Case Report

Dermoid Cysts of the Floor of the Mouth in the Pediatric Patient: Report of Two Cases
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Abstract
Dermoid cysts of the floor of mouth are rare in the pediatric population and account for only 0.01% of all oral lesions. They arise as a result of failure of the surface ectoderm to separate from the underlying structures. In the pediatric patient, the initial differential diagnosis is often challenging due to multiple similar appearing lesions, which can occur in the floor of mouth. We report two cases of dermoid cysts in the floor of mouth, and briefly discuss the etiology, differential diagnosis, and treatment in the pediatric patient.

Keywords: Dermoid cyst; Floor of mouth; Oral cavity; Pediatric; Embryonic; Congenital.

Introduction
Dermoid cysts developing in the floor of mouth result from an abnormal embryonic growth, due to the entrapment of the ectodermal layer during midline fusion of the first and second brachial arches. Generally, dermoid cysts are found in the ovaries and testes. Majority of head and neck dermoids, however, are located in the area of the lateral third portion of the eyebrow, which is the most common site in the pediatric population. Floor of mouth dermoid cysts are rare, comprising only 1.6% of all body dermoids. Although congenital in nature, most cases become clinically apparent around the second or third decade of life. They most commonly develop in the midline of the floor of mouth and present as slow and progressive growing lesions, which have the potential to cause dysphagia and airway compromise. Treatment for a dermoid cyst is surgical excision. Diagnosis is confirmed via histopathologic examination, which shows the cyst lined by orthokeratinized stratified squamous epithelium with a prominent granular cell layer. Abundant keratin is often found within the cyst lumen. The purpose of this paper is to report two pediatric cases of floor of mouth dermoid cysts, and to briefly review the most recent literature pertaining to this very uncommon pediatric lesion.

Case Report 1
A 14 year old male with an unremarkable past medical history presented with a floor of mouth swelling. He began to notice the lesion about three months prior, which led his parents to seek treatment. Upon further questioning, the patient denied any fever, pain, or chills. Physical examination showed a well-nourished male with no facial asymmetry. His neck was soft and non-tender without any palpable masses or cervical lymphadenopathy. Intraorally a lesion measuring approximately 1 cm was located to the center-right of the floor of mouth. The mass was firm upon palpation without any tenderness. Bilateral Wharton’s ducts were patent with a noticeable decrease salivary flow to the right duct. There were no symptoms of dysphagia, difficulty with speech, or airway compromise. A computed tomography (CT) scan of the neck with intravenous contrast was obtained depicting a cystic mass within the right sublingual space, measuring 4.0 x 2.2 x 3.8 cm. (Figure 1a & b) The lesion was contained between the mylohyoid and genioglossus muscles. The radiologic and clinical impression was thought to be a right sublingual ranula.

A trans oral surgical approach was chosen to excise the suspected ranula. As the dissection proceeded, it was noted that a separate smooth surfaced cystic lesion was appreciated medially to the sublingual gland. A small area of the lesion was lanced during excision and expressed copious amounts of thick, yellow, keratin-like debris. (Figure 1c) The right sublingual gland and cystic lesion were removed without complication and the incision site was primarily closed. (Figure 1d) Wharton’s duct and the lingual nerve were both preserved. The patient was admitted for
overnight observation and was discharged home the next day with an uneventful stay. At the one week postoperative visit, the incision site was healing well and the patient was able to return to his normal diet without difficulty. Histologic examination established a diagnosis of a sublingual dermoid cyst. Sebaceous glands and hair follicles were present within the wall of the epithelial lined cyst as well as an abundance of keratinous debris which was located in the lumen. Figure 1e

Figure 1: The axial (a) and sagittal (b) computed tomography sections at the level of the oral floor, showing a well-defined, hypodense unilocular collection with normal adjacent structures. Cystic lesion with yellow keratin-like debris (c) and right sublingual gland (d) seen. The Hematoxylin and eosin stain (e) shows the cystic lumen lined by stratified squamous epithelium with sebaceous glands (S) and hair follicles (H) within the cyst wall.

