Unilateral Submandibular Gland Aplasia with Ipsilateral Sublingual Ranula - A Case Report

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Abstract

Background: Congenital aplasia of major salivary glands is a very rare entity, especially if it concerns an ipsilateral aplasia in a nonsyndromic patient. **Key points from the case:** The aim of this report is to present a case of an aplasia of the left submandibular gland, which was incidentally diagnosed during presurgical imaging for an ipsilateral sublingual ranula. Histopathological evidence of the lack of sublingual gland tissue in the excised specimen of the ranula is discussed. **Main lessons to be learned from this case report:** Unilateral submandibular aplasia has unknown etiology. Clinicians should be aware of this condition mainly to be able to differentially diagnose a hypertrophy/dysplasia of the contralateral or other major salivary glands, or when xerostomia is the main patient's symptom.

Keywords: Nonsyndromic unilateral aplasia, salivary gland aplasia, submandibular gland

Introduction

Congenital aplasia of major salivary glands is a very rare entity. Unilateral aplasias of an isolated major salivary gland are even rarer, with few case reports published.[1] The first case of a bilateral submandibular gland aplasia was reported in 1885.[2] Since then, approximately 40 cases of aplasias have been published, and approximately 13 that involve unilateral aplasias of the submandibular gland.[3]

Aplasia of more than one salivary glands is connected to other developmental anomalies, in particular lacrimo-auriculo-dento-digital (LADD) syndrome.[4] This condition is inherited, with a possible autosomal-dominant type. The mechanism which results to an isolated salivary gland aplasia is not known.

The aim of this report is to present a case of an aplasia of the left submandibular gland, which was incidentally diagnosed during presurgical imaging for an ipsilateral sublingual ranula.

Case Report

A 24-year-old male patient visited the outpatient clinic of the University Department of Oral and Maxillofacial Surgery, with a relapsing sublingual swelling on the left side of the floor of the mouth. In his medical history, The patient reports a laser excision of the lingual frenulum 2 years ago and after that, the presence of a swelling below the tongue, at the left side. A previous excision of the swelling was also reported, with a subsequent recurrence 3 months ago and since then the lesion showed exacerbations and recessions. Clinically, a ranula of the left sublingual gland was the probable diagnosis for his condition. Wharton's duct seemed clinically normal, and during the bimanual examination of the floor of the mouth, no abnormal clinical sign was recorded. A computed tomography (CT) was performed to exclude a submerged ranula [Figure 1]. The CT did not reveal a ranula, possibly due to recession of the inflammation, but did show an absence of the ipsilateral submandibular gland while the contralateral submandibular gland was of normal size. A magnetic resonance imaging (MRI) was then performed for further assessment. The MRI confirmed the absence of the ipsilateral submandibular gland and revealed a small cystic lesion in the

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DOI: 10.4103/ams.ams_63_20

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Received: 04-03-2020 **Revised:** 01-04-2020
Accepted: 17-04-2020 **Published:** 23-12-2020

How to cite this article: Tatsis D, Mantevas A, Kilmpasani M, Karafoulidou I, Venetis G. Unilateral submandibular gland aplasia with ipsilateral sublingual ranula - A case report. Ann Maxillofac Surg 2020;10:543-6.

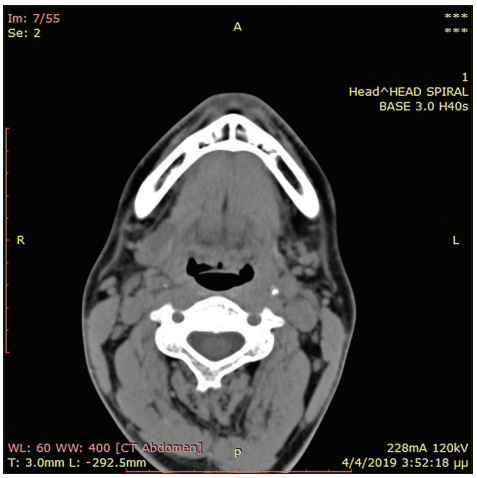

Figure 1: Computed tomography, axial plane, absence of the left submandibular gland

