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Case Report

A rare case of paediatric sublingual keratinous cyst

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ABSTRACT

Background: The occurrence of keratinous cyst in the oral cavity is extremely rare. Keratinous cyst in the floor of the mouth is painless, doughy or fluctuant lesion and causes no symptoms until it is large enough to interfere with speech or eating. Cyst generally presents as slow and progressive growth and often not diagnosed until the second or third decade of life.
Case Report: We report an uncommon presentation of keratinous cyst of the sublingual space in a 6 year old female child. The swelling was mimicking ranula on inspection. On complete examination, the swelling appeared as epidermoid cyst. After a routine evaluation of the sublingual mass, surgical excision was performed under general anaesthesia.
Conclusion: This case report addresses novel information regarding simple yet effective way of managing a sublingual keratinous cyst in a paediatric patient.

1. Introduction

In paediatric age group of under 15 years 90% of the neck masses are benign and of these up to 55% are congenital.[1] During third and fourth embryonic weeks, fusion of the first and second branchial arches takes place. The keratinous cysts occur secondary to embryonic entrapment of endodermal and ectodermal remnants. As a result, most often these keratinous cysts are seen in the midline. The intraoral keratinous cysts are rare and accounts for <0.01% of all the cysts.[2] The keratinous cysts are classified as. These keratinous cysts need thorough clinico-pathological examination. The diagnosis of have been one of the challenges for the surgeons as they mimic many other benign cysts of the head and neck and also due to their relative rarity. All though the final diagnosis of keratinous cyst is made by histopathological analysis, thorough examination and additional imaging is of great importance for preoperative analysis of the swelling character and its relation with the surrounding structures.

2. Case Report

A 6 year-old otherwise healthy girl presented to the Otorhinolaryngology outpatient services for second opinion of a persistently increasing mass in the floor of oral cavity, after a formal evaluation of the mass initially at an outside institution. A small solitary asymptomatic swelling in the floor of oral cavity was present since birth. For the past 2 years, mother had noticed gradual increase in size of the swelling. Mother also gave history of difficulty in chewing and swallowing large bolus of food. The child had no difficulty in breathing or no history of difficulty in articulation of speech.

Examination of the girl revealed a well-defined solitary swelling in the sublingual region, measuring around 4x3cms. The buccal mucosa, lining the swelling appeared to be normal. [Figure 1]

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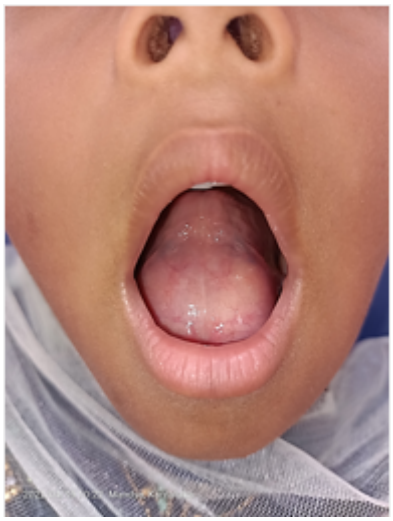

Fig. 1: "Second tongue" appearance. Intra-oral view, when the tongue is pushed against the hard palate.

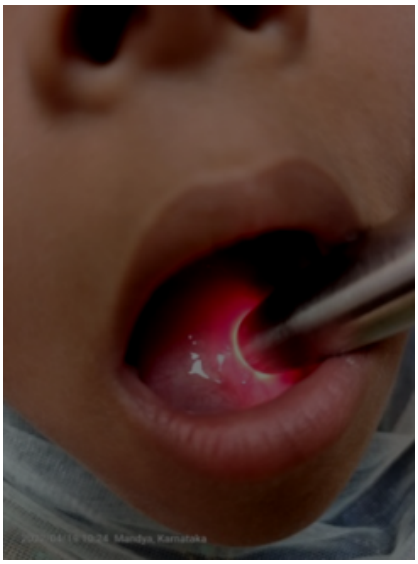

Fig. 2: Transillumination test demonstrating the swelling to be non transilluminant.

