



Congenital Epidermoid Cyst in the Uvula of a Child: Case Report and Literature Review

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Abstract

Epidermoid cysts are benign stratified-squamous epithelium-lined lesions that may be congenital or acquired (trauma or surgery). They are rare in the oral cavity, being more frequent on the mouth's floor, and extremely rare in the uvula. To the best of our knowledge, only 14 cases of epidermoid cysts in the uvula have been reported to date. We present a case of a 3-year-old boy with an incidental finding of a uvular epidermoid cyst. The pathogenesis, diagnosis, and treatment of this rare entity are discussed.

Keywords: Epidermoid cyst; Uvula; Congenital; Oral

Introduction

Epidermoid cysts are benign lesions with stratified-squamous epithelial lining containing fluid or semisolid material. Approximately 7% occur in the head and neck, and only 1.6% occur in the oral cavity, mainly on the mouth's anterior floor [1,2]. A comprehensive review of English and Spanish literature by searching the databases using the terms "epithelial cyst uvula" and "epidermoid cyst uvula", without date restrictions, disclosed only 14 confirmed cases [3-13]. We report a case of a 3-year-old male with a mass at the uvular tip. We provide a comprehensive review

of existing cases.

Case Report

A 3-year-old male with a history of isolated episodes of regurgitation came for consultation for a slowly-growing small white mass in the uvula that had been noticed by his mother for the past three months. The patient was in good health, and his medical history was uneventful. Physical examination revealed a well-circumscribed, smooth, yellow-white, oval lesion that measured approximately 1 cm located on the tip of the uvula (Figure 1).



Figure 1: Whitish 1 cm cyst in the tip of the uvula.

The rest of the physical examination was unremarkable. A clinical diagnosis of uvular cyst was made, and the lesion was completely excised under general anesthesia without surgical or postoperative complications. Grossly, the surgical specimen consisted of a smooth-surfaced, oval, soft, yellowish-white nodule that measured 7 x 6 x 5 mm. A thin fibrous tissue capsule formed the

cyst's wall, and the cut surface showed a central cavity filled with yellowish-white, pasty material. Microscopically, sections of the specimen showed a cyst with luminal keratin debris (Figure 2A), lined by stratified squamous epithelium without skin appendages and an underlying loose fibrous connective tissue (Figure 1A, 2B).

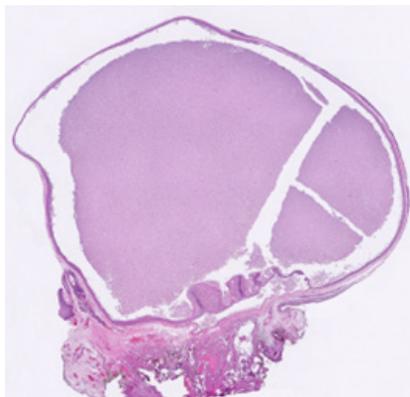


Figure 2A: Low-power view of the cyst. The lumen is packed with keratin debris (H&E, x10).

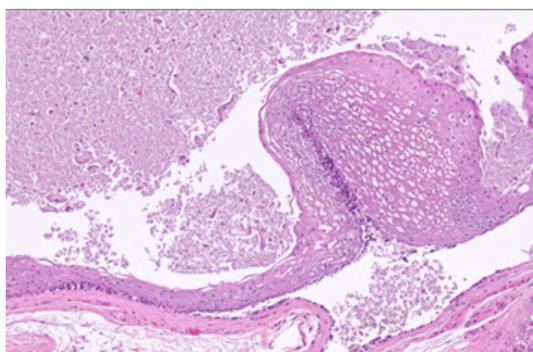


Figure 2B: High-power view of the cyst wall. Uniform stratified squamous epithelium without skin adnexa and loose fibrous connective tissue capsule (H&E, x100).

Based on these findings, the case was diagnosed as a completely excised epidermoid cyst of the uvula. The patient had no recurrence at a one-month follow-up.

Discussion

Epidermoid cysts are benign, slowly-enlarging lesions, either congenital or acquired, due to trauma or surgery. These cysts can arise anywhere in the body, with an incidence of 7% in the head-and-neck region and 1.6% in the oral cavity [2]. Only 14 cases have been described in the uvula (Table 1) [3-13].

In this rare location, they are predominant in males with a male:female ratio of 4:1, all in the pediatric population, with a median age of 9 months. Most epidermoid cysts are acquired lesions, most likely due to trauma or iatrogenic introduction of epithelial cells into deep tissue during surgery [10]. However, in the uvula, the pathogenesis seems to be congenital, as all cases have occurred in pediatric patients with no history of trauma or prior intubation. The origin is, therefore, likely due to a developmental defect where

remnants of ectodermal debris are trapped during the fusion of the maxillary process to form the palate between the 6th and 12th week of embryological development [4,8,10,11].

In this zone, they are generally asymptomatic, slowly enlarging lesions [4,5,7,10,12] that can eventually grow, causing obstructive symptoms such as snoring [3,8,11], gagging [3,11], dysphagia [3,11], mouth breathing [3,8], poor sucking reflex [6,8], and insufficient weight gain [6,8]. Some cases presented with simultaneous upper respiratory infections, which caused airway obstruction [11,13] and even required emergency surgery in one case [13]. Our patient's new onset regurgitation was likely caused as a result of this entity. Imaging is not required for diagnosis as a cyst in the uvula is easily identified on physical examination and accessible for surgical resection. They are usually located at the tip [10,13]; however, they may be present laterally [6,9], or behind the uvula [8]. MRI may be necessary if malignancy is suspected; however, no cases of malignant transformation have been described in the uvula [7,12].

