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CASE REPORT

VOMER AGENESIS AS A RARE CAUSE OF HYPERNASALITY

Adam MOHAMAD [1 2], Rohaida IBRAHIM [2], Khairul Azhar MOHD RAJET [2], Irfan MOHAMAD [1], Anura AMAN [2]

[1] Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, 16150 Kota Bharu, Kelantan, Malaysia

[2] Department of Otorhinolaryngology, Hospital Tengku Ampuan Afzan, 25100 Kuantan, Pahang, Malaysia

ABSTRACT
Hypernasality which is rare symptom commonly occur as a consequence of velopharyngeal insufficiency (VPI). VPI usually manifested as nasal air emission and hypernasal resonance during speech. The cause can be divided into congenital, neuromuscular disorder and surgical complication. Congenital cause of VPI includes cleft palate, nasal septum malformation such as vomer agenesis, submucous cleft palate and velar dysplasia, while neuromuscular VPI can be due to cerebral palsy or cerebrovascular accident. Surgical cause of VPI could be due to adenoidectomy and scarring of the velum post palatoplasty in cleft palate repair. We present a 17-year-old man who was diagnosed of congenital left nasolacrimal duct obstruction referred to us for left endoscopic dacrocystorhinostomy in which during nasoendoscopic examination revealed absence of vomer.

KEY WORDS: Hypernasality, Velopharyngeal insufficiency, Vomer, Velum.

Corresponding author: Dr Adam Mohamad, Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, 16150 Kota Bharu, Kelantan, Malaysia. Email: persona522115@gmail.com

INTRODUCTION
Vomer agenesis is one of congenital structural nasal septum defect. It is rarely seen nowadays. Less than 22 cases were reported and mostly it is seen incidentally during nasoendoscopic examination (1). In most of the case, they had no history of maxillofacial trauma, nasal surgery, syphilis, cauterization or drug abuse (2). A few cases also encountered during computed tomograph (CT) done for facial skeleton evaluation (3). Because of the large defect at the posteroinferior part of septum, it indirectly will cause some degree of velopharyngeal insufficiency. This patient manifested as hypernasality which already presence since his childhood.

CASE REPORT
A 17-year-old man who was diagnosed of congenital left nasolacrimal duct obstruction who present with persistent left epiphora since childhood, referred for left endoscopic dacrocystorhinostomy (EDCR). Apart from that, he also had rhinitis symptoms such as nasal blockage, nasal itchiness, and rhinorrhoea for many years in which he had allergy to seafood. He also noted to have hypernasality since childhood as well as intermittent fluid regurgitation, but never been investigated before.

On examination, patient was alert and conscious. His vital signs were stable. There was left eye epiphora noted. Endoscopic nasal examination shows absence of vomer. Other than that, there were congested nasal mucosa with lots of secretion seen as well as presence of grade II nasal polyp coming from left osteomeatal complex (OMC) and grade I nasal polyp seen in right OMC. Flexible nasopharyngolaryngoscopy (FNPLS) showed inadequate closure of the velopharynx.

He was diagnosed of congenital vomer agenesis, velopharyngeal insufficiency (VPI), chronic

rhinosinusitis with bilateral nasal polyposis and left nasolacrimal duct obstruction. He was started with intranasal steroid which is mometasone furoate nasal spray 2 puffs 12 hourly, tablet loratadine 10 mg once daily. Computed tomograph (CT) of paranasal sinus (PNS) was done shows presence of mucosal thickening in both maxillary sinus ethmoid sinuses as well as absence of posteroinferior part of the nasal septum (Figure 2). He is planned for EDCR and functional endoscopic sinus surgery (FESS). However, in term of the VPI, it was decided for conservative management.

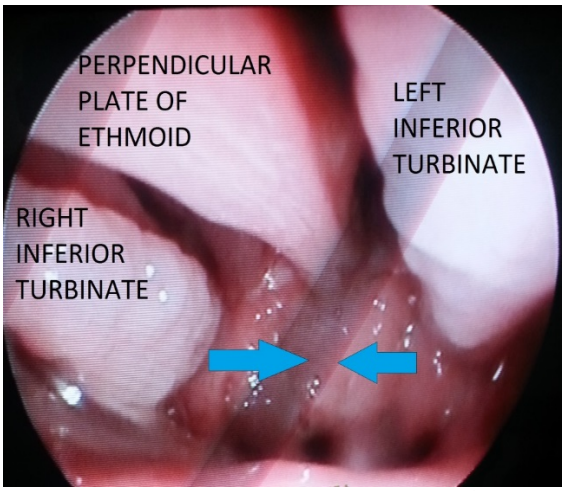

Figure 1: The posterior septum (arrow) showed absence of vomer.

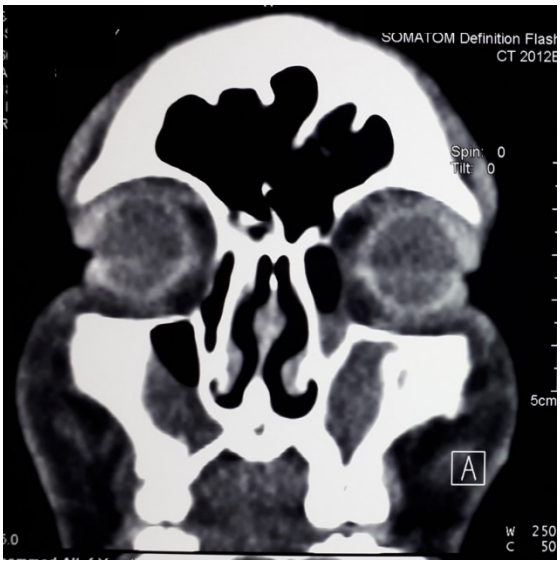

Figure 2a : Coronal view of the CT scan (anterior cut) showing presence of the anterior part of the nasal septum.