On palpation the swelling was nontender with rise in temperature. The swelling were tense and non-mobile. Fluctuation test and trans-illumination test of the swelling was found to be non-fluctuant and non-trans-illuminant respectively. [Figure 2]

On neck examination, there was fullness of sublingual and submandibular region. Bimanual examination of it confirmed that the fullness of sublingual and submandibular region was secondary to the mass in the floor of oral cavity. No other swellings were palpable in the oral cavity or neck. [Figure 3]

During the process of evaluation, Non-Contrast Computerized Tomography (NCCT) reported as

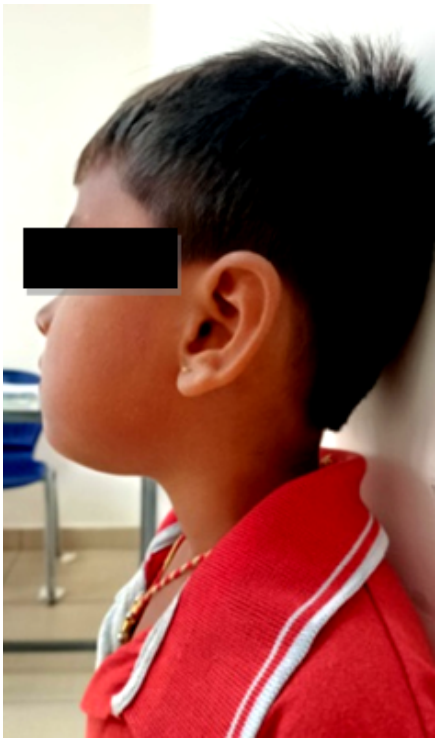

Fig. 3: Lateral view. "Double chin" appearance. Fullness of sublingual and submandibular region

4x2.7x4.1cm well circumscribed cystic lesion located in the sublingual space indenting the mylohyoid muscle with extension to the adjacent submandibular space. Radiological features were suggestive of Ranula. [Figure 4a,b]

Since clinical examination was not characteristic of ranula, differential diagnosis of dermoid or keratinous cyst was made.

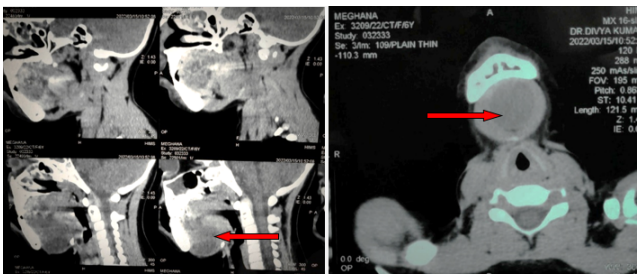

Fig. 4: a and b: NCCT imaging showing a well circumscribed sublingual swelling.

As Ranula and dermoid/ keratinous cyst were the differential diagnosis, excision biopsy under general anaesthesia was planned rather than Marsupialization.

Steps of the surgical procedure were as follows,

1. Adequate mouth opening was achieved using self-retaining Doyen's mouth gag. [Figure 5]

2. Sublingual region was exposed retracting the tongue superiorly with a stay stitch. [Figure 5]
3. Hydro dissection was carried out to get the plane between mucosa and the sublingual mass using diluted 1:1 local infiltration of 2% LOX with 1:2,00,000 adrenaline and normal saline. [Figure 6]
4. A vertically elliptical incision was given around the lingual frenulum in the midline and care was taken to avoid injury to the Wharton's duct opening in the floor of oral cavity. [Figure 7]
5. The sublingual mass was dissected all around from the floor and base of the tongue and excised completely. The specimen was sent for histopathological examination. [Figure 8]
6. Genioglossus, hyoglossus, lingual nerve and Wharton's duct were visualized, traced bilaterally and preserved.
7. Surgicel was used to fill up the potentially in the sublingual region. This was intended to avoid post operative hematoma formation and asphyxia secondary to airway obstruction in the child. [Figures 9 and 10]

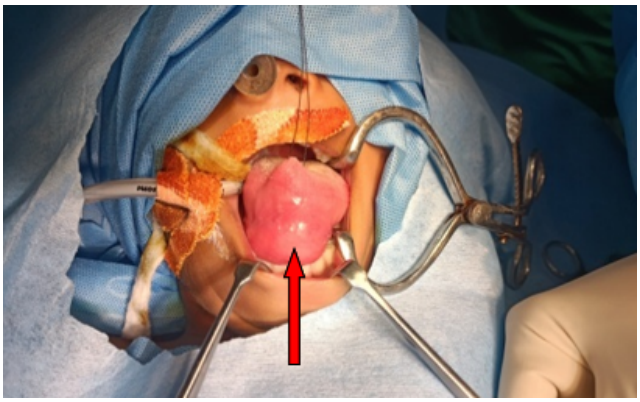

Fig. 5: Adequate mouth opening with Doyen's mouth gag and tongue sutured.

The child recovered well with no intraoperative or postoperative immediate or delayed complications. No recurrence was observed in the follow-up.

