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Recurrent right sublingual ranula, concomitant with ipsilateral submandibular salivary gland aplasia

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ABSTRACT

INTRODUCTION: Oral ranula is a retention cyst that arises from the salivary gland with recurrence rate of up to 25% after complete excision of ranula and up to 2% in case of complete excision of ranula and sublingual gland.

Major salivary gland aplasia is a rare finding that is usually associated with other developmental anomalies.

PRESENTATION OF CASE: We report a 15-year-old female patient presented with recurrent intraoral cystic swelling that was documented to be sublingual ranula. CT scan revealed also the absence of right submandibular salivary gland with persistence of its Whartons duct. This combination has never been reported previously.

DISCUSSION: The combination of recurrent sublingual ranula associated with aplasia of ipsilateral submandibular salivary gland and persistence of Whartons duct has never been reported before in the literature, a finding that may provide the base for future research.

CONCLUSION: Further research may prove similar associations between oral ranula and salivary gland aplasia, which may have clinical implications on diagnostic and management plan decisions.

1. Introduction

Ranulas are mucoceles that occur in the floor of the mouth and usually involve the major salivary glands. Specifically, the ranula originates in the body of the sublingual gland, in the ducts of the sublingual gland, in the Wharton's duct of the submandibular gland or infrequently from the minor salivary glands at this location. The gender predilection of oral ranulas slightly favors females, with a male-to-female ratio of 1:1.4, while cervical ranulas have a predilection for males.[1] Ranulas usually occur in children and young adults, with the peak frequency in the second decade. The cervical variant tends to occur a little later in the third decade.[1] These lesions have not known to have malignant potential, but one report of a squamous cell carcinoma exists.[2] Congenital absence of the salivary glands is an infrequent disorder, which has been described to affect the parotid or submandibular glands associated with multiple other developmental anomalies. The management of ranula and recurrence after surgical or non-surgical management were a matter of interest in the literature. In this case, we report an incidentally detected unilateral submandibular salivary gland aplasia associated with an ipsilateral recurrent sublingual ranula. To the best of our knowledge, this is the first report of such an association.

2. Presesntation of case

A 15-year-old young girl who's not known to have any chronic medical illnesses, presented to our outpatient clinic complaining of an intraoral lesion at the right side of the mouth floor since 2 months, which had increased in size with no significant associated symptoms of pain, fever, mouth dryness or upper respiratory tract infection. No history of dental caries, or family history of such condition. On examination, there was a rounded cystic swelling (2 cm) located at the right side of the floor of the mouth, blue to translucent, soft in consistency, not-tender, without any signs of inflammation. No other intraoral lesions or cervical lymphadenopathy. Our initial diagnosis was a right sublingual ranula. The patient was operated upon using a trans-oral approach where the cystic mass was denuded from the surrounding mucosa and totally excised with primary closure of the mouth floor. Histopathological study revealed a cyst that was devoid of lining epithelium and the wall was mildly infiltrated by inflammatory cells, histiocytes and foreign body giant cell reaction with no signs of malignancy confirming the preoperative diagnosis. After three months of an uneventful postoperative period the mass had recurred, and ruptured spontaneously three to four times according to the patient.

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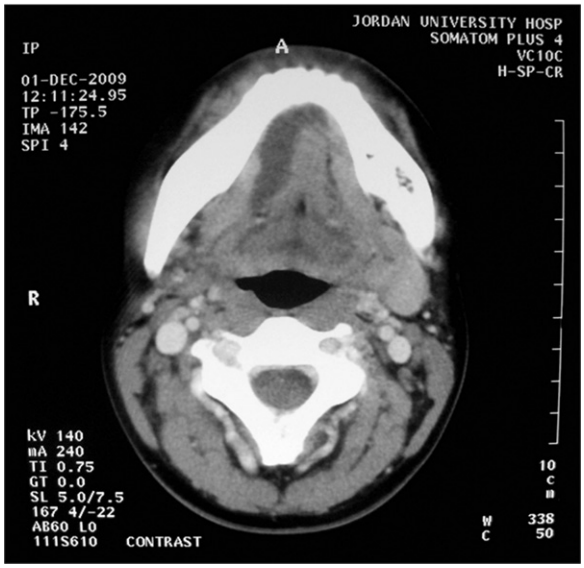

Fig. 1. Axial CT scan images with I.V. contrast showing right sublingual ranula and absence of right submandibular salivary gland, while the left submandibular gland is present with normal size.

