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Case Challenge
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Smooth-surfaced Nodule on the Ventral Tongue

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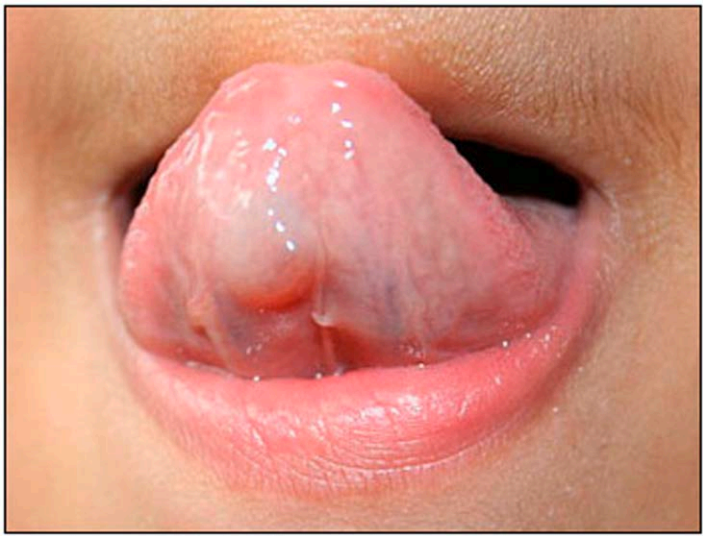

The following Case Challenge is provided in conjunction with the American Academy of Oral and Maxillofacial Pathology.

Case Summary

A four-year-old girl presented to the Dental Clinic at Seattle Children's Hospital in Seattle, Washington, USA with a single, smooth-surfaced, well-demarcated, grayish nodule on the right side of the ventral surface of the tongue. It was described initially as small (4 X 4 mm) and fluctuant, but over a six to eight week period it progressed to a 1 X 1 cm firm mass. The overlying mucosa was distended and smooth with no evidence of ulceration.

After you have finished reviewing the available diagnostic information, make the diagnosis.

Diagnostic Information

Medical History

The patient's medical history was not significant for any diseases or allergies. There was no history of trauma to the area.

Oral Findings

Upon examination, a well-demarcated, dome-shaped, smooth-surfaced, exophytic, pink to focally gray nodule was identified on the ventral surface of the tongue (Figure 1). The lesion was firm to palpation but was not painful, and there was no evidence of calcification associated with the lesion.

Excisional Biopsy Findings

Under general anesthesia, the lesion was completely excised and was found to be a fluid filled cyst-like structure. The fluid was viscous, honey-colored, and was expressed from the central cystic structure. Histologic examination revealed a cyst-like architecture lined by granulation tissue rather than epithelium (Figure 2) and filled with mucoid material, sheets of foamy macrophages, and a few neutrophils (Figure 3). This structure was focally surrounded by skeletal muscle bundles (Figure 2) and minor salivary gland lobules. The latter showed evidence of focal fibrosis and chronic inflammation (Figure 4).

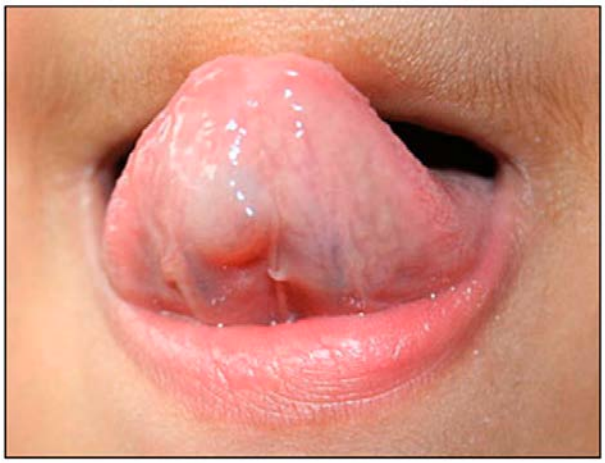

Figure 1. Clinical photograph taken at the first visit. Note the dome-shaped smooth surfaced exophytic grayish nodule on the ventral surface of tongue.