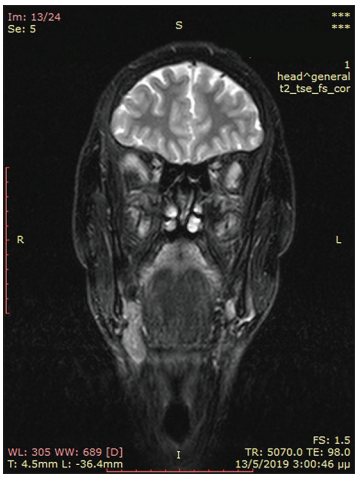

Figure 2: Magnetic resonance imaging, coronal plane, absence of the left submandibular gland without the presence of a submerging ranula

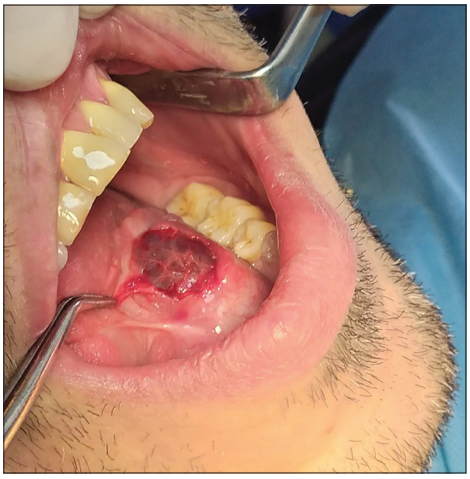

Figure 3: Intraoperative photograph of the dissection of the mucosa revealing the ranula

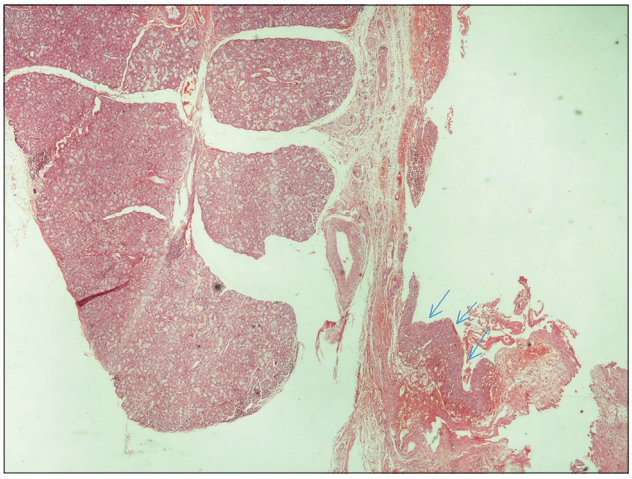

Figure 4: Sublingual salivary gland with adjacent pseudocyst wall (arrows), consisting of granulomatous inflammatory connective tissue (H and E stain, ×20)

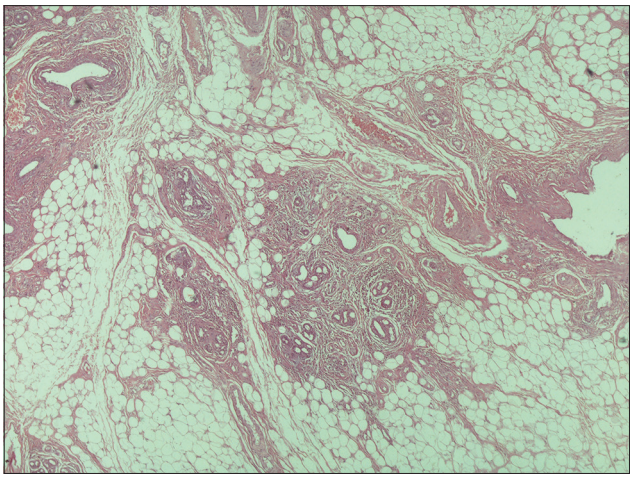

Figure 5: Sublingual salivary gland with evident atrophy, elimination of acini, and extensive replacement of adipose tissue (H and E stain, ×40)

left sublingual gland [Figure 2]. Since the patient continued to have the symptoms of the left side of the floor of the mouth, he underwent under general anesthesia a sublingual gland excision including Wharton's duct. A ranula formation was discovered in the floor of the mouth and was excised [Figure 3]. Informed consent of the patient was provided before surgery.

The histopathology report of the specimen revealed a sublingual gland sized 3 cm × 2 cm × 1 cm with an adjoining cystic formation. The gland has the regions of chronic inflammatory infiltrations and duct fibrosis and the cystic formation comprises of inflammatory connective tissue. This report is compatible with sublingual ranula with chronic inflammation of the sublingual gland [Figures 4 and 5].

