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The longest uvula - stretched to the epiglottis from normal swallowing reflexes activated by a fibroma on its tip

Abstract

Background: An enlarged uvula is known to result from more than a dozen different toxic, infectious or traumatizing phenomena, including pharyngitis, smoking cocaine or crack, snoring/sleep apnea, and several sexually transmitted infections. When thus affected, the uvula can become 2-3 times wider than normal (from edema and congestion) and as much as 50% longer. This structure is, of course, an integral part of the swallowing mechanism, a process requiring constant coordination of almost 30 muscles controlled by five cranial nerve systems. It works so well, of course, that we almost never think about it, but what would happen if a mass developed on a uvular surface? We present the first case of such a mass, an irritation fibroma, attached to the tip of a uvula. The coordinated swallowing system apparently repeatedly tried to swallow it, for many months, resulting in the longest, thinnest uvula every reported, extending down to the epiglottis.

Case summary: A 29-year-old male was referred to a dentist by his family physician for evaluation of a "thing dangling down his throat." The patient first noticed an enhanced gag reflex nine months earlier, with the gagging became more severe and more frequent over time, especially when eating. In recent weeks he has also noticed that his speech had a "nasal sound." He has had "a snoring problem" since college and was recently diagnosed with sleep apnea and started using a CPAP device. At examination a very thin, elongated uvula was noted and, when asked, the patient was able to cough it up into his mouth. The uvula was somewhat erythematous and had a sausage-shaped irritation fibroma attached to its tip. The uvula was approximately 2.5 cm. long and the 0.6 mm fibroma was seen to rest just above the epiglottis when in its "normal" hanging position. Surgical removal eliminated the patient's snoring and speech issues. Histology confirmed the fibroma diagnosis, and frictional keratosis of its surface was consistent with repeated mild friction/trauma.

Conclusion: We report the first example of a uvula severely "stretched" by normal pharyngeal muscle action attempting to "swallow" an irritation fibroma attached to its tip.

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Keywords: Uvula; Irritation fibroma; Swallowing reflexes; Oropharynx.

Introduction

Swallowing is a hugely complex process, involving almost 30 different symmetrical muscles moving in coordination with each other, controlled by 5 different cranial nerve systems, all working together over and over and over again during a meal [1,2]. This requires continuous and smooth pushing and pulling of food from the mouth into the hypopharynx and then into the esophagus, where peristaltic movement will bring it down into the stomach [3]. This coordinated action is different for liquids than for solids, works even when a person is upside down, and must take place while constantly protecting the airways (both nasopharynx and bronchial tubes). Additionally, while the food is reaching the esophagus at the end of its travels, more food is being placed into the mouth to start the process all over again, requiring synchronization with the next "wave" of activity.

Swallowing solid food starts with the "oral propulsive stage," in which the food is pushed down along the palate and uvula [1]. If the uvula is deformed or enlarged for any number of reasons (Table 1), or there is a traumatic or developmental abnormality, such as a cleft palate, the normally well-coordinated effort to forcefully bring food into the lower structures can be interrupted or interfered with, leading to a chocking sensation and even interrupted breathing [3]. Sometimes this can lead to a bolus of food becoming chronically attached to a pharyngeal surface, a phenomenon called "pooling [4]."

What would happen if there was a mass attached to the oral-facing surface of a uvula, interrupting the smooth push by the tongue along the uvula? Almost never has a uvula mass been discussed, other than constantly enlarging neoplasms, typically of adjacent tissues and of salivary origin [5]. Interference with speech or swallowing is not mentioned except very generically. Only one case of a non-enlarging mass this has been reported thus far: a 4×3 mm squamous papilloma was attached approximately halfway down the length of the uvula. This resulted in edema and thickening of the uvula and a sensation of "needing to swallow," but the uvula was not lengthened significantly [6]. We present the first case with a mass on the very tip of a uvula, leading to, apparently, repeated attempts to swallow it, resulting in a very long, narrow uvula.

Case summary

A 29-year-old male visited a dentist, at the suggestion of his family physician, for evaluation of a "thing dangling down his throat." The patient first noticed a gag reflex nine months earlier. The gagging became more severe and more frequent over time, especially when eating, and in recent weeks he had also noticed that his speech had developed a "nasal sound." An abnormally long uvula was seen to extend down from the soft palate, out of sight into the oropharyngeal region. He has had "a snoring problem" since college and was recently diagnosed with sleep apnea and started using a CPAP device. He was subsequently referred to an oral and maxillofacial surgeon.

