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A swelling of the floor of the mouth

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CASE PRESENTATION

A 69-year-old man was referred to the department of Oral and Maxillofacial Surgery by an ear, nose, and throat surgeon for diagnosis and management of a painless "lump" on the floor of the mouth, which extended to the submandibular region. The intraoral lesion had been noticed 3 months before consultation. According to the patient, the growth had recently enlarged, interfering with his mastication, but caused no difficulty in swallowing or speech. No changes of size or development of intraoral swelling during meal times were reported. The patient was in apparent good health and his medical history was noncontributory.

Clinical examination showed a 3 × 2 cm swelling of the left submandibular area (Fig. 1). It was covered by skin that was normal in both texture and color. Intraorally, elevation of the floor of the mouth was noted, which was covered by normal mucosa (Fig. 2). On palpation, the nontender mass was soft to rubbery in consistency. Although tongue movement was marginally restricted by the physical presence of the swelling, the mass per se was freely movable anteroposteriorly and medially in relation to the tongue and the bone of the mandible. The patient was totally edentulous and wearing complete acrylic dentures. Normal saliva could be expressed from the left submandibular salivary gland and there was no sign of hypoesthesia or paresthesia of the area corresponding to the ipsilateral lingual nerve.

A computerized tomography scan (CT) with enhancement, which had been carried out before consultation, confirmed the existence of a well demarcated dense lesion lying between the mylohyoid and genioglossus muscles (Fig. 3). The lesion was solid, without relation to the mandibular bone. Intravenous contrast medium enhanced the image of the lesion. The remainder of the head and neck scan did not reveal any other pathology or lymph node enlargement. Although it was thought that magnetic resonance imaging could give more precise and accurate information for the diagnosis and management of the lesion, the patient refused any further imaging studies.

DIFFERENTIAL DIAGNOSIS

The floor of the mouth and submandibular area may be affected by numerous pathologic conditions, which can be broadly classified as developmental, inflammatory-obstructive, or neoplastic in origin. Lesions in this area may be present for a prolonged period of time before the patient seeks medical advice, usually as a result of interference with swallowing or speech.

The floor of the mouth is the most common intraoral location for developmental lesions of the oral soft tis-

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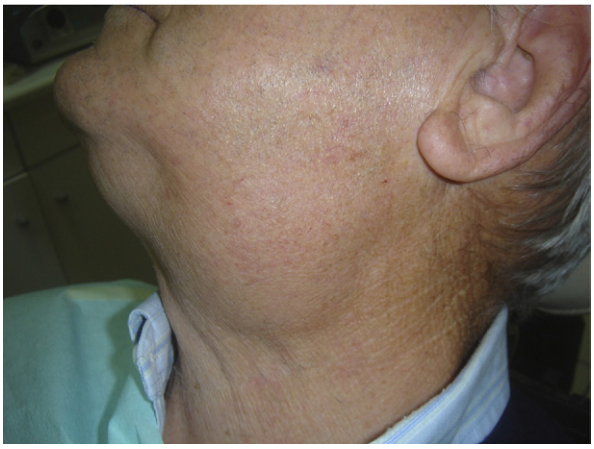

Fig. 1. Extraoral examination revealed a left submandibular swelling with a diameter of 3 cm. The soft to rubbery swelling was nontender and not firmly attached to overlying skin, the lower jaw, or the tongue.

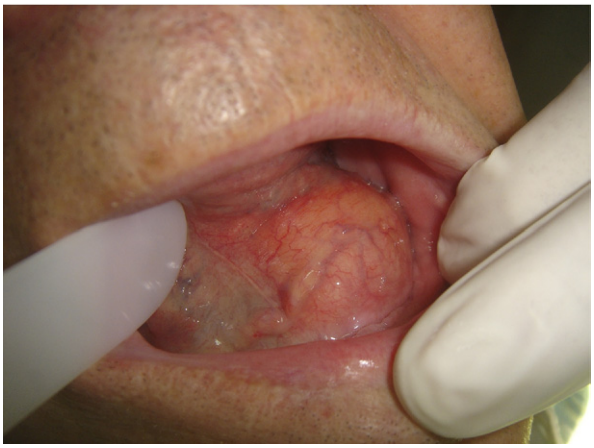

Fig. 2. Intraoral photograph showing ipsilateral elevation of the floor of the mouth. The swelling was covered by normal mucosa and was freely movable anteroposteriorly and medially.