Discussion

The embryological derivation of salivary glands is from the buccal epithelium of the ectoderm of the first branchial cleft. The parotid gland starts forming at the fourth embryonic week from the ectoderm, whereas the submandibular and sublingual glands start developing later during the 6th–9th week, but finish

their formation before the parotid. The salivary buds extend into the adjacent mesoderm and develop the gland and its duct in the course of its distal movement. Conventionally, aplasia or agenesis of a salivary gland is associated with other congenital malformations of ectodermal origin, such as cleft lip and palate, LADD syndrome, Treacher-Collins, or hemifacial microsomia. Another rare genetic entity, aplasia of lacrimal and salivary glands, may cause aplasia of all salivary glands. The exact mechanism which results to this gland malformation is not known.[1] Our patient did not mention any relative with a known similar aplasia.

The case reports published concerning the unilateral aplasias of submandibular glands are presented in Table 1. There is no sex predominance, since seven female patients and six male patients have been recorded, including our case. The mean age of the diagnosis is 35.5 years, ranging from 12 to 62 years old. Since this is a congenital condition, the age of diagnosis does not coincide with the age of onset.

If the aplasia occurs to more than one gland, symptoms deriving from decreased saliva excretion may be the main complaint of the patient.[7] Our patient did not have any symptoms of xerostomia or poor dental hygiene, as far as the aplasia of the submandibular gland is concerned. Patients with aplasia of only one major salivary gland are usually asymptomatic. Although without sequences on saliva production, some patients develop a hypertrophy of the contralateral gland that occasionally manifests as a mass, which can be mistaken for malignant pathology of the contralateral area.[13] Hypertrophy of the contralateral submandibular gland may mimic cancer metastasis, as Shipchandler and Lorenz report.[8] In thin patients, submandibular gland aplasia may produce clinically a depression of the submandibular region.[15]

A similar case by Srinivasan *et al*.[16] reports a unilateral submandibular aplasia with an ipsilateral sublingual hypertrophy, initially presenting as a mass of the floor of the mouth, and a normal-sized contralateral submandibular gland. In a similar case, a 1-year follow-up period of an ipsilateral hypertrophy of a sublingual gland with submandibular aplasia was also reported, stating a stable size of the gland as assessed by ultrasound sonography.[17] In our patient, ranula of the ipsilateral sublingual gland could be attributed to dysplasia of the ductal element of the gland. Correlation to previous lingual frenulum 2 years ago may be simply coincidental.

Bimanual examination of the floor of the mouth should typically demonstrate the absence of a submandibular gland. A definitive diagnosis of gland aplasia requires a thorough imaging technique, such as ultrasound, CT, or MRI.

Cases of early atrophy of the submandibular glands have been reported, proposing that an obstruction in an early phase of formation of a salivary gland can induce atrophy and subsequent secondary aplasia, leading the deposition of fatty tissue in the area where the gland should be normally cited.[7] Literature shows a quite frequent hypertrophy and/or dysplasia of another major salivary gland as possibly happens in our case of submandibular aplasia synchronous to sublingual dysplasia.

In conclusion, unilateral submandibular aplasia is a rare entity with a not-known etiology. It is usually diagnosed incidentally during the imaging of the visceral cranium for other reasons, and seldom can have symptoms involving xerostomia. Possibly, its true incidence in the population is higher, due to its asymptomatic nature. Clinicians should be aware of this condition mainly to be able to differentially diagnose a hypertrophy/dysplasia of the contralateral or other major salivary glands.

Table 1: Summary of the unilateral submandibular glands reported to the literature

Report	Year	Age	Sex
Bruno[3]	1894	Unknown	
Abdel-Dayem[5]	1978	62	Female
Kubo *et al*.[6]	1990	34	Male
Yılmaz *et al*.[3]	2002	32	Female
Shipchandler and Lorenz[8]	2008	60	Male
Gallego *et al*.[9]	2009	35	Female
Gupta *et al*.[10]	2009	35	Female
Yilmaz *et al*.[11]	2010	41	Female
García-Consuegra *et al*.[7]	2010	39	Male
Damar *et al*.[12]	2013	55	Female
Bhoil *et al*.[13]	2016	20	Female
Kandemirli[14]	2019	12	Male
		13	Male
Present case		24	Male

Financial support and sponsorship

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflicts of interest

There are no conflicts of interest.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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