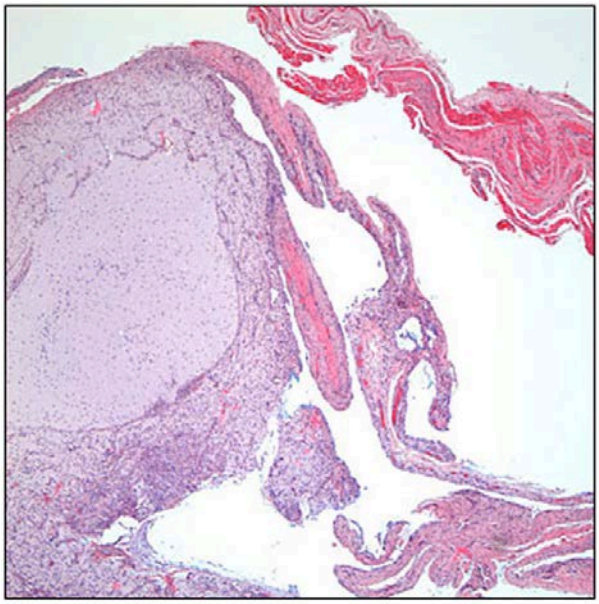

Figure 2. Low power (x100) histology shows a cyst-like structure lined by granulation tissue and filled with mucoid material and foamy macrophages. The lesion was focally surrounded by skeletal muscle bundles.

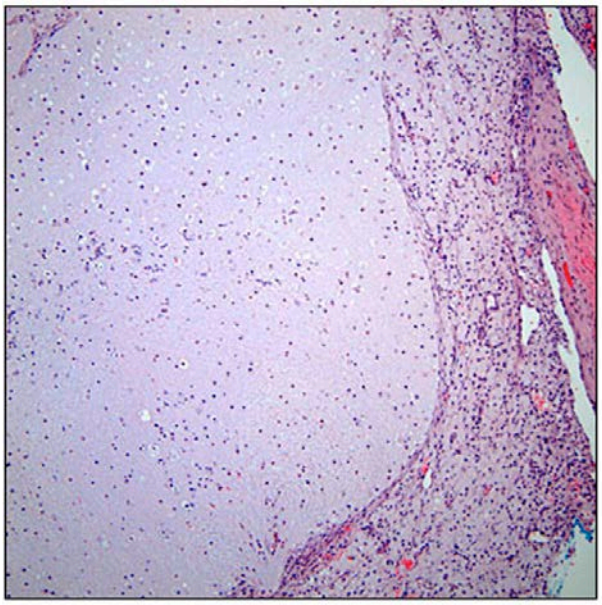

Figure 3. High power (x200) histology shows a closer view of the lumen of the cyst-like structure filled with mucoid material and foamy macrophages.

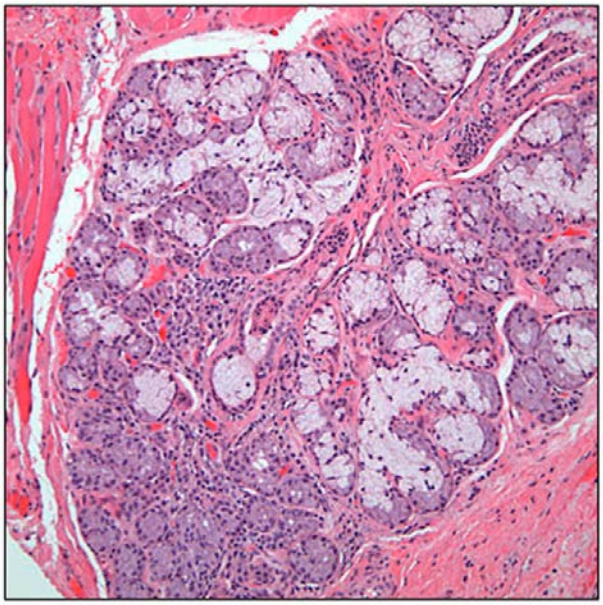

Figure 4. High power (x200) histology shows minor salivary gland lobules (Blandin-Nuhn glands) exhibiting focal stromal fibrosis and chronic inflammation.