Clinical features

At exam the elongated uvula was obvious and, when asked, the patient was able to cough it up into his mouth (Figure 1). The uvula was very thin and somewhat erythematous, but not hemorrhagic or tender. It had an oval or "saugage-shaped" soft tissue mass attached to its tip, measuring 5 mm in greatest length. The uvula was approximately 2.5 cm. long and rested just above the epiglottis when in its "normal" or resting position, as per the oral surgeon. The soft palate itself appeared unremarkable, as were all pharyngeal walls. This patient also presented with a coated tongue, mostly likely secondary to long-term mouth breathing and its subsequently xerostomia.

The surgeon excised the mass as well as most of the elongated uvula. Microscopically, the uvula was not particularly abnormal throughout its length, with minimal edema, mild congestion and a few chronic inflammatory cells beneath an atrophic epithelium. The mass on the tip (Figure 2) proved to be different, being comprised of dense and rather avascular fibrous tissue with scattered chronic inflammatory cells (more than in the uvula "stalk") beneath a covering of hyperplastic squamous epithelium. A diagnosis of irritation fibroma was made. The epithelium had a slight excess of keratin on the surface, consistent with frictional keratosis, apparent evidence of chronic surface irritation.

The surgical site healed well, and the patient's gagging problem and nasal speech no longer were present within days after the surgery.

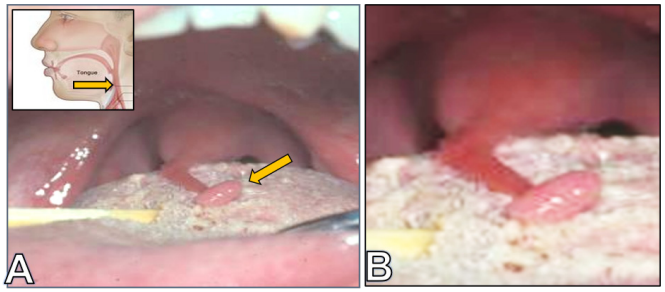

Figure 1: Clinical appearance of a mass on a long, narrow uvula; for the photo, the patient coughed the mass from his throat onto his tongue. **(A)** Arrow points to oval shaped soft tissue mass on the end of a 2.5 cm. long uvula, with the arrow in the inset pointing to the estimated location of the mass when it is in its normal position in or just above the larynx (inset photo from NCI); **(B)** Close-up of the mass. Patient also has a coated tongue.

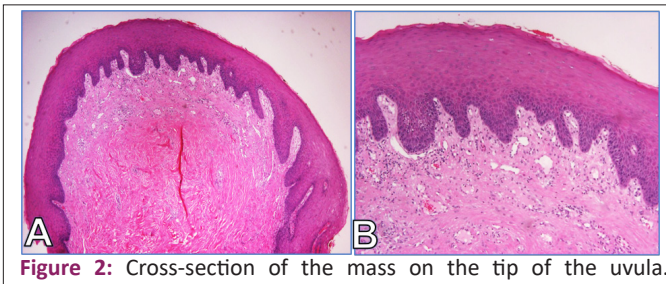

Figure 2: Cross-section of the mass on the tip of the uvula. **(A)** Pedunculated mass consists of dense fibrous connective tissue covered by hyperplastic squamous epithelium with mild hyperkeratosis (frictional keratosis); **(B)** Higher power shows lymphocytes (small dark blue dots) beneath the epithelium and partially detaching surface keratin.

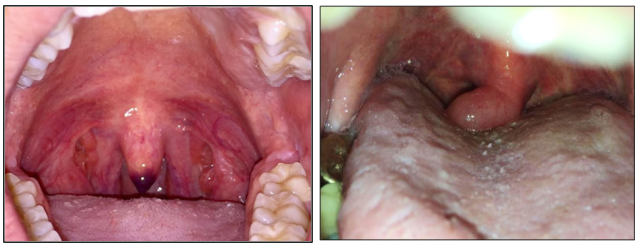

Figure 3: Examples of edematous, elongated uvulas. **(A)** Secondary to fellatio trauma, with a hematoma at the tip (courtesy of Dr. Jose Tapia, University of Buffalo, Buffalo, NY, used with permission; **(B)** Cocaine uvula, with considerable semitranslucent edema and considerable elongation.

Table 1: Phenomena previously reported to be associated with, perhaps causing, an elongated uvula [1-3]. It should be noted that some individuals will have more than one of the reported issues.

