Subsequently, number 3.5 endotracheal tube was left in place for 5 days in both cases. No packing, stenting or anti fibrosis agent was applied. Antimicrobial therapy was prescribed for one week after surgery and saline irrigations were performed regularly post-operatively. For evaluation of nasal patency we passed a flexible, 4-mm fiber-optic endoscope through the passage at 3rd and 6th month and found complete nasal patency without any complications and restenosis.

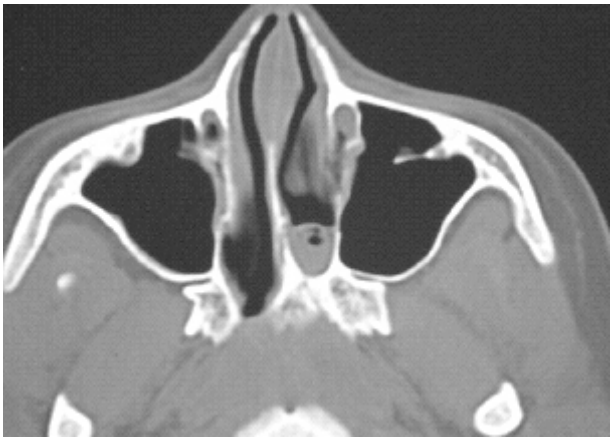

Image 2: Axial CT scan nose and paranasal sinuses of case 2, showing Left-sided mixed bony and membranous Choanal atresia

Case 3

Our third case was an 18-months-old boy who presented with progressive deterioration of respiratory function and right-sided nasal discharge. Past medical history was significant for feeding difficulties and recurrent rhinorrhea. Family history was positive for hypertension but not significant for any congenital anomalies. An Endoscopic examination was done which revealed obstruction and CT scan confirmed right sided mixed Choanal atresia. Although, there were no associated symptoms, but cardiovascular examination was abnormal so an initial chest x-ray was done, which showed Dextrocardia. The patient was advised cardiology consult and transnasal endoscopic repair of atresia but due to concerns of the parents, the patient was referred to a tertiary care hospital for further treatment.

DISCUSSION

Choanal atresia results from the developmental failure of posterior nasal cavity to communicate with the nasopharynx[5]. Anatomically, the atresia area is normally at the palatal-maxillary union and consists of an abnormally enlarged vomer and medialized lateral pterygoid plate[3]. Most cases

present as isolated malformations, but it may also be associated with other congenital malformations in 20-50% of cases, such as CHARGE syndrome (coloboma, heart defect, atresia choanae, retarded growth, genitourinary anomalies and ear anomalies) or other isolated anomalies. For appropriate treatment decisions these anomalies should be identified before surgical correction of atresia[8].

The previous studies in literature have reported the prevalence of Choanal atresia in the range from 32% to 38 %[3,7].Unilateral Choanal atresia (as present in all three of our cases) is often diagnosed later in life due to a long asymptomatic course as compared to bilateral CA which presents as a surgical emergency at birth.

Currently, the most important diagnostic tool for CA is computed tomography (CT), which not only reveals the location, extent, type and nature of the atresia, but also guides for surgical approach. Confirmatory diagnosis is usually achieved with the help of a nasoendoscopic examination.

For Surgical correction of atresia, transnasal endoscopic repair has recently emerged as the most safest and efficacious method[3]. Advantages over other approaches include reduction in operative time, less bleeding, reduced risk of palatal fistula and less disruption of growth centers of the bony plates and the pyramid of the nose[5]. The main complication of endoscopic approach is restenosis of the choanae with a reported incidence from 9% to 36%, depending mainly on the surgical skill of the operating surgeon[2]. In our experience, we find this endoscopic cauterization approach to be highly useful and didn't find any evidence of restenosis in our follow-ups.

Currently, different other techniques along with surgery like post-surgical stenting and use of anti-granulation drugs like mitomycin C are under debate. Post-nasal stenting is often suggested especially in bilateral cases although stent material, fixation and care vary greatly depending on the surgeon. Some believe that stenting prevents restenosis and avoid traditional major surgical approaches, whereas others claim that they are associated with high risk of scars, decubitus, infections and are not cosmetically acceptable. However, information regarding new stents reports them to be highly effective and free of these complications. Mitomycin C on the other hand, acts as an anti-fibroblastic agent and prevents formation of granulation and hence subsequent scarring and restenosis.Its suitability for reducing restenosis in low doses is justified by some studies. However, there is a potential risk of systemic side effects due to overdose and so far there is no consensus regarding exact dose[9]. We therefore didn't employ any of these, owing to lack of knowledge and no protocol regarding dosage and use.

We believe that complications occur regardless of surgical techniques and recurrence depends on other factors including age of the patient, nature of obstruction(bony vs. membranous; unilateral vs. bilateral), antacid treatment for physiological GERD, presence of other congenital anomalies and good post-operative care (involving frequent nasal rinsing and regular follow-ups). The surgeon's learning curve regarding these prognostic factors and understanding of the importance of follow-up care is rising resulting in deceased recurrence of restenosis[2]. Still, more focused protocols regarding the appropriate techniques and dosage of agents like mitomycin C are mandatory for convenience and better understanding.

Although this report has the limitation of a small number of cases, but our results are still comparable to cases reported in literature and may provide useful information to guide treatment decisions among physicians. We have been quite successful in preventing restenosis in our patients with a simpler technique and believe that, at least unilateral choanal atresia can be diagnosed and treated at low facility units.

We also conclude to suspect the unlikely possibility of unilateral choanal atresia in any age of child with long-standing unilateral nasal obstruction in the absence of a foreign body or

adenoidal hyperplasia".

REFERENCES

1. Mantovani, M., et al., **A new dynamic endonasal stent for bilateral congenital choanal atresia.** Acta Otorhinolaryngol Ital, 2009. 29(4): p. 209-12.
2. Teissier, N., et al., **Predictive factors for success after transnasal endoscopic treatment of choanal atresia.** Arch Otolaryngol Head Neck Surg, 2008. 134(1): p. 57-61.
3. Al-Noury, K. and A. Lotfy, **Role of multislice computed tomography and local contrast in the diagnosis and characterization of choanal atresia.** Int J Pediatr. 2011: p. 280763.
4. Holland, B.W. and W.F. McGuirt, Jr., **Surgical management of choanal atresia: improved outcome using mitomycin.** Arch Otolaryngol Head Neck Surg, 2001. 127(11): p. 1375-80.
5. Irfan, M. and S.A. Suzina, **A missed bilateral choanal atresia.** Ann Acad Med Singapore. 39(1): p. 72.
6. Cedin, A.C., et al., **Surgery for congenital choanal atresia.** Cochrane Database Syst Rev. 2: p. CD008993.
7. Al-Ammar, A.Y., **Presentation of choanal atresia in Saudi children.** Saudi Med J, 2006. 27(11): p. 1680-2.
8. Burrow, T.A., et al., **Characterization of congenital anomalies in individuals with choanal atresia.** Arch Otolaryngol Head Neck Surg, 2009. 135(6): p. 543-7.
9. Pardo Romero, G., et al., **[Endoscopic treatment for choanal atresia].** Acta Otorrinolaringol Esp, 2007. 58(1): p. 34-6.