Case Report 2
A 16 year old healthy African-American female was referred for evaluation of a large floor of mouth swelling extending into the submental region (Figure 2a). The patient stated the mass had been present “for as long as one can remember and it has gotten bigger as I have grown”. She occasionally had difficulty swallowing but denied any airway compromise. Extra oral examination showed a soft, doughy, non-pulsatile, submandibular and sub-mental swelling measuring 6cm x 5cm in greatest dimension. Intra orally, there was a bilateral compressible, non-tender, swelling of the entire floor of the mouth. The mucosa was pink and moist with no evidence of ulceration (Figure 2b). CT scan showed an 8 cm bilobed cystic mass in the midline involving the floor of mouth displacing the tongue superiorly and mylohyoid muscle inferiorly (Figure 2c). Initial radiological interpretation could not rule out a ranula. Our clinical diagnosis suspected this lesion as being congenital in nature due to the early presence in the oral cavity since childhood. A MRI was obtained which showed a hyper intense T2 weighted image suggestive of a congenital cystic lesion (Figure 2d).

The patient was taken to the operating room for surgical excision. Due to the large size of the mass, a transcutaneous approach was chosen to allow for adequate exposure (Figure 2e). An enlarged submental lymph node was also detected upon dissection and was sent for frozen section to rule out malignancy. The cyst was kept intact and excised, measuring 7.5x6x3.5 cm in size (Figure 2f). The incision was then closed in layers and a drain was placed for prevention of any fluid collection. Histopathology confirmed the lesion as a dermoid cyst. The patient was seen one week postoperatively for follow up. Her tongue had a full range of motion without any neurological
deficits. She was able to return to her everyday habits and eat and swallow without difficulty.

Figure 2: A clinical photograph showing a “double-chin” appearance (a), with an intraoral midline submental swelling in the floor of mouth (b). The sagittal CT image of the radiolucent unilocular mass (c) and Sagittal MRI: T2W1 hyper intense, well circumscribed mass superior to the mylohyoid muscle and displacing the geniohyoid superiorly (d). Intraoperative view of the specimen delivered via an extra oral approach (e) with resected sublingual mass measuring 7.5cm x 6cm x 3.5cm (f).

Discussion
Although head and neck dermoid cysts are common in the pediatric population, the cysts are usually limited to the periorbital region, appearing along the naso-optic groove between the maxillary and mandibular processes during embryonal closure. Less common areas of occurrence are noted to develop over the dorsum of the nose and between the mid-ventral and mid-dorsal fusion in the suprasternal, thyroidal, and suboccipital regions. In 1976, Pollard et al. reported a study of 231 pediatric patients that underwent surgical removal of dermoid cysts. Of these lesions, 84% (194 cases), were located in the head and neck, with 43% confined to the periorbital region. The remaining 57% were scattered throughout the scalp, ears, and neck. A recent Mayo clinic study of pediatric head and neck dermoid cysts, recorded an even higher number of 61% occurring in the peri orbital region. There were no reported cases of intraoral dermoid cysts over their 22 year study. The most unusual and least common area for dermoid cysts to develop, as in our cases, is the floor of mouth. The cyst develops as the first and second brachial arches fuse during the third and fourth week of embryologic gestation.

Dermoid cysts are described as “dysontogenetic,” referring to the disturbance in its embryologic development. Over the last century, we have seen multiple terminologies...
used when referring to the dermoid cyst. Traditionally, dermoid cysts have been classified into three variants based upon their unique histopathology: 1) Epidermoid – epithelial-lined cyst without adnexal structures. 2) Dermoid – epithelial-lined cyst with dermal appendages, such as hair follicles, sebaceous glands, or sweat glands, in the underlying connective tissue. 3) Teratoid - epithelial-lined cysts containing epithelial and non-epithelial elements, such as bone, muscle, respiratory and gastrointestinal tissues.5,6