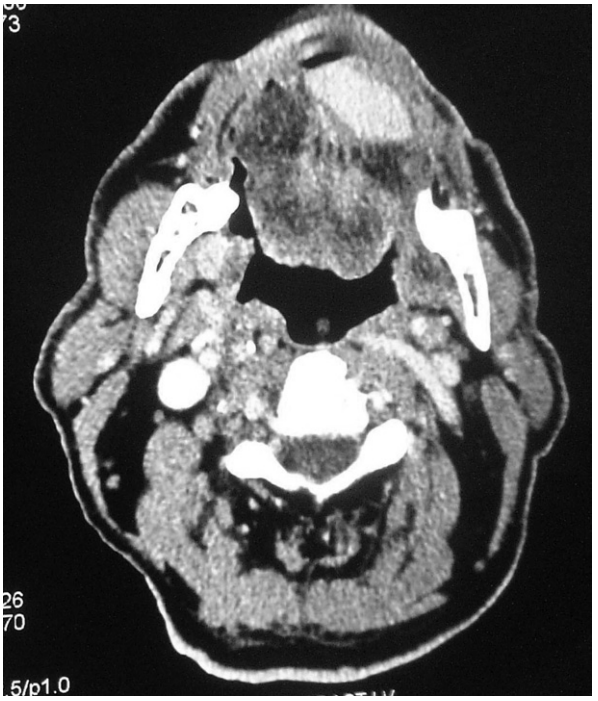

Fig. 3. Transversal computerized tomography scan with contrast enhancement depicting a well delineated dense lesion of the submandibular triangle.

sues, particularly dermoid cysts, branchial cleft cysts, heterotopic gastrointestinal cysts, and thyroglossal duct cysts.[1-3] Dermoid cysts are considered to be a variation of teratomas and are thought to arise from entrapment of epithelial remnants during closure of the branchial arches or as a result of trauma.[4,5] Almost one-fifth of the dermoid cysts that occur in the head and neck area are located on the floor of the mouth,[1,4] where they may cause tongue elevation, submental protrusion, or both.[4,5] They are predominantly seen in young persons, presenting as soft to rubbery swellings in the midline or laterally.[1] Branchial cleft cysts are developmental anomalies which arise from incomplete closure of branchial arches.[1,2,4] They usually appear in relatively young patients as fluctuant swellings located anteriorly to the sternocleidomastoid muscle.[1,6,7] Heterotopic gastrointestinal cysts are choristomas of head and neck and mostly affect the sublingual area or the floor of the mouth. They appear to have a male preponderance and are usually lined with gastric mucosa.[3] Thyroglossal duct cysts arise from remnants of the embryonic thyroid.[1,2,4] They typically present in the midline in close contact to the hyoid bone, often producing a characteristic movement during swallowing,[3] which was not seen in the present patient. In most cases, a fluctuant or soft swelling is evident in the neck region, although some lesions may appear intraorally[1,2] and cause dysphagia.[4] Half of the patients are younger than 20 years at the time of diagnosis.[1] Developmental lesions present a relatively silent course before causing any symptoms, as with the present patient. However, this patient's age made the probability of such an entity less likely. Additionally, the CT findings were suggestive of a solid rather than a cystic lesion.