Problems associated with (Causing?) an Elongated Uvula
Allergies (pollens, pet dander, dust, etc)
Angioedema (angioneurotic uvula edema, Quincke's edema)
Autoimmune attack against mucosa, e.g. lichen planus
Chronic cough
Continual hawking (from foreign body sensation in throat)
Dehydration (may be result of mouth breathing)
Fellatio trauma
GERD (acid reflux)
Hereditary angioedema
Intubation, throat surgery
Mouth breathing (sinusitis may be involved)
Obstructive sleep apnea, snoring, especially with obesity
Professional singing or oration
Pycnodysostosis
Smoking tobacco, cocaine, crack, marijuana, methamphetamine
STI (gonorrhea, syphilis, chlamydia, herpes)
URI, Strept throat
Uvula angioedema (rare consequence of Covid-19 vaccine)
Vaping

Discussion

The earliest detailed discussion of diseases of the uvula is an 1849 review paper which combined several theses and articles from as early as 1822 [1]. These authors specifically described a uvula that was "loose, and hangs down much farther than in the natural and healthy state" as a feature of pharyngitis, especially as seen in the 1830 "epidemic influenza." Recent reports have also emphasize an association with laryngitis and edematous epiglottitis, with some authors suggesting that epiglottitis is actually the most common cause of uvulitis [7-9]. The 1849 report noted that "disease of the uvula. is one of the most frequent maladies of the voice to which singers and public speakers are liable," it "may diminish the volume and compass of the voice" and mentions "some professional singers, in which one, two, or even three notes were lost in consequence of disease of the uvula." They also reference earlier works suggesting that the "elongated uvula acts most on the larynx and glottis, and consequently induces cough and feeble or disordered voice."

Old and new reports discuss an enlarged uvula altered by edema and increased vascularity (congestion or hyperemia or both), not a thin, "stretched" structure, as seen in the present case [10,11]. (Figure 3) illustrates typical examples of this, showing a considerable difference between our case and the typical case, although we have found at least two other examples in which the uvula was so long that it could be flipped forward onto the lingual dorsum [12,13].

Today we know of many additional causes and/or associations with enlarged uvulae, as outlined in (Table 1). Snoring, with its subsequent dehydration of oral and pharyngeal membranes, is probably the most common cause of this phenomenon, especially in persons with sleep apnea, similar to the man in our present case. Also common is uvulitis is epiglottitis, and both are frequently associated with dyspnea, pharyngitis, erythema of all pharyngeal structures, fever [14,15].

The present case appears to be a uvula that became elongated from snoring (sleep apnea) and when it subsequently developed an irritation fibroma on its tip, the mass was repeatedly "dragged" down into the hypopharynx in a misguided attempt to swallow it as if it was a bolus of food. In doing so, the uvula was not only stretched extremely but became quite thin in the processes, with a subsequent hyperkeratosis created in response to the repeated friction or compression. This eventually created the longest uvula ever reported (as per our literature review), at 2.5 cm in length, but it was also very unusual, even unique because it was so thin; typically, an elongated uvula is widened, as much as 3-4 times its normal width, from edema, with only a few mm of elongation (Figure 3).

As previously mentioned, swallowing solid food starts with the "oral propulsive stage," in which the anterior tongue rises to meet the hard palate, the posterior tongue drops and opens the oropharynx and the bolus is pushed down the palate by the tongue toward the oropharynx; this requires coordination of 5 different muscles, controlled symmetrically by cranial nerves III, X and XII [15]. Once in the oropharynx, 23 different muscles (mostly working symmetrically) are involved, controlled by cranial nerves III, VII, IX, X, XII. This muscle activity is designed to bring a bolus of food from the top (oral) to the bottom (esophagus) of the pharynx, and it works remarkably well, unless the "bolus" is actually attached to the end of a uvula. The present uvula-tipped fibroma was most likely repeatedly "dragged down" into the pharynx, even into the larynx, with each swallowing reflex, a process which would have stretched the uvula and kept it thin.

Regardless of the cause, simple surgical excision of the enlarged part of the uvula is the appropriate treatment, just as it was recommended in 1849, although today perhaps one of the available sleep apnea therapies is added. Our patient had recently begun using a CPAP mechanical device, which should prevent a recurrence of the uvula elongation in the future.

Declarations

IRB exemption: The West Virginia University IRB rules exclude individual case reports from their purview.

Conflict of interest: None of the authors have conflicts of interest for this research.

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