Can you make the diagnosis?

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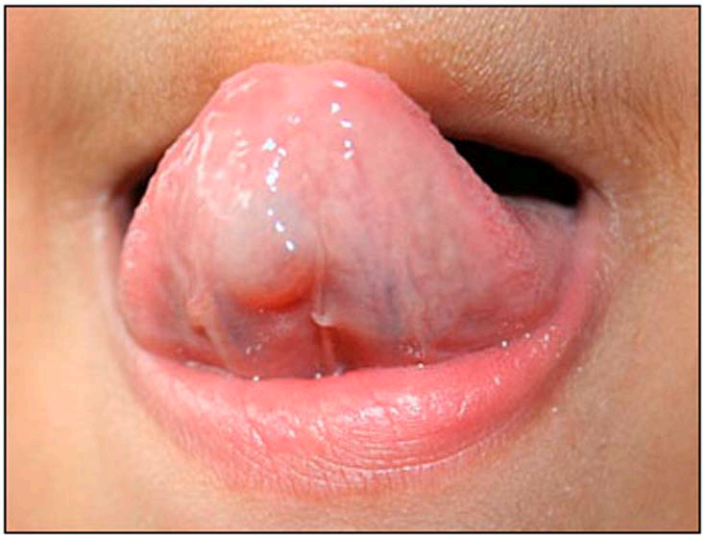

Select the Correct Diagnosis

A. Lymphangioma
B. Mucocele
C. Mucoepidermoid Carcinoma
D. Granular Cell Tumor

Lymphangioma

Choice A. Sorry, this is not the correct diagnosis.

Lymphangiomas are benign congenital vascular developmental anomalies of the lymphatic system that commonly occur in the head and neck area, especially on the dorsal tongue. [1,2] Lymphangiomas are best classified as being either superficial or deep. Deep lymphangiomas are divided into cavernous and cystic types. The cavernous type is more common in the oral cavity. Cystic hygroma is a term used to describe a particularly large cavernous lymphangioma.[1,2] The superficial type is also known as lymphangioma simplex or lymphangioma circumscriptum. The head and neck area is the most common location for lymphangiomas, followed by the extremities and buttocks. They are usually diagnosed in infancy, with 60% diagnosed at birth and 80-90% developing within the first two years of life.[1,2] In the oral cavity lymphangiomas most commonly affect the anterior two thirds of the tongue.[1,2] The superficial (circumscriptum type) tongue lymphangiomas have a “cobblestone” appearance produced by fluid filled superficially located, dilated lymphatic vessels. They are usually the same color as the surrounding mucosa; but occasionally, a focal dark red color is described. This is believed to be due to trauma-induced bleeding into the dilated lymphatic vessels. Deep lymphangiomas are more diffuse and soft in consistency.[1,3] Lymphangioma may also occur on the lips, resembling angioedema. They are usually asymptomatic but can be painful on palpation, and they may drain clear fluid if traumatized. Depending on the histologic subtype, lymphangiomas demonstrate dilated lymphatic vessels that are either present directly beneath the surface epithelium or extend into the underlying deep connective tissue. Conventional scalpel surgery or CO2 laser ablation are the preferred treatments for this lesion.[3,4] Sclerosing agents such as OK-432 and steroids have been used with some success.[4] Local recurrence is common, especially with the deep and hard-to-reach lesions. The superficial lesions have better success and fewer incidences of recurrence.

Given the location of the lesion and the age of the patient, lymphangioma should be seriously considered. However, the histology is not supportive of the diagnosis.

Please re-evaluate the information about this case.

Mucocele

Choice B. Congratulations! You are correct.