2.1. Histopathology

Macroscopic examination of the revealed solitary globular encapsulated sublingual mass with smooth regular surface. Pearly white in colour. The capsule was thick and as a result, the mass was non-transparent. It was 5.5x4x3cms in dimension. [Figure 12] The mass was doughy, cystic in consistency and fluctuant in nature. Transillumination was found to be negative. The cut surface revealed thick pultaceous material with one small white hard nodule measuring 0.25cm noted in the cyst wall.

Microscopic examination of the specimen was as follows-

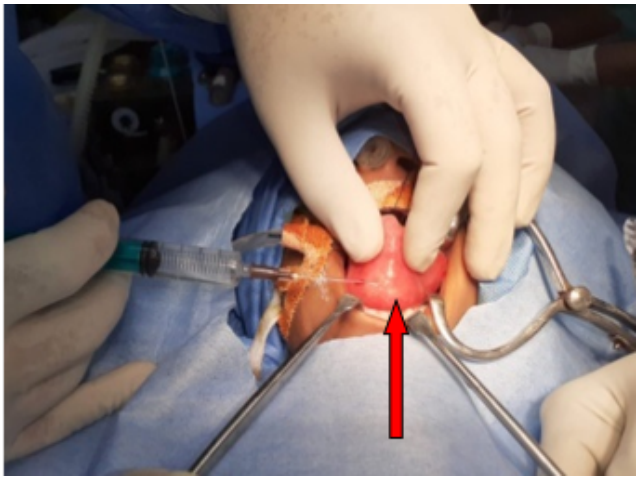

Fig. 6: Hydro dissection

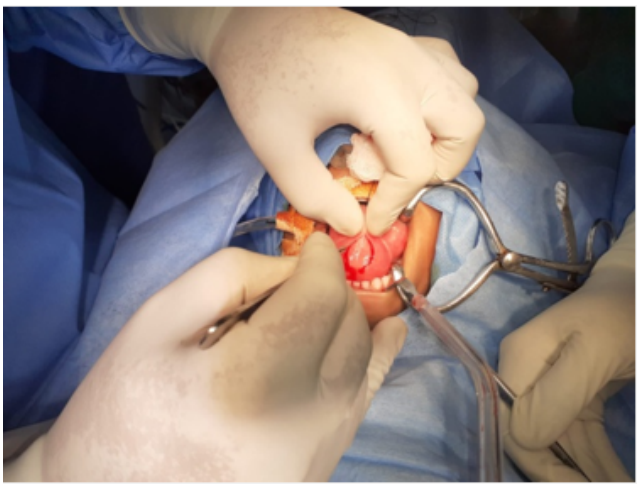

Fig. 7: Elliptical incision along the midline.

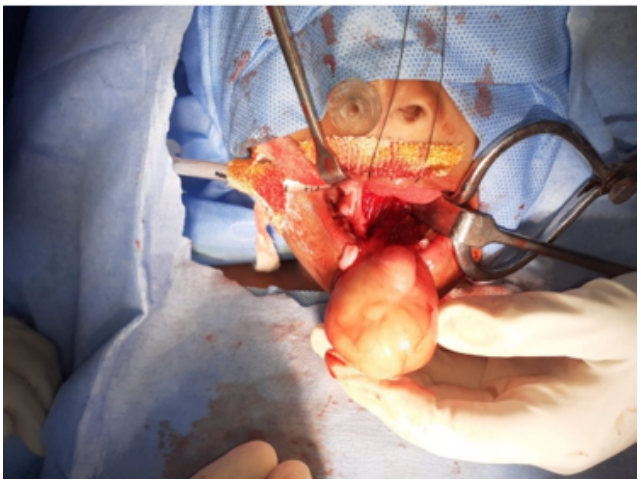

Fig. 8: Encapsulated sublingual mass intoto.

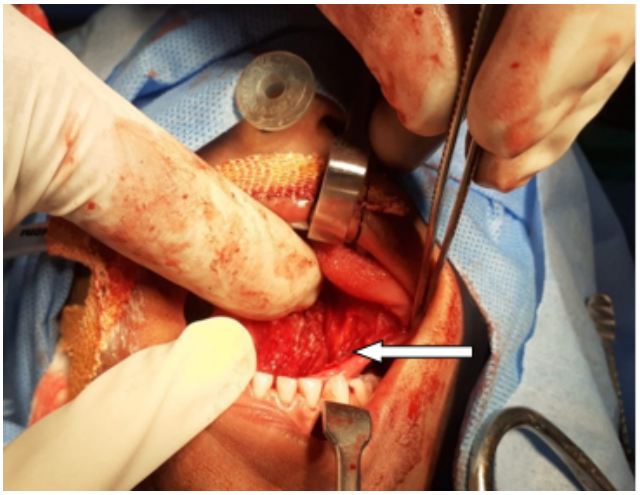

Fig. 9: Potential sublingual dead space.