Table 1: Epidermoid Cysts in The Uvula.

Author, year	#	Sex	Age	Location	Size	Symptoms	Treatment / follow-up
Andrews, 1962p [3]	1	Male	9 months	Uvula	1.0 cm	Gagging, snoring, dysphagia, noisy breathing. 3 months of growth.	Surgical excision. No recurrence at 6 months.
Yoshinari et al., 1986 [4]	2	Female	12 months	Uvula	0.6 x 0.4 cm	Asymptomatic.	Surgical excision.
Mu et al., 2002 [5]	3	Male	Neonate	Uvula	0.4 cm	Asymptomatic. Vascular insult in middle meningeal artery.	Surgical excision. No recurrence at 2 months.
	4	Female	1 month	Uvula	0.5 cm	Asymptomatic. Bronchiolitis.	Spontaneous rupture 24 hours post-examination. No recurrence at 2 years.
Suga et al., 2010 [6]	5	Male	1 month	Uvula (left base)	0.8 x 0.8 x 0.7 cm	Poor sucking reflex, insufficient weight gain.	Surgical excision. No recurrence at 5 years.
Hernández-Paz et al., 2011 [7]	6	Female	18 years	Uvula	1.0 cm	Asymptomatic. 8 years of growth.	Surgical excision.
Tsai et al., 2013 [8]	7	Male	7 months	Uvula (behind)	0.8 x 0.8 cm	Abnormal sucking and swallowing reflexes, snoring, noisy breathing, low body weight.	Surgical excision.
Kim et al, 2014 [9]	8	Male	14 months	Uvula (right side)	0.4 x 0.4 cm	Cleft lip and palate.	Surgical excision.
Daram et al., 2016 [10]	9	Male	5 years	Uvula (tip)	0.6 x 0.6 x 0.4 cm	Asymptomatic.	Surgical excision.
Sheshul et al., 2016 [11]	10	Male	12 months	Uvula	1.0 x 1.0 cm	Snoring.	Surgical excision.
	11	Male	1 month	Uvula	0.5 cm	Gagging.	Surgical excision.
	12	Male	3 months	Uvula	0.5 cm	Fever, drooling, elevated WBC.	Surgical excision.
Medina et al., 2017 [12]	13	Male	6 months	Uvula	1.0 cm	Asymptomatic.	Surgical excision.
Lim et al., 2020 [13]	14	Male	10 months	Uvula (tip)	2.0 x 1.0 x 0.5 cm	Dyspnea after URI.	Emergency surgical excision. No recurrence at 6 months
Sanchez et al., 2022 (current case)	15	Male	3 years	Uvula	0.7 x 0.6 x 0.5 cm	Regurgitation.	Surgical excision.
							No recurrence at 1 month.

WBC = White blood cells, URI = Upper respiratory infection

Macroscopically, epidermoid cysts in the uvula range from 0.4-2 cm (average 0.78 cm). They are well-circumscribed, spherical to oval, yellowish-whitish in appearance with a thin capsule. On the cut surface, they contain fluid or semisolid material. A histologic examination is required for a definitive diagnosis. Epidermoid cysts are characterized by a fibrous connective tissue wall lined by a layer of stratified-squamous epithelium without skin adnexa. The lumen is filled with variable amounts of keratin debris. Mass lesions in the uvula can be caused by dermoid cysts, teratoid cysts, mucoceles, and branchial cleft cysts. In 1955, Meyer established a morphologic classification for cysts on the floor of the mouth as either dermoid, epidermoid, or teratoid. Epidermoid cysts are lined only by stratified-squamous epithelium; Dermoid cysts also include skin adnexa; and Teratoid cysts include mature tissue representing at least two embryonic layers (endoderm, mesoderm, or ectoderm) [14]. Mucoceles are not lined by any kind of epithelium. Branchial cleft cysts are lined by stratified-squamous or ciliated columnar epithelium and have a fibrous wall with lymphoid follicles.

Complete transoral surgical excision is the treatment of choice, even in neonates [3-13]. Early excision can prevent clinical complications such as airway obstruction [13]. The well-defined borders of the cysts facilitate surgical resection, and no recurrence has been described. Fine needle aspiration and fenestration are not recommended due to the risk of infection, which can cause pain and worsen clinical symptoms [6]. On rare occasions, the cyst can spontaneously rupture without further recurrence [5].

Conclusion

Epidermoid cysts in the uvula are rare benign tumors formed by ectodermal entrapment during the fusion of maxillary processes to form the palate. It presents more commonly in males, and all cases have presented in pediatric patients. Clinical presentation is usually asymptomatic but may cause obstructive symptoms when size is considerable. In the evaluation of cystic lesions of the uvula, the differential diagnosis should include epidermoid cyst, dermoid cyst, teratoid cyst, mucocele, and branchial cleft cyst. Complete surgical excision provides both the definitive diagnosis and curative treatment, and the procedure is usually well tolerated. Pediatricians, otolaryngologists, and pathologists should be aware of the occurrence of epidermoid cysts in the uvula, as early detection and management prevent complications.

Conflict of Interest

No conflict of interest.

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