In 1973, Spouge reclassified epidermoid, dermoid, and teratoid cysts into simple, compound, and complex respectively.7 Recently, in an attempt to resolve the ambiguity in the literature, Gordon et al.8 suggested in the utilization of a more specific term, congenital germ line fusion cyst, which is inclusive of all three histologic variants.8 Floor of mouth dermoid cysts are generally diagnosed in young adults in their second and third decades of life. There have been reports of occurrence as early as infancy. Gibson and Fenton reported a total of 27 infants with congenital intraoral dermoid cysts from 1949-1981.7 Floor of mouth dermoid cysts are mainly located in the midline spaces (sublingual 52%, submental 26%).9 They initially present as a progressive, painless, doughy mass in the floor of mouth or base of tongue. When the cyst occurs below the mylohyoid muscle, the patient can present with a submental mass resembling a “double-chin”. Dermoid cysts are usually asymptomatic but have the potential to cause dysphagia, dysphonia, and airway compromise as they grow. The cyst may become infected and secondary fistulas can develop.7 The cystic mass can vary in size from a few millimeters up to 12 cm in diameter and are considered benign lesions.5 There have been reports of malignant transformation to squamous cell carcinoma documented only in the teratoid variant; however, this is extremely rare.

Differential diagnosis should include a ranula, thyroglossal duct cyst, lymphatic malformations (cystic hygroma) and benign or malignant salivary gland neoplasms. Ranulas are common in children and are referred to as “pseudocysts,” because they lack an epithelial lining. They arise from the traumatic rupture of the sublingual salivary gland duct resulting in spillage of mucin into the surrounding tissues. Clinically, ranula appear as a blue, dome-shaped, fluctuant swelling in the floor of mouth.2 Occasionally, ranula can dissect through clefts or defects in the mylohyoid muscle and present as a cervical neck swelling which can mimic the presentation of a dermoid cyst.10 This is referred to as a cervical or plunging ranula. They are treated by excision or marsupialization.

Cystic hygroma are usually seen in infants and present as a fluctuant mass located in the sublingual space and anterior third of the tongue. They represent a sequestration of embryonic lymphatic tissue which fails to communicate with the venous system during development. This developmental abnormality is present at birth with 90% present by the age of 2 years.11 Diagnostic imaging used for any floor of mouth lesion would include a CT, ultrasonography (US), and magnetic resonance imaging (MRI). Because MRI and US have superior soft tissue resolution compared with CT, they are able to better depict the internal structure of a mass lesion.12 For Case 2, the initial CT could not rule out other cystic lesions, such as a ranula. A MRI was obtained of the neck and showed a high-intensity mass on the T2-weighted images, consistent with a dermoid cyst. For pediatric patients, US is recommended as the first choice imaging technique due to the reliability, cost efficiency, and elimination of radiation.13 Longo et al.13 demonstrated the successful use of US in 5/16 floor of mouth midline cysts without the need for further imaging prior to excision.13

Aspiration of dermoid cysts for diagnostic value is a controversial topic. Eppeley14 stated that aspiration of the cystic content is usually not helpful and in doing so can lead to infection that complicates surgical excision. There have been multiple case reports of pediatric patients where aspiration was used as temporary relief if postponement of surgery was necessary. Surgical removal is the treatment of choice for dermoid cysts. The surgical approach is dictated by the location and size relative to the geniohyoid and mylohyoid muscles. Makos et al., reported that out of 120 floor of mouth dermoid cyst case reviews, 58% were removed intra orally, 31% extra orally and 11% via a combination of intra and extra oral approaches.15 Prognosis is very good, and the recurrence rate is virtually non-
existent. In the neonate, early excision is important to establish normal nursing, speech development, and to possibly prevent osseous growth abnormalities.5

Conclusion

Dermoid cysts of the oral cavity are uncommon, representing 0.01% of all oral lesions and often pose a challenge with the initial differential diagnosis in the pediatric population.16 Our initial differential in both cases were proven wrong, which show why it is imperative to educate the medical community. Literature has shown an overwhelming success in treating these lesions via surgical excision with a low rate of morbidity or mortality. Ultimately, a variety of imaging tools allows us to determine the anatomical makeup and location of the lesion and histopathology will confirm the final diagnosis.

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