CASE REPORT

Simultaneous Occurrence of a Midline Sublingual Dermoid Cyst with Respiratory Epithelium and Submental Dermoid Cyst in a Paediatric Patient: A Case Report and Review of Literature

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Abstract

Aim To present the case of a simultaneously occurring sublingual dermoid cyst with respiratory epithelium and a submental dermoid cyst, and also to review literatures on the same.

Methods A complete examination, radiograph, MRI and excision biopsy of both lesions were carried out and diagnosis was arrived at histopathologically.

Results The histopathology of the submental swelling revealed orthokeratinized stratified squamous epithelium with underlying connective tissue consisting of dense irregularly arranged collagen fibres with fibroblasts along with chronic inflammatory cell infiltrate of lymphocytes and plasma cells. There was presence of sebaceous glands and sebum. Sublingual swelling showed non-keratinized stratified squamous epithelium with a fibrovascular connective tissue. Areas of pseudostratified ciliated columnar epithelium were also evident in some areas. Presence of sebaceous glands also seen. Based on these findings a confirmatory diagnosis of sublingual dermoid cyst with respiratory epithelium and submental dermoid cyst was made.

Conclusion This report throws light on midline lesions of the oral cavity and hopes to add this rare case into the current differential diagnosis of the same.

Keywords Sublingual cyst · Midline swelling · Dermoid cyst · Histopathology · Simultaneous lesions

Introduction

Swellings of the head and neck have a plethora of reasons which could be congenital, developmental, traumatic, inflammatory, infectious or idiopathic. Simultaneously occurring swellings intrigue the diagnostician of its origin and diagnosis. Very few cases have been published in literature on simultaneously occurring swellings and even more rare are for them to occur in the midline. Midline swellings usually point towards fusion defects and lesions arising from midline structures. Here we present a very rare case report of simultaneously occurring midline sublingual dermoid cyst with respiratory epithelium and a submental dermoid cyst. The presence of respiratory epithelium in the sublingual cyst is also very intriguing and its possible pathogenesis has been described. Review of literatures till date have also been added for a better understanding of this rare entity. Here below we report this atypical presentation.

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Case History

A 5 year old boy was brought to our Craniofacial Centre by his parents for evaluation of two swellings in the submental and sublingual area. Patient also complained of difficulty in swallowing. History revealed that the sublingual swelling was present since birth and the submental swelling was

noted 3 days after birth. There was a gradual increase in size in the sublingual swelling leading to progressive dysphagia and difficulty in closing the mouth. The submental swelling also had a similar progression in size till the presentation date. Parents did not give a history of trauma, pain or fever from childhood.

Patient was not in respiratory distress and immediate intervention was not warranted. Patient was moderately built and nourished. The medical history and review of systems was unremarkable.

Extra oral clinical examination of the anterior submental region revealed a rounded midline swelling measuring 1.5×1.5 cm. The surface of the anterior submental swelling was smooth and the skin over the swelling was pinchable. On palpation the swelling was non tender, soft, compressible and mobile along all planes. The margins of the swelling were diffuse.

Extra oral clinical examination of the posterior submental swelling; which was an indentation of the sublingual swelling revealed a single, rounded, diffuse swelling measuring 4×3 cm². Intraorally the floor of the mouth was raised and the tongue was elevated. The mucosa over the swelling was normal. On palpation the swelling was non-tender, soft, compressible and immobile. On bimanual palpation the sublingual swelling was continuous with the posterior submental swelling, whereas the anterior submental swelling was clinically distinct from it. No surface changes were seen. Function of Whartin's duct was normal and no paresthesia or anesthesia was noted. Transillumination test was negative.

Orthopantomogram and occlusal radiographs were non-contributory. MRI study showed a T2 weighted hyperintense cystic area, measuring 4×3 cm² in the floor of the mouth above the level of mylohyoid. A smaller hyperintense cystic area was seen below the mylohyoid and geniohyoid measuring 1.4×1.2 cm.