Congenital lesions, such as vascular malformation or lymphangioma (cystic hygroma), are generally included in the differential diagnosis of neck masses extending to the floor of the mouth. Both lesions are most commonly seen in childhood, with ~90% of lymphangiomas of the head and neck diagnosed by the age of 2 years.[5] They demonstrate a variety of clinical

features, ranging from small varicosities to large bluish or red masses that may reach large dimensions and cause disfigurement.[1,2] Nevertheless, such lesions are not likely to initially present in an older patient and were not compatible with the CT images of our case.

Infections and inflammatory lesions of perioral tissues may also present as a swelling of the submandibular area extending to the floor of the mouth.[1,2] Acute infections, such as Ludwig angina, cellulitis, submandibular, or sublingual space infections, which may derive from an odontogenic focus of infection, osteomyelitis, or trauma, were not considered in the differential diagnosis, because of the chronic nature of the condition. In addition, fever, malaise, and pain would normally accompany the clinical presentation of an acute infection, and tenderness to palpation and fixed or fluctuant overlying skin would be apparent in the clinical examination. Another entity which should be considered is lymphadenitis of different causes. Submandibular lymph nodes may commonly be affected by cat-scratch disease, tuberculosis (scrofula), or actinomycosis.[1,8] The present patient's noncontributory past medical history, the CT findings, and the absence of any skin sinuses, precluded these entities from our clinical differential diagnosis. Additionally, lymphadenitis typically does not produce this degree of intraoral swelling.

Salivary gland lesions are commonly seen intraorally and in the submandibular triangle.[2] The ranula is located exclusively on the floor of the mouth. It is considered to be a mucus retention cyst of the sublingual gland[4,5] or a mucous extravasation phenomenon of either the major salivary glands or the minor sublingual salivary glands.[1] It appears as an enlargement of normal to bluish color, varying from a fluctuant process to a lesion of soft consistency, depending on the thickness of the overlying tissues. Children and young adults are more frequently affected, and a history of repetitive rupture and recurrence is a common finding. Plunging ranula is a variation of the ranula, produced by dissection of extravasated mucin through the mylohyoid muscle.[1,5] It appears as a lump in the submandibular neck area, with or without intraoral signs.[1,5] Although ranulas appear well defined and unilocular on CT imaging,[9] they are characteristically hypodense.

Inflammatory disorders of the salivary glands merited consideration in the differential diagnosis of the present lesion. Acute sialadenitis of the submandibular gland usually presents with swelling associated with meals and pain and purulent discharge from the Wharton duct,[1,2,4] and chronic sialadenitis runs a prolonged course of remissions and exacerbations. Both conditions are more frequent in the sixth decade of life and present no gender predilection.[4] Moreover, both conditions usually involve a number of predisposing factors, such as sialolithiasis, chronic illness, hospitalization, or medications, all of which may be associated with xerostomia. Especially regarding drug-induced xerostomia, it is worth mentioning that the geriatric population is more susceptible, because the risk increases with the number of drugs taken.[1,2,10] Although in most cases of sialadenitis the infection is bacterial, viral causes, i.e., cytomegalovirus or paramyxovirus, are also not uncommon. The present patient presented no such risk factors and reported no previous episodes of swelling in the area. The size and asymptomatic nature of the swelling would argue against an inflammatory salivary gland origin. However, a Kuttner tumor, a subset of chronic sclerosing sialadenitis, can achieve considerable size and was included in the clinical differential diagnosis.[11]

Sialosis is often related to disorders such as diabetes, acromegaly, hypothyroidism, alcoholism, or general undernourishment.[1] Medications affecting the autonomous nervous system, such as antihypertensive or psychotropic drugs, are also associated with sialosis.[1] Benign lymphoepithelial lesions commonly occur as part of Sjögren syndrome.[1] Although the lesion affects mostly the parotid glands, involvement of the submandibular glands may be seen.