Six months after her first surgery, the patient was readmitted with the same complaint. CT scan was performed and showed a cystic lesion at the right aspect of the mouth floor with absence of the right submandibular salivary gland. The left submandibular and both parotid glands were seen and were of normal size. There was no cervical lymph node enlargement (Fig. 1). Intraoperatively we found the cystic mass to be connected to a well defined duct which was blind-ended upon insertion of a metal probe probably representing the right submandibular salivary gland duct (Wharton's duct), confirming the CT finding of an aplastic right submandibular salivary gland. The cystic mass was denuded from the surrounding mucosa, totally excised, and the mentioned duct was marsupialized into the floor of the mouth. The right sublingual salivary gland was excised, and the lingual nerve was observed and secured (Fig. 2). Histopathologically, the cystic mass was consistent with the previous findings, and the excised sublingual salivary gland showed signs of chronic sialadenitis. There was no evidence of recurrence in 2 years follow up after second surgery.

3. Discussion

The term "ranula" is derived from the Latin word "rana" (meaning frog) and is a descriptive of the bluish color. It causes gradual enlargement of the floor of the mouth to form a painless, fluctuant, translucent, dome-shaped swelling, which is said to resemble the underbelly of a frog.[3] These lesions are divided into two types: oral ranulas and cervical or plunging ranulas. Oral ranula is a mucus retention cyst arising from the sublingual gland on the floor of the mouth as a result of ductal obstruction and fluid retention,[3] whereas cervical ranulas are associated with mucus extravasation beyond the mylohiod muscle along the fascial planes of the neck. There are two different concepts in the pathogenesis of ranula. One is a true cyst formation due to ductal obstruction with an epithelial lining, and the other is a pseudocyst formation due to ductal injury and extravasation of mucus without an epithelial lining. Ranula may also uncommonly present as a rapidly enlarging swelling following infection.[3] Oral and plunging ranulas, if large, may affect swallowing, speech, or mastication and may result in airway obstruction. The very rare thoracic ranula may compromise respiratory function and may be life threatening.[4] Aside from ranula, a number of other lesions may be encountered in the floor of the mouth or submandibular space region. These include congenital abnormalities (cystic hygromas, branchial cysts, and thyroglossal duct cysts), benign lesions (epidermoid cysts, dermoid tumors, and lipomas), malignant neoplasia, and other lesions (abscess, mucocele, and acidosis).[3] The diagnosis of ranula is largely clinical.[5] Treatment of an intraoral ranula consists of surgical excision,[6] marsupialization with and without packing[7] or currently Intracystic injection therapy with OK-432, streptococcal preparation[8] or botulinum toxin.[9] Alternatively, the ranula can be treated with the placement of a silk suture or seton into the dome of the cyst.[10] Optimal management of pediatric oral cavity ranulas may include observation for five months for spontaneous resolution.[11] Definitive treatment yielding the lowest recurrence and complication rates for all ranulas is transoral excision of the ipsilateral sublingual