DISCUSSION

Hypernasality in general is a resonance disorder caused by VPI(4). VPI is defined as an incomplete closure of velopharynx which consist of tensor veli palatine, levator veli palatine, muscularis uvulae, palatoglossus and palatopharyngeus muscles(5). VPI can be manifested as hypernasality as well as nasal regurgitation which presence and demonstrated in our patient. During the FNPLS, with the help of our speech pathologist, our patient was asked to read few sentences which project the production of consonant sounds. During the procedure, it show minimal inadequate closure of the velum to the posterior pharyngeal wall which consistent with VPI.

The cause of VPI can be divided into congenital, neuromuscular disorder and surgical complication. Congenital cause of VPI includes cleft palate, nasal septum malformation such as vomer agenesis, submucous cleft palate and velar dysplasia, while neuromuscular VPI can be due to cerebral palsy or cerebrovascular accident. Surgical cause of VPI could be due to adenoidectomy and scarring of the velum post palatoplasty in cleft palate repair. In adenoidectomy, VPI can occur due to the fact that most children have velo-adenoidal closure. When the adenoid is removed, it caused deeper nasopharynx and increase in the distance between velum to the nasopharynx to achieve complete closure (6).

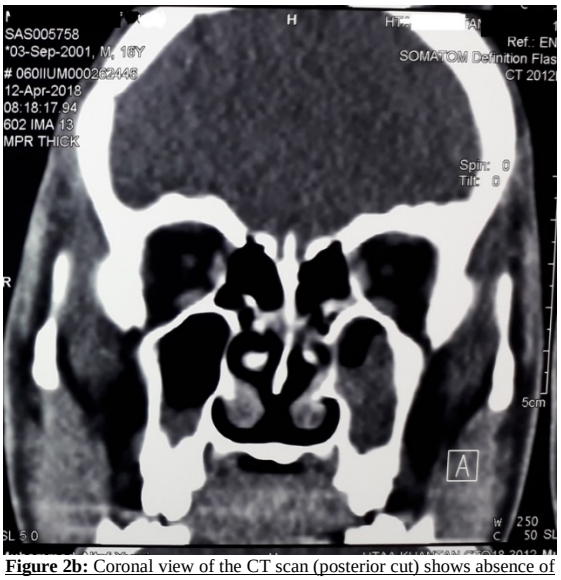

Figure 2b: Coronal view of the CT scan (posterior cut) shows absence of vomer.

Nasal septum defects can be due to many causes such as nasal surgery which causes septal perforation, trauma, infection such as syphilis or tuberculosis, chronic inflammatory diseases such as Wegener's granulomatosis, neoplasm such as natural killer T-cell lymphoma or drug abuse; all of these usually affecting the anteroinferior part of nasal septum(7). In contrast to vomer agenesis, it involves the posteroinferior part of nasal septum.

Defect of the posteroinferior part of nasal septum occur when there is interference of the normal stages of vomer development which will lead to VPI and subsequent hypernasality. Two theories of vomer agenesis were postulated. The first one is "immature ossification center" theory which postulates the presence of an incomplete or immature ossification center while the latter is the "incomplete downward growth" theory, which stated that the posterior extension and downward growth of the primary nasal septum are interrupted(1).

Though in our case there is no active surgical intervention for the vomer agenesis itself, it is important that we can established the diagnosis or the root cause of hypernasality and early referral to the speech therapist given. There are few surgical options for velopharyngeal surgery such as pharyngeal flap, sphincter pharyngoplasty and pharyngeal augmentation(6). However, since there was very minimal inadequate closure of the velopharynx we decided for conservative management for the VPI.

Possible VPI in this case is likely due absence of vomer causes weakness closure of the velopharynx by the soft palate muscles which directly cause VPI.

CONCLUSION

Hypernasality is multifactorial. If such case present, nasoendoscopy and flexible nasopharyngolaryngoscopy should be performed to find the possible cause of it.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript and provided approval for this final revised version.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTERESTS

The authors declare no competing interests.

REFERENCES

[1] Mohri M, Amatsu M. Congenital defects of the vomer. Annals of Otology, Rhinology & Laryngology. 2000;109(5):497-9.

[2] Lee JH. Congenital vomeral bone defect: report of two cases and a review of the literature. Acta Oto-Laryngologica. 2006;126(11):1229-31.

[3] Yan DJ, Lenoir V, Chatelain S et al. Congenital Vomer Agenesis: A Rare and Poorly Understood Condition Revealed by Cone Beam CT. Diagnostics. 2018;8(1):15.

[4] Golabbakhsh M, Abnavi F, Kadkhodaei Elyaderani M et al. Automatic identification of hypernasality in normal and cleft lip and palate patients with acoustic analysis of speech. The Journal of the Acoustical Society of America. 2017;141(2):929-35.

[5] Rudnick EF, Sie KC. Velopharyngeal insufficiency: current concepts in diagnosis and management. Current Opinion in Otolaryngology & Head and Neck Surgery. 2008;16(6):530-5.

[6] Kummer AW, Marshall JL, Wilson MM. Non-cleft causes of velopharyngeal dysfunction: implications for treatment. International Journal of Pediatric Otorhinolaryngology. 2015;79(3):286-95.

[7] Lanier B, Kai G, Marple B et al. Pathophysiology and progression of nasal septal perforation. Annals of Allergy, Asthma & Immunology. 2007;99(6):473-80.