Based on clinical and radiographic examination, a provisional diagnosis of sublingual and submental dermoid cysts was derived.

The patient underwent surgical excision of both swellings under general anaesthesia (Fig. 1). Under endotracheal intubation after standard preparation and draping, the patient underwent excision of both the lesions separately submental followed by sublingual intraoral mass. A 3 cm vertical incision was placed in the submental area. Careful dissection was carried out around the capsulated mass to remove it in toto. No breach in capsule was done to avoid leaving behind any residual lining which may cause recurrence. The wound was closed in 2 layers with vicryl and 5–0 ethlion sutures for best esthetic results. A pressure dressing also applied submental area to prevent hematoma collection.

The intraoral sublingual mass was approached with similar vertical incision clearing the submandibular ducts 5 mm away from it. Meticulous submucosal dissection was performed as the lesion was big and had a chance for lining perforation. Sublingual hemostasis achieved with bipolar cautery and the mass was totally enucleated from the bed of muscle of tongue and underlying structures. Primary closure was done by using 5–0 Vicryl by leaving some area for drainage of collected blood to avoid any sublingual hematoma which may jeopardize the airway of the patient. Postoperative edema was present for 2 days, however was managed with simple anti edema drugs. No recurrence or any complications reported for 1 year following surgery.

The histopathology of the submental swelling revealed orthokeratinised stratified squamous epithelium with underlying connective tissue consisting of dense irregularly arranged collagen fibres with fibroblasts along with chronic inflammatory cell infiltrate consisting of lymphocytes and plasma cells. There was presence of sebaceous glands and sebum. Numerous endothelium lined blood vessels of various sizes and shapes were present. Vascularity is increased. These findings were consistent with the diagnosis of Submental Dermoid cyst (Fig. 2a, b).

Sublingual swelling showed non-keratinized stratified squamous epithelium with a fibrovascular connective tissue. Areas of pseudostratified ciliated columnar epithelium is evident in some areas. The epithelium is 4–8 cell layers in thickness and exhibit hyperplasia in few areas. Connective tissue showed loosely arranged collagen fibres with fibroblasts and increased vascularity. Sebaceous glands are also evident (Fig. 3).

Based on these findings a confirmatory diagnosis of sublingual Dermoid cyst with respiratory epithelium was made.

Discussion

Epidermoid and dermoid cysts are lesions derived from the germinal epithelium and represent less than 0.01 % of all the oral cavity cysts. The hypothesis of their etiology is that they arise from entrapment of epithelial debris or rests during the midline fusion of the first two branchial arches [1, 2]. This has also given rise to the term “dysontogenetic” used first by Erich [3].

Histologically, Meyer classified oral dermoid cyst into 3 groups [4]

1. Epidermoid cysts- A keratinizing squamous cell epithelial lined cavity surrounded by a capsule with no skin appendages.
2. Dermoid cyst- A keratinizing squamous cell epithelial lined cavity with skin appendages such as hair

Fig. 1 Excision of submental and sublingual cysts

Fig. 2 a Submental cyst showing orthokeratinized stratified squamous epithelium with underlying connective tissue showing sebaceous glands (under $\times 10$ magnification). **b** Submental cyst showing sebaceous glands (under $\times 60$ magnification)

Fig. 3 Sublingual cyst showing respiratory epithelium with underlying connective tissue showing sebaceous glands (under $\times 10$ magnification)

follicles, sweat and sebaceous glands in the underlying connective tissue.

3. Teratomas- A squamous cell or respiratory tract epithelial lined cavity with skin appendages and mesodermal elements such as bone, muscle, respiratory and gastrointestinal tissues.