Pleomorphic adenoma and monomorphic adenoma are usually included in the differential diagnosis of painless and slowly growing intraoral masses. However, salivary gland neoplasms of the floor of the mouth are more often malignant than benign.[4] The present patient's age conforms to the higher incidence of salivary gland neoplasms.[1,4,12] However, the posterior hard palate/soft palate is the site of predilection for minor salivary gland tumors.[2,4] The most common malignant neoplasms of salivary glands are the mucoepidermoid carcinoma and the adenoid cystic carcinoma.[1] Acinic cell carcinoma is also seen, especially in older patients.[2] Nevertheless, in the case of a malignant neoplasm, the growth is often fixed to adjacent structures. Adenoid cystic carcinoma also frequently invades neighboring nerves, resulting in hypoesthesia of the lingual nerve.

Another possible cause of a cervical swelling is lymphoma, both Hodgkin and non-Hodgkin.[13,14] Although more common in children and young adults, lymphoma may present in older patients as well.[13,15]

The 3-month course of asymptomatic growth, the lack of neurologic signs or evidence of attachment to surrounding structures, and the soft consistency directed the differential diagnosis toward a benign tumor. Furthermore, the normal appearance of the overlying mucosa is suggestive of a benign tumor of mesenchymal origin, such as lipoma, neurilemmoma, or leiomyoma, or of salivary gland origin.[1,2] Lipoma is quite rare in the oral cavity. The buccal mucosa is the site of predilection, with 50% of the cases, but the floor of the

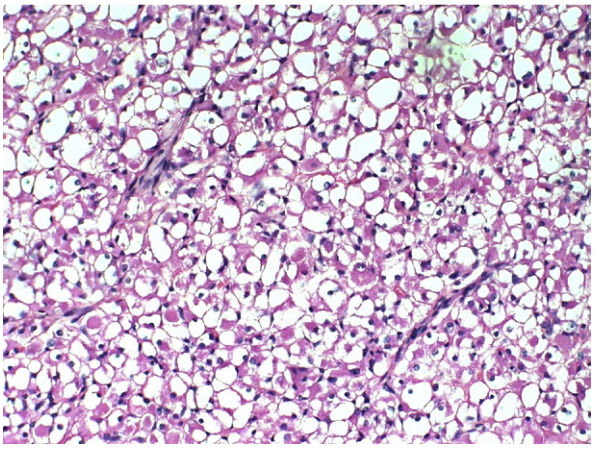

Fig. 4. Histologic examination revealed a lesion consisting of large polygonal cells with eosinophilic to clear cytoplasm (hematoxylin and eosin stain, original magnification ×200).

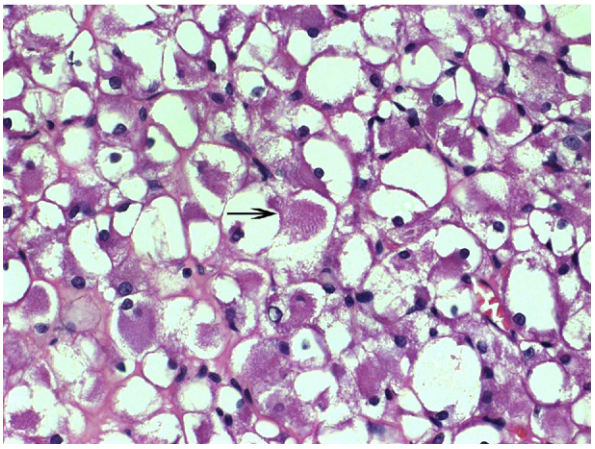

Fig. 5. Higher power histologic examination shows cross-striations (*arrow*) in lesional cells (hematoxylin and eosin stain, original magnification ×400).

mouth may also be involved. It typically presents as a slowly enlarging mass covered by normal appearing mucosa of yellowish hue. Middle-aged persons are more prone to develop lipomas, and no gender predilection has been reported.[1,2] The CT imaging of a lipoma is that of a hypodense nonenhancing lesion, in contrast to the present case.

