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CASE REPORT

Schwannoma arising from the sublingual glandular branch of the lingual nerve radiologically masquerading as sublingual gland tumor

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Abstract
We report a rare case of schwannoma arising from the sublingual glandular branch of the lingual nerve radiologically masquerading as sublingual gland tumor. A 42-year-old female was referred to our department with a painless swelling in the left submandibular region. Contrast-enhanced computed tomography showed a well-circumscribed, heterogeneous low-density tumor with cystic change in the left sublingual region. Magnetic resonance imaging showed a well-circumscribed, heterogeneous sublingual tumor with low-signal intensity on T1-weighted image and high-signal intensity in T2-weighted image. The lesion was diagnosed radiologically as benign sublingual gland tumor. The patient underwent resection of sublingual gland tumor under general anesthesia. There was no definitive continuity between the tumor and the sublingual gland, and the tumor originated from sublingual glandular branch of the lingual nerve. Pathological examination of the specimen showed schwannoma with highly cellular areas (Antoni A) and hypocellular areas (Antoni B). The postoperative course was uneventful without lingual nerve palsy, and there was no recurrence 4 years after surgery.

Keywords Glandular branch · Lingual nerve · Schwannoma · Sublingual gland

Introduction

Schwannoma is a benign, slow-growing, usually solitary and encapsulated tumor which is composed of Schwann cells. Nearly half of all schwannomas occur in head and neck region [1], and about 25–45% of extracranial schwannomas are found in the head and neck region [2, 3]. This tumor is more likely to be seen in females [3], and there is no significant preference at any age. Intraoral schwannoma accounts for 20–60% of extracranial schwannomas of head and neck [2]. Most common site of intraoral schwannoma is the tongue, followed by the palate, floor of the mouth, buccal mucosa, gingiva, lips, and vestibule in decreasing order [4]. The origin of schwannoma in floor of the mouth is commonly the lingual or hypoglossal nerve. To our knowledge, there are a few reports of schwannoma arising from the sublingual glandular branch of the lingual nerve [5]. We report a rare case of schwannoma arising from the sublingual glandular branch of the lingual nerve radiologically masquerading as sublingual gland tumor.

Case report

A 42-year-old female presented to a private ENT clinic with a painless swelling in the left submandibular region. Because ultrasonography (US) showed sublingual mass, fine needle aspiration cytology (FNAC) was performed. The specimen was inconclusive of the diagnosis, but malignancy was not seen. For the treatment of the sublingual mass, she was referred to department of oral and maxillofacial surgery in our hospital. The elastic hard mass, measuring approximately 4 × 3 cm, was mobile and covered by normal mucosa without inflammation. Contrast-enhanced CT showed a well-circumscribed, heterogeneous low-density tumor (43 × 30 × 17 mm) with

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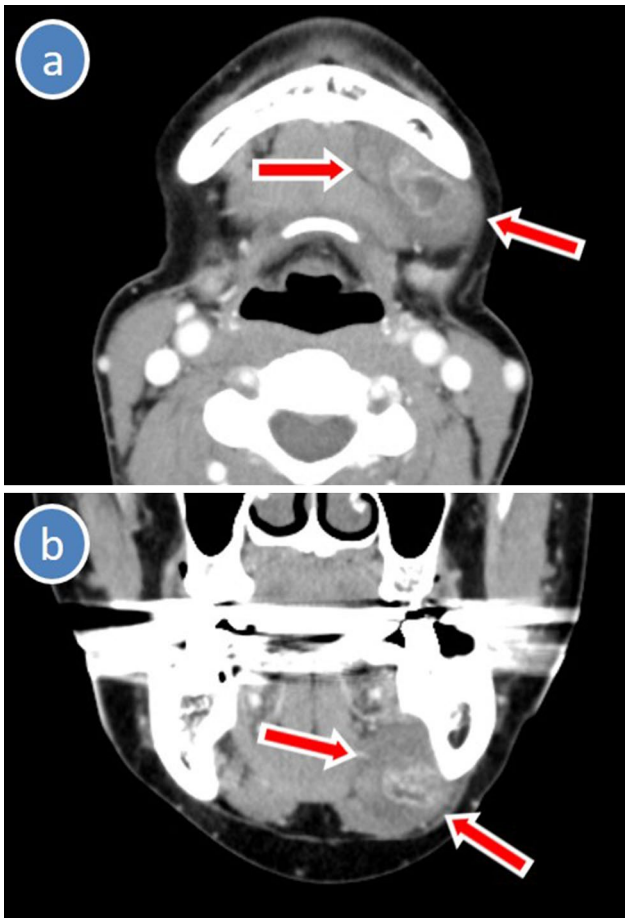

Fig. 1 CT images. Contrast-enhanced CT showed a well-circumscribed, heterogeneous low-density tumor mass in the left sublingual region. **a** Axial image; **b** coronal image