Mucoceles are common reactive lesions associated with minor salivary gland tissue in the oral cavity. They are not common, however, in the ventral tongue area. Mucoceles typically present as an exophytic, fluid filled, fluctuant nodule most commonly occurring as small lesions in the lower lip and the floor of the mouth.[5,6] The vast majority of mucoceles are found in the lower lip. The anterior and ventral tongue are affected much less frequently but are cited as the next most frequent location. Mucoceles rarely occur in the upper lip.

The term ranula is used for a very large mucocele occuring in the floor of the mouth. Histologically, they show an area of mucous extravasation into the connective tissue with an associated inflammatory response. Ranulas constitute 6% of all mucoceles.[5-7] The majority of ranulas are found above the mylohyoid muscle, but occasionally lesions penetrate the muscle layers and may extend into the tissues of the neck. These lesions are referred to as plunging or cervical ranulas.[7,8] Ranulas can be of either minor or major salivary gland origin. Mucoceles show a wide age distribution ranging from infancy to the ninth decade of life[6,7] but are most commonly reported in children and adolescents with an equal sex distribution.

The etiology of the mucocele is usually sharp trauma, such as a lip bite. The sharp trauma can also be secondary to surgery. This is especially true with mucoceles in the tip of the tongue. The trauma results in severing of the salivary gland duct and release of mucous into the extracellular tissue.[5-7] However, this patient did not have a history of a sharp bite, surgery, or any trauma to the area.

Clinically, about 20% of mucoceles present as superficial swellings with a light blue color. The majority present as deep swellings and appear pink in color, similar to the surrounding mucosa.[9] This patient's clinical presentation fits mostly into the deep mucocele category (Figure 1) except in one area where the color is slightly gray, indicating a focal superficial component.

Mucoceles may fluctuate in size, usually being described by the patient as "going up and down." This patient's mucocele started as a small fluctuant swelling and grew to a large firm nodule. When they are small and superficial, mucoceles can also mimic vesiculobullous-type diseases because they look like vesicles.[9] This is especially true when the mucoceles present as multifocal lesions. While this is a rare presentation, it is a reported pattern of occurrence.

Histologically, the mucocele consists of a cyst-like structure lined by granulation tissue and filled with mucoid material, foamy macrophages, and at times small clusters of neutrophils (Figures 2 and 3).

Surgical excision of the lesional tissue with the associated minor salivary glands is the preferred treatment for deep mucoceles. For superficial mucoceles, no treatment is recommended since they can heal spontaneously within two to three weeks.[7-9] The lesion in this patient was completely excised. The surgical site healed within two weeks. A small and superficial mucocele subsequently appeared in the third week. It lasted two weeks and healed without surgery. The clinical presentation five months after surgery (Figure 5) shows a completely healed area with very little scarring.

Simple (non-plunging) ranulas are best treated by marsupialization.[7,8] Plunging ranulas require complete excision via an extraoral approach. The technical difficulties associated with the complete removal of some ranulas and mucoceles result in a relatively high recurrence rate.

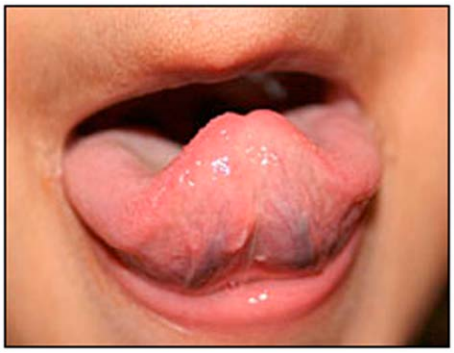

Figure 5. Clinical photograph was taken five months after surgery. Note the area is completely healed.

A lesion that is usually clinically indistinguishable from a mucocele is the salivary duct cyst. This lesion is also sometimes referred to as a mucous retention cyst. Salivary duct cysts are less common than mucoceles and usually occur on the buccal mucosa, palate, and floor of the mouth. Thus, it should be included in the differential diagnosis for lesions that clinically resemble a mucocele when the lesion is found in a location unusual for a mucocele. In particular, lesions in the palate and upper lip diagnosed clinically as mucoceles often prove to be salivary duct cysts. The salivary duct cyst, as the name would suggest, develops as a result of a duct blockage. The blockage can be caused by trauma, fibrosis, sialolith, or pressure from an overlying tumor.[6-8] In contrast to a mucocele it presents histologically as a true cystic structure with an epithelial lining, connective tissue wall, and a lumen filled with mucoid material.