Malignant soft tissue tumors were considered to be a remote possibility. Rapid growth, lack of circumscription, fixation to adjacent structures, and ulceration of the overlying mucosa[1,12] are common features of malignancy that were not noted in the present patient.

MANAGEMENT AND DIAGNOSIS

With the provisional diagnosis of a benign soft tissue tumor, an incisional biopsy under local anesthesia was planned. During the initial incision and identification of Wharton duct, it became apparent that the growth was encapsulated and clearly separated from nearby structures, allowing for the excision of the lesion in toto.

The surgical specimen, consisting of a brown encapsulated lobulated tumor measuring 3.0 × 1.5 cm, was fixed in 10% buffered formalin and processed for routine histopathologic examination. Hematoxylin and eosin–stained sections showed a benign lesion encapsulated by fibrous connective tissue. The lesion consisted of large polygonal cells with eosinophilic to clear cytoplasm (Fig. 4), several presenting with discrete cross-striations (Fig. 5). No nuclear or cellular pleomorphism or mitoses were seen. The cells stained red with Masson trichrome, and were immunohistochemically reactive for desmin and myoglobin.

The final diagnosis was adult rhabdomyoma.

The patient's postsurgical recovery was uneventful, without salivary gland obstruction or lingual hypoesthesia. Further management included endoscopy of the larynx and pharynx to exclude multifocal type of rhabdomyoma.[16] The patient's stomach, prostate, and heart were investigated with ultrasonography. Moreover, the patient had no features associated with tuberous sclerosis, such as childhood seizures, hypomelanotic macules, facial angiofibromas, or bone cysts.[17]

At the time of writing, the patient had been under regular follow-up for 2 years and remained free of recurrence or new lesion.

DISCUSSION

Rhabdomyomas are rare benign tumors originating from striated muscle[18,19] and classified as cardiac and extracardiac.[18] Cardiac rhabdomyomas affect the myocardial fibers and produce a diffuse distortion of the heart muscle. They are generally considered hamartomas rather than true neoplastic lesions and are usually associated with tuberous sclerosis.[17,18]

The number of extracardiac rhabdomyomas reported in the literature exceeds 160 cases, since the first report in 1897.[16,19,20] Extracardiac rhabdomyomas are of 3 types: adult, fetal, and genital. The majority of extracardiac rhabdomyomas are of the adult type and occur in the head and neck.[5,16,19] The fetal type, also occurring more commonly in the head and neck region, is considered to be a developmental abnormality.[19,21] The adult type usually presents at a mean age of 50 years with a male preponderance of 4:1.[16,20] The genital type, in contrast, is a solitary lesion that occurs more commonly in women.[20,22]

On microscopic examination, the fetal rhabdomyoma is composed of slightly differentiated polygonal cells admixed with spindle-shaped cells.[23] This type is typ-

ically more cellular than the adult type and often has a myxoid stroma. Lesions with pleomorphic characteristics and increased mitotic activity can rarely be mistaken for rhabdomyosarcomas.[23] The adult type presents a simpler structure compared with the fetal subtype, with large ovoid or polygonal cells with granular eosinophilic cytoplasm. Some cells present a vacuolated cytoplasm due to the accumulation of glycogen.[16,23,24] There are usually a large number of blood vessels, scant stroma, and a well defined capsule.[18] Routine hematoxylin and eosin histologic features are usually sufficiently characteristic. Immunohistochemical markers of skeletal muscle differentiation, such as desmin, myoglobin, and muscle-specific actin, may occasionally be of value in confirming the diagnosis.[16,25]

The treatment of choice for rhabdomyoma is surgical excision. Multifocal presentation[26,27] and local recurrence[16,28] are possible, mandating long-term follow-up, especially for lobulated tumors or tumors showing invasive growths.[16,20] Recurrence is usually associated with inadequate resection and not locally aggressive biologic behavior.[16,24,28]

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