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Case Report

Bifid Uvula—An Enigma

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Submitted: 07-Oct-2022
Revised: 01-Dec-2022
Accepted: 14-Dec-2022
Published: 05-Jul-2023

ABSTRACT

Facial development involves an intricate regulatory mechanism that accounts for numerous craniofacial abnormalities, common being orofacial clefts. Although cleft in the secondary palate accounts for one-third of orofacial clefts stills remains an under-researched domain. Hence, in this work, the authors put forth two non-syndromic, asymptomatic cleft uvulae reported among bimodal male patients of the Indian-Asiatic population who came up for dental screening. Most of the time, isolated/asymptomatic cleft uvula patients will be reluctant to further investigations and treatment. Although bifid uvula looks benign in most patients, it may sometimes be associated with catastrophic complications. To conclude, whenever bifid uvula is an incidental finding, it is the responsibility of the healthcare worker to plan a thorough patient workup as a primary preventive measure to rule out any complications whenever feasible. It can help us overcome many future unforeseen sequelae and emergency management due to bifid uvula.

KEYWORDS: *Bifid, cleft uvula, non-syndromic, uvula*

INTRODUCTION

The uvula is a vital organ in the craniofacial complex concerned with speech, deglutition, and mastication.[1] The literature revealed numerous abnormalities of this small grapes-shaped structure, the

Access this article online

Quick Response Code:

Website: https://journals.lww.com/jpbs

DOI: 10.4103/jpbs.jpbs_464_22

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How to cite this article: Prasad P, Khalil E, Desai VB, Varma SR, Gunasekaran L, Kumar K, et al. Bifid uvula—An enigma. J Pharm Bioall Sci 2023;15:S806-9.

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most common being split uvula. The split uvula, also commonly known as the bifid uvula, is a milder form of cleft palate.[2]

The etiology of cleft palate occurrence is multifactorial. It involves both major and minor genetic variations synergistic with interplay from the environmental domain.[2] Human orofacial cleft incidence remains a Bermuda Triangle among researchers. We cannot get a wholesome picture of its molecular mechanisms despite numerous case-based and genomic studies researched in this area.

Evidence reported that the prevalence of split uvula is 2% in Whites; however, isolated epidemiological data globally on this condition remains under research. It may be because bifid uvula is mainly an incidental finding in asymptomatic patients. Further, symptomatic/asymptomatic bifid uvula clinically presents itself with either or a combination of the following conditions: submucosal cleft; inability to form a seal with pharyngeal wall leading to frequent regurgitation; velopharyngeal insufficiency; nasal intonation; a systemic disease like aneurysm and syndromes like Loyez-Dietz syndrome, Marfan Syndrome.[3-5]

In this report, the authors have illustrated two cases of incidental bifid uvula in the Indian population.

Case Report 1

An eight-year-old male child who came for dental screening at a private dental clinic accompanied by his mother, who was visiting for her dental treatment, was incidentally diagnosed with Type D bifid uvula [Figure 1]. The kid was on-term and typically delivered the baby with no other relevant history. There was no history of consanguineous marriage. The baby was delivered normally after a full-term pregnancy. On further examination, there was the absence of a cleft in the lip or palate.

Case Report 2

A 44-year-old male with no relevant family history reported to the private clinic for dental screening and was incidentally diagnosed with bifid uvula (Type C) [Figure 2]. The dentist did not find any other associated physical signs and symptoms in the orofacial complex. Further, none in the family pedigree were reported to have bifid uvula. The adult male does not remember having any difficulties in speech or swallowing while growing up.

Discussion

Indeed, a complex regulatory mechanism is involved in embryonic facial development. Among the craniofacial abnormalities, orofacial clefts hold a prime position. Evidence suggested cleft involving the secondary palate stills remains under-researched domain.[6,7] Hence we reported the occurrence of two non-syndromic cleft uvula reported among the Indian-Asiatic population. CPO (MIM 119540) includes cleft uvula as one entity under its classification and further declares cleft in the secondary palate accounts for one-third of orofacial clefts reported. Numerous pieces of evidence demonstrated a web of risk factors and etiology of both genetic and environmental domains for CPO.[8]

Although cleft uvula and cleft palate prevalence vary from 1.5% to 10%, isolated cleft uvula epidemiological distribution remains unveiled.[7] Cleft uvula may range from a developmental malformation to symptoms of several chromosomal syndromes like trisomy.