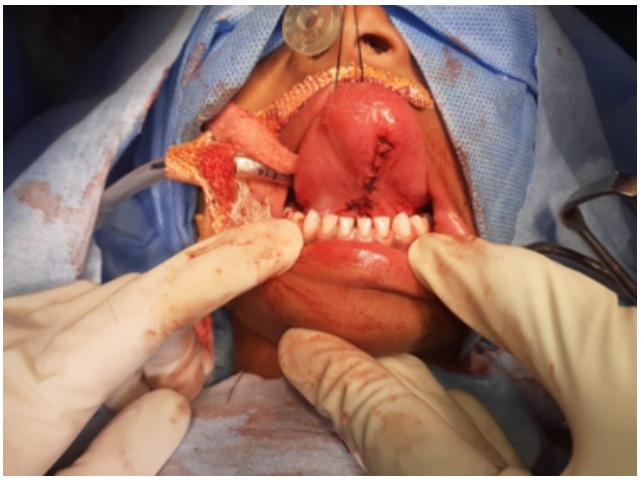

Fig. 10: Incision sutured in layers

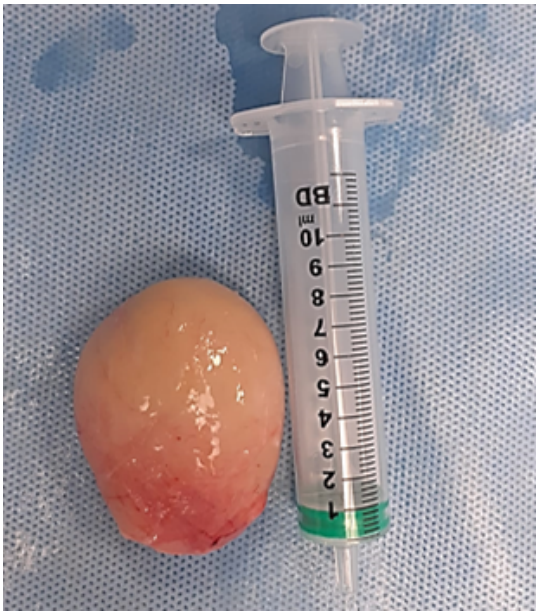

Fig. 11: Gross appearance of the excised sublingual mass.

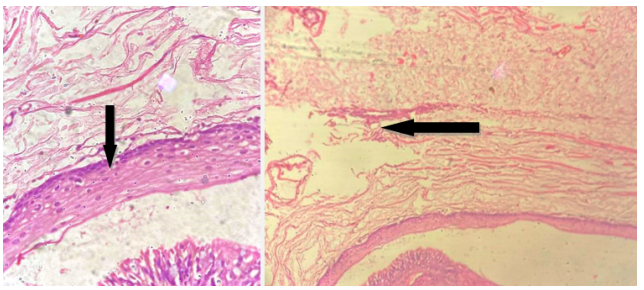

Fig. 12: Stratified squamous epithelium lined cyst with keratin flakes.

The cyst was lined by stratified squamous epithelium. The subepithelial tissue shows congested blood vessels and skeletal muscle bundles. Within the lining epithelium, mucus secreting cells were also found to be embedded. Keratin flakes were noted. Focal areas of ciliated columnar metaplasia were also observed. [Figure 12]

3. Discussion

Sublingual masses reside within a space bounded laterally by the mandible, inferiorly by the mylohyoid muscle, and superiorly by the floor of mouth mucosa

The differential diagnosis for sublingual masses also includes ranula, keratinous cyst, epikeratinous cyst, branchial cleft cyst, lymphatic malformation, mucocele, haemangioma, thyroglossal duct cyst, and infection.[1]

Overall, only 7% of all the epikeratinous and keratinous cysts are found in the head and neck region. 1.6% are found in the oral cavity.[2] Sublingual epikeratinous and keratinous cysts are uncommon, with keratinous cysts accounting for 1.6% and epikeratinous cysts representing 0.01% of the sublingual cysts.[1]

Keratinous cysts are lined by stratified epithelium. They may keratinize either by forming keratohyalin or without formation of keratohyalin. Thus, keratinous cysts are of two types:

1. Epikeratinous cysts (Stratified squamous epithelium with keratohyalin granules).
2. Tricholemmal cysts (Stratified squamous epithelium without keratohyalin granules).[3]

Keratinous cysts are classified as epikeratinous, true keratinous and teratoid cysts depending on the types of tissues identified pathologically by Meyer in 1995.[4]

1. Epikeratinous cyst: Contains only skin, but no other adnexal structures. It is lined with squamous epithelium with or without keratinous material.