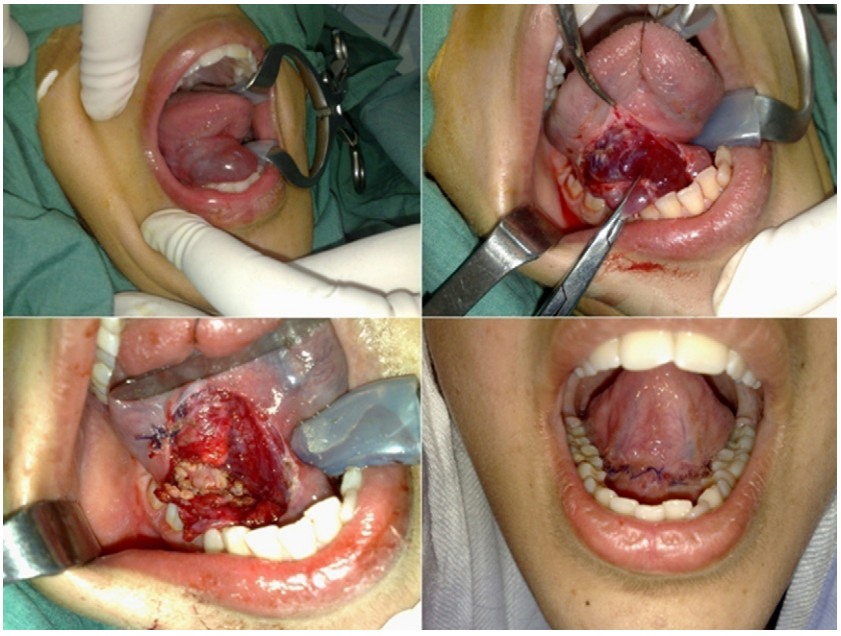

Fig. 2. Clinical photographs showing the right sublingual ranula and transoral approach for removal of ranula and excision of right sublingual salivary gland.

gland with ranula evacuation.[12] If symptoms are minimal in young age group, aspiration of the lesions and periodic follow-up for 6 months have been suggested as an alternative to surgery.[13] Laser ablation and cryosurgery, either alone or after marsupialization, have been used for some patients with oral ranula.[12] The recurrence rates of an oral ranula with various surgical treatment methods are as follow: Incision and drainage (71–100%); Ranula excision only (0–25%); Marsupialization only (61–89%); Marsupialization with packing (0–12%) (Limited studies), Complete excision of the ranula and sublingual gland (0–2%).[14]

Congenital absence of the salivary glands is an infrequent disorder which has been described to affect the parotid or submandibular glands. It can be associated with other developmental anomalies, especially in the face which can be diagnosed with a variety of imaging techniques.[15,16] Clinically, patients may be asymptomatic or may present with dryness of the mouth, difficulty in chewing and swallowing, or dental caries,[17,18] which is thought to be due to reduction of the protective effect of saliva within the oral cavity. The most common pattern seen is absence of the submandibular gland, which may be associated with hypertrophy of the contralateral submandibular gland.[15,18] Isolated unilateral major salivary gland aplasia is a rare entity with only a few cases reported in the literature to date.[18,19,21] The aplasia is likely due to arrest in organogenesis, but the exact etiology is unknown.[20] Salivary gland aplasia may be associated with first branchial arch defects in the Treacher-Collins syndrome (mandibulofacial dysostosis) or orbital abnormalities like lacrimal hypoplasia, canalicular atresia, and absence of the lacrimal puncta.[15,16,22,23] Absence of right submandibular salivary gland with existence of its Wharton's duct alone was reported,[20] But the association with ipsilateral recurrent ranula was never reported before.

4. Conclusion

We report, we believe, the first time association between recurrent sublingual ranula and aplasia of the ipsilateral submandibular gland with persistence of its Wharton's duct in otherwise healthy adolescent. This could call for analytic research in the upcoming future looking for possible similar results and the clinical implications of such an association and whether this could affect our diagnostic and management plans in such case.

Conflict of interest statement

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors' contributions

Nader M. Albsoul performed study design, writing and the surgeon who underwent the operations.

Raed N. Altaher performed study design, writning, and assistant in the operation.

Fatima O. Obeidat is the pathologist who made the histopathological studies of the case.

Azmy M. Hadidy and Shams A. Jubouri are the radiologists who reported the CT images of the case.

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