The embryological development of the uvula takes place through the proliferation of cells between the distal palatine shelves resulting in smoothening of the soft

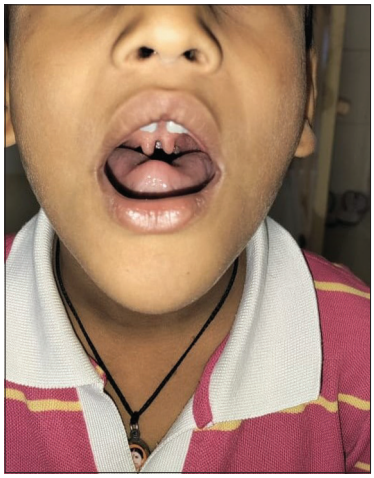

Figure 1: An eight-year-old male child with Type D bifid uvula

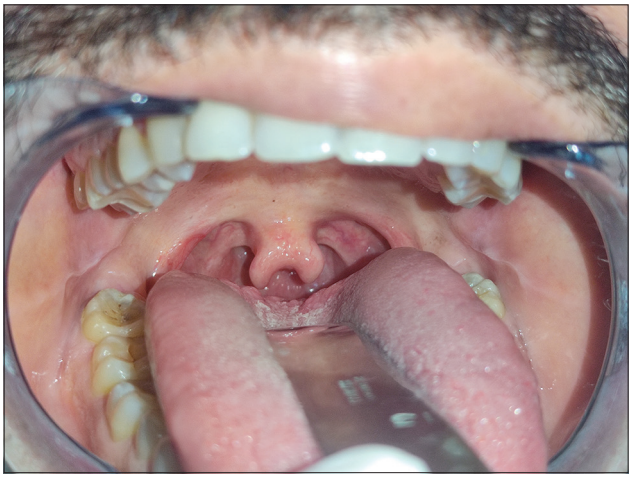

Figure 2: A 44-year-old male with Type C bifid uvula

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palate and its associated structures. The uniqueness of the uvula formation lies in fact; it develops from the mesenchymal merging of two distinct masses, which originate from the posterior part of the palatine shelves, unlike the fusion mechanism of the primary palate that takes place all along the length of the palatine shelves. Failure of the merging process in the soft palate and uvula development can result in complete or partial clefts of the soft palate and uvula. Classification of the types of cleft uvula as discussed by Meskin (1964) based on morphology are Type A: normal uvula, Type B: uvula bifurcated up to one-fourth of its total length, Type C: uvula bifurcated from one-fourth to three-fourths of its size, and Type D: uvula bifurcated from three-fourths to its entire length.[7]

The bifid uvula is a frequently observed anomaly that has served as a clue for clinicians to detect the earliest signs of various anomalies. It has been correlated to a clinical sign of many distinct syndromes manifesting with varied patterns and prevalence worldwide.[4]

Although bifid uvula looks benign in most patients, it may sometimes be associated with anomalies leading to catastrophic complications. It has been reported that bifid uvula increases the odds of numerous conditions like chromosomal schizophrenia and milder forms of mental retardation. Many congenital syndromes like Cornelia de Lange syndrome, Loeys-Dietz syndrome, and Marfan syndrome manifest this condition. Differential diagnosis of Loeys-Dietz syndrome and Marfan syndrome becomes challenging except for the chromosomal deletion of the TGFB2 gene.[9-11]

A classic example depicts "bifid uvula may be a cautioning sign of the syndrome with internal anatomical or functional changes without any physical manifestation," as reported by Samanta. In 2013, a 16-year-old boy who underwent retinal surgery was noted to have a bifid uvula and a milder form of hypertelorism. During retinal surgery, the patient failed to be extubated and had anisocoria. It was an emergency condition when CT revealed a ruptured aneurysm in the brain, following which immediate craniotomy and clipping were performed as a life-saving procedure.[12]

Another case report in 2008 revealed an association between a case of the split uvula and an aortic aneurysm. In their work, the authors reported a positive family history of the aortic disease, which, when further investigated, was diagnosed as Loeys-Dietz syndrome.[13]

In this work, the authors reported that both patients presented asymptomatic bifid uvula and that no first-degree relatives had a congenital anomaly. On the contrary, Khasbage SD 2017 wrote an incidental finding of the bifid uvula in a 58-year-old man. Snowball sampling of this patient revealed that both his sons had the same clinical condition.[9]

As discussed earlier, bifid uvula was found to be more common in males than in females.[7,14] The present work also reported that the incidental finding of bifid uvula was in the male gender.[15]

The bifid uvula is a marker for the submucous cleft palate.[3,7] In the present work, no accompanying submucosal cleft was identified in either of the cases. Our study aligns with Brazilian research on 1206 children; none had the event.[4]

Symptomatic severity of the bifid uvula determines its treatment planning. Very few opt for either the removal or surgical reconstruction of these abnormal tissues. Most of the time, isolated/asymptomatic cleft uvula patients will be reluctant to further investigations and treatment.[9] Similarly, both cases refrained from any diagnostic or treatment protocol in the present scenario.

A clear-cut knowledge of any congenital abnormality can be attributed to our understanding of its distribution, determinants, covariates, and precise phenotyping. A more complex spectrum of phenotyping patterns was observed in the non-syndromic bifid uvula, making every case unique, necessitating its reporting for future analysis.

Conclusion

To conclude, whenever bifid uvula is an incidental finding, it is the responsibility of the healthcare worker to elicit a detailed history and evaluate any anatomical or physiological variation and genetic predisposition. Whenever feasible, it is essential to plan a thorough patient workup as a primary preventive measure to rule out any submucosal cleft, aneurysm, or anesthetist complications. It can help us overcome many unforeseen sequelae and emergency management due to bifid uvula.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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