Our reported case contained sebum and sebaceous glands in the submental swelling consistent with a dermoid cyst. The sublingual swelling consisted of respiratory epithelium in certain areas along with stratified squamous epithelium and underlying connective tissue containing loosely arranged collagen fibres with fibroblasts and also few sebaceous glands were evident. A diagnosis of sublingual dermoid cyst with respiratory epithelium was made. The reported case gives history of sublingual swelling from birth and submental swelling 3 days after birth which proves both the swelling are developmental in origin. The probable reason for this cyst could be developmental derangement of the foregut though there exist various theories regarding embryogenesis of respiratory lined cysts.

Cysts with respiratory epithelium are rare in the oral cavity and are termed as lingual choristomas. They usually present as a small, soft, compressible nodule lying entirely within the tongue or in floor of mouth and maybe also called foregut duplication cysts [5, 6]. A review of literature shows involvement of tongue and few cases involving floor of mouth lined by respiratory epithelium and there is lacking of gastrointestinal epithelium [5] similar to our reported case. As both the respiratory and gastrointestinal are of endodermal origin, it is conceivable that primitive uncommitted endoderm could differentiate in a multidirectional manner [7]. Due to the proximity of the primitive foregut to the pharyngeal arches, the embryonal rests may

be misplaced and entrapped between any parts of tongue in the fourth week. The pluripotential entrapped rests are exposed to different environments and inductive influences and can differentiate to respiratory or gastrointestinal epithelium. Terminology for the cyst is also based on descriptive histopathology rather than the pathophysiology [8].

According to Gorlin [9] in 1957 even odontogenic cysts can contain respiratory epithelium, and similarly respiratory epithelium in an apical periodontal cyst of mandible has also been reported [10].

These cysts are usually benign, however there have been dermoid cysts with lymph node metastasis noted only in cysts located outside oral cavity [11].

Respiratory epithelium lined cysts are asymptomatic in floor of mouth but there has been reports of about 30 % of cases who had difficulty in swallowing, speech, respiration and feeding. Our case reported to us with similar symptoms except for respiratory difficulty probably because of the slow expansion and late presentation of the swelling.

The most common causes for sublingual swellings include [12]

1. Ranula
2. Dermoid cysts
3. Thyroglossal cyst
4. Haemangioma
5. Lymphangioma
6. Sialoliths obstructing submandibular duct
7. Sialadenitis
8. Oral alimentary tract cyst
9. Cystic teratoma

Literature revealed oral cysts lined by respiratory epithelium and squamous epithelium with varying age, sex and histopathological criteria as tabulated below [13].

Author of article	Age of patient	Sex of the patient	Histopathology
Manor et al. [8]	11 years	Male	Respiratory epithelium and cuboidal cells
Kim et al. [14]	27 years	Male	Respiratory epithelium, squamous epithelium and calcium deposition
Ho and Crean [15]	6 weeks	Male	Keratinized squamous epithelium with fibrous tissue, adipose tissue, sebaceous glands and hair follicles
Burkart et al. [16]	Infant	Male	Squamous epithelium, interspersed focally by respiratory type epithelium, mucous glands, and dermal appendages
Burkart et al. [16]	Infant	Male	Squamous epithelium and focal areas of ciliated pseudostratified respiratory epithelium, mucus glands, and lymphoid tissue

Our reported case reveals, simultaneous occurrence of two swellings one after other within a short period of time in the oral cavity, with similar literature evidence. In spite of its developmental origin, these cysts are of varied age presentation. Simultaneous occurrence of a midline sublingual dermoid cyst with respiratory epithelium and submental dermoid cyst are likely to occur.

Conclusion

This case report aims to add to existing evidence of the likelihood of such simultaneous occurrences of midline swellings and the possibility of respiratory epithelium to be taken into the diagnostic probability. This paper also reaffirms the various inductive influences oral ectoderm may face during development. Thorough clinical examination is mandatory while evaluating midline lesions as there may be possibility of having two lesions masquerading as a single lesion. Although dermoids are usually solitary, we recommend multiple cyst evaluation

immaterial of age with Magnetic Resonance Imaging scan before attempting a surgery.

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