2. True keratinous cyst: Contains skin with appendages such as hair, hair follicles, sebaceous glands and sweat glands. It is lined with squamous epithelium.
3. Teratoid cyst: Contains all three embryological elements: ectodermal, endodermal and mesodermal elements, such as nails, teeth, brain and glandular tissue. It is lined either with squamous or respiratory epithelium.

Epikeratinous, true keratinous, and teratoid cysts are non-odontogenic benign lesions. They are derived from the germinative epithelium. These cysts can be found anywhere in the body. These cysts often remain asymptomatic.[4]However they may become symptomatic as a result of enlargement or superinfection.

Conventional keratinous cysts are <5cms. Cysts measuring 5cms or more than 5cms are referred as Giant epidermal cysts.[3]

A Keratinous cyst can either be congenital or acquired based on its etiopathology.

1. A congenital epikeratinous cyst is formed by trapping displaced embryonic epithelium occurring in the head and neck area.
2. An acquired epikeratinous cyst results from implantation of viable epidermal cells in the dermis or subcutis either secondary to surgery or trauma.[5] The posttraumatic cysts are also called as implantation keratinizing keratinous cysts.[4]

3.1. Secondary malignancy

The most common malignancy associated with keratinous cysts is squamous cell carcinoma and basal cell carcinoma is the second most common malignancy transformation seen in the keratinous cysts. Other malignancies reported are as follows- Bowens disease, Paget's disease and Markel cell carcinoma.[3,6]

3.2. Etiopathogenesis

1. HPV 57 or 60 may act as an additional factor for the development of palmoplantar keratinous cysts.[3]
2. Epikeratinous cysts are also found to be associated with Gardner syndrome/ familial adenomatous polyposis, Gorlin syndrome/basal cell nevus syndrome, Ehlers-Danlos syndrome and pachyonychia congenital.[3,6]
3. Patients with BRAF inhibitors can develop epikeratinous cysts on the face.[6]
4. Imiquimod and cyclosporine have been noted to be able to cause epidermal inclusion cysts.[7–9]

3.3. History and examination

They are usually slow growing progressive swelling. Always tend to be asymptomatic in nature. Clinical presentation of the keratinous cyst in the floor of oral cavity depends on the size and positional relationship of the cyst to the mylohyoid muscles. Keratinous cysts above the mylohyoid muscle manifests as a swelling in the sublingual region. Displace the tongue secondary to the size of the cyst, symptoms as dysphagia, dysphonia and dyspnoea can occur. Keratinous cysts below the mylohyoid muscle manifests as a swelling in the submental region, causing a double chin appearance.[10,11] On palpation the cyst appears dough like painless mass without any lymphadenopathy.[11]

3.4. Management

Aspiration biopsy is commonly used but is not definitive. To differentiate between vascular, salivary and mucosal lesions and also to diagnose, specialized imaging techniques such as ultrasonography (USG), computed tomography (CT) and magnetic resonance imaging (MRI) are carried out.[4]

Ultrasonography is the first choice of imaging technique as it is quick, non-invasive, reliable and economical. Ultrasound examination may help to differentiate between solid, vascular and cystic lesions. the keratinous cyst are well-circumscribed, smooth mass with a heterogeneous interior.[11]

On computed tomography (CT), the cyst appears as moderately thin walled, unilocular mass filled with a homogeneous, hypo-attenuating fluid substance with numerous hypo-attenuating fat nodules. This gives the pathognomonic "sack of marble" appearance.[12]

On magnetic resonance imaging (MRI), the lesion appears as a well-circumscribed mass. The signal intensity of keratinous cysts is high in T2-weighted images and low in T1-weighted images.[4,13]

4. Conclusion

Big keratinous cysts involving floor of mouth in paediatric age group poses many clinical challenges in diagnosis and definitive treatment. Proximity of important structures such as salivary duct and lingual nerves makes the definitive surgery challenging. Radiology may not always give diagnosis or even confuse the treating surgeon as happened in case presented above where in CT scan was suggestive of ranula. In such a situation surgeon needs to consider findings of clinical examination and prepare for alternative surgical procedure depending on findings during the surgery.

5. Source of Funding

None.

6. Conflict of Interest

None.

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