Mucoepidermoid Carcinoma

Choice C. Sorry, this is not the correct diagnosis.

Mucoepidermoid carcinoma (MEC) is the most common malignant salivary gland neoplasm. It accounts for 10% of all salivary gland neoplasms.[10-12] While the majority of MECs occur in the parotid gland, approximately 18% occur in the oral cavity. The palate is the most common intraoral site followed by the tongue, buccal mucosa, lips, and retromolar pad areas.[10] Two thirds of intraoral MECs present as smooth-surfaced swellings, gray-blue or pink in color in the posterior-lateral hard palate or anterior-lateral soft palate. Less than 20% of MECs intraorally occur in the tongue.[10] It is reported in a wide age range with a predilection for young people.[10,13]

One study from the Armed Forces Institute of Pathology (AFIP) demonstrates that 44% of MECs occur in patients under the age of 20.[10] The youngest patient was nine months old.

Three histologic grades within this neoplasm are reported: low, intermediate, and high; the low-grade type is more common in the oral cavity.[10,13] Low-grade lesions are slow-growing and painless but are not encapsulated. They sometimes resemble a mucocele, especially those in the retromolar pad area.[10,11] High-grade lesions tend to be found more commonly in the parotid gland. They often present as rapidly growing, painful lesions with facial nerve paralysis and sometimes with regional lymph node metastasis.

Complete surgical removal with clean margins is the preferred treatment for the low-grade type. Radiotherapy has also been successfully used, especially when the tumor involves the surgical margins.[10-13]

Given the focal gray color and history of a fluctuant lesion, MEC should be included in the differential diagnosis. The histology, however, is not supportive of this diagnosis.

Please re-evaluate the information about this case.

Granular Cell Tumor

Choice D. Sorry, this is not the correct diagnosis.

The granular cell tumor (GCT) is a benign neoplasm of neural crest origin.[14] The tongue is the most common site of occurrence of this tumor with almost one third of cases occurring there. Lesions tend to occur more often on the lateral aspects of the dorsum of the tongue than on the ventral surface. In the oral cavity the buccal mucosa is second to the tongue in site predilection, and the lesion is more common in females than males (2:1 ratio). Although the skin is another common location, this lesion can occur in a variety of sites and is almost exclusively benign. A malignant form is described, but it is rare.

Although GCT tends to occur in adults over 30 years of age, it has also been described in children. Lesions in children have been reported between age 3 and 19, with an average age of occurrence of 14.5 in this group.[15] In children there is a 3:1 female/male sex predilection, and 50% of the lesions occur in the tongue.[14,15]

The GCT is usually asymptomatic and can be of long duration, ranging from months to years. The color of the clinical lesion is variable ranging from pink to white with a keratotic surface to yellow-orange.

Histologically, GCT is composed of strands and fascicles of large cells with distinct cell borders containing abundant granular cytoplasm. The nuclei are small, round to oval, and are eccentrically located. At times, the cells are intimately related to surrounding skeletal muscle fibers and nerve bundles. The lesion can be well demarcated and circumscribed or infiltrative.

The overlying epithelium can be normal in thickness or proliferative with psuedoepitheliomatous hyperplasia.[14,15] The pseudoepitheliomatous hyperplasia, which may be extensive in rare cases, can be mistaken for well differentiated squamous cell carcinoma, especially if the biopsy is superficial. The tumor cells are positive for S-100 protein and neuron specific enolase (NSE) indicating a neural crest origin.

Treatment includes conservative surgical excision. Recurrence is extremely rare.

While GCT is a consideration in the differential diagnosis for nearly any tongue lesion, the age, the gray color of the lesion, and the histology are arguments against the diagnosis.

Please re-evaluate the information about this case.

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Note: Bio information was provided at the time the case challenge was developed.

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