cystic change in the left sublingual region (Fig. 1). MRI showed a well-circumscribed, heterogeneous sublingual tumor (40 × 25 × 19 mm) with low-signal intensity on T1-weighted image, relative high-signal intensity in T2-weighted image and high-signal intensity in fat suppression T2-weighted image (Fig. 2). Target sign was not showed, and diffusion-weighted image was not performed. US showed a 40 × 25 mm solid tumor with blood flow and cystic lesion (Fig. 3). The lesion was diagnosed radiologically as benign sublingual gland tumor, such as pleomorphic adenoma. The patient underwent resection of sublingual gland tumor under general anesthesia. A mucosal incision was made over the mass in the left side of floor of the mouth. After identification of the lingual nerve and Wharton's duct, the sublingual gland was dissected from the mylohyoid muscle. The main trunk of the lingual nerve or hypoglossal nerve had no direct attachment to the tumor. There was no definitive continuity between the tumor and the sublingual gland, and the tumor originated from sublingual glandular branch of the lingual nerve, which were thought to be the parasympathetic nerve of the sublingual gland. After the cutting of the sublingual glandular branch posterior the tumor, sublingual gland with tumor was removed completely (Figs. 4, 5). Because intraoperative rapid pathological diagnosis showed schwannoma with no malignancy, the lingual nerve, Wharton's duct, and hypoglossal nerve could be preserved. Pathological examination of the specimen showed highly cellular areas (Antoni A) and hypocellular areas (Antoni B) in the tumor (Fig. 6). There were bundles of spindle-shaped cells and palisading nuclei in some areas, and Antoni type A was predominant. There was low grade of nuclear pleomorphism in some areas, but no mitosis was seen. The postoperative course was uneventful without lingual nerve palsy, and there was no recurrence 4 years after surgery.

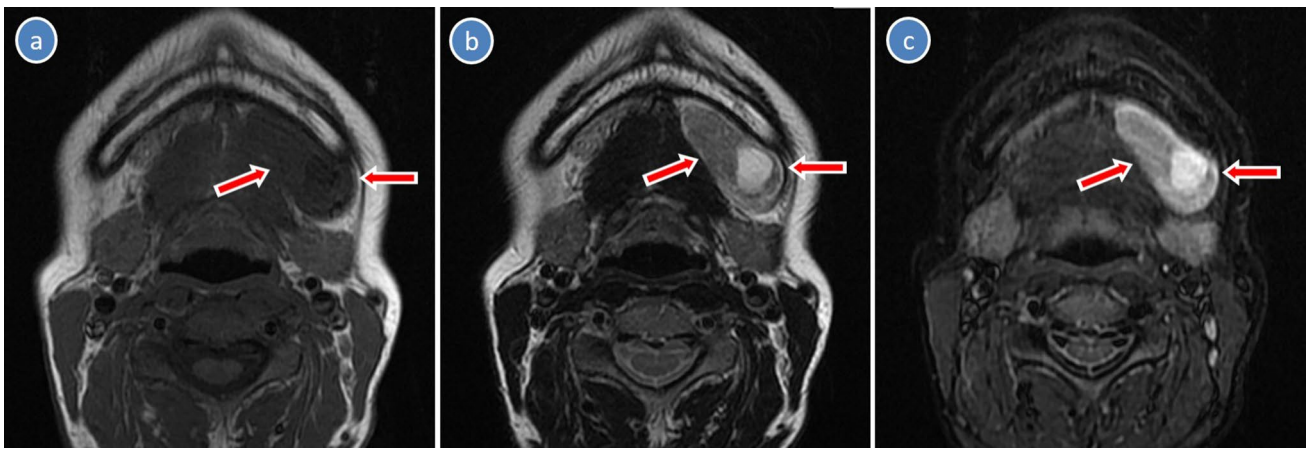

Fig. 2 MR images. MRI showed a well-circumscribed, heterogeneous sublingual tumor mass (arrows) with low-signal intensity on T1-weighted image (**a** TR/TE = 400/10), high-signal intensity in T2-weighted image (**b** TR/TE = 3067/107), and high-signal intensity in fat suppression T2-weighted image (**c** TR/TE = 3300/23)

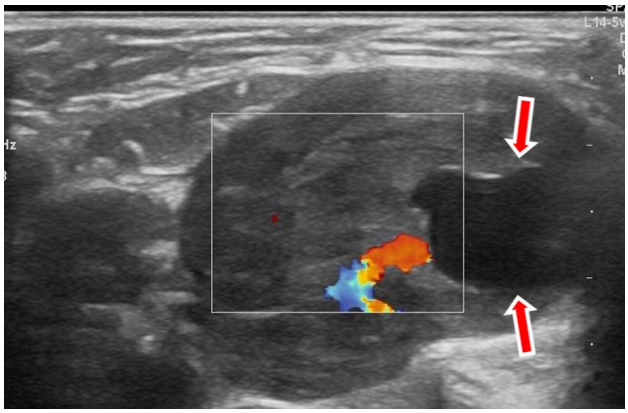

Fig. 3 US image. US showed a 40×25 mm solid tumor with blood flow and cystic lesion (arrows)

Discussion

The origin of intraoral schwannoma is commonly the lingual or hypoglossal nerve, but the origin in floor of the mouth is not always identified [3, 4]. Schwannoma arising from the sublingual gland is extremely rare, and only two cases have been reported in the English literature [6, 7]. The evidence of gland remnant or duct without a capsule within tumor is highly confirmatory that the tumor is arising from the gland itself [6, 7]. In the present case, there were no sublingual gland remnants with no demonstrable capsule. There are a few reports of schwannoma arising from the sublingual glandular branch of the lingual nerve [5]. We identified the nerve of origin as the branch of the lingual nerve, which carried the postganglionic parasympathetic nerve fibers. To our knowledge, the present case was the second report of schwannoma arising the sublingual glandular branch of the lingual nerve. In contrast, there were only two cases of schwannoma of the submandibular gland originating from the submandibular branch of the lingual nerve, which carries the preganglionic parasympathetic nerve fibers [1, 8]. Schwannoma arising from the parasympathetic nerve fibers may masquerade as sublingual gland tumor like present case.

Diagnosis of schwannoma is performed by clinical features, such as long-standing history and firm mass which is more mobile along the plane perpendicular to the course of the nerve and nerve palsy [9]. Furthermore, FNAC is performed for the diagnosis of schwannoma, but is commonly not helpful for diagnosis of schwannoma [9–11]. In schwannomas arising from sublingual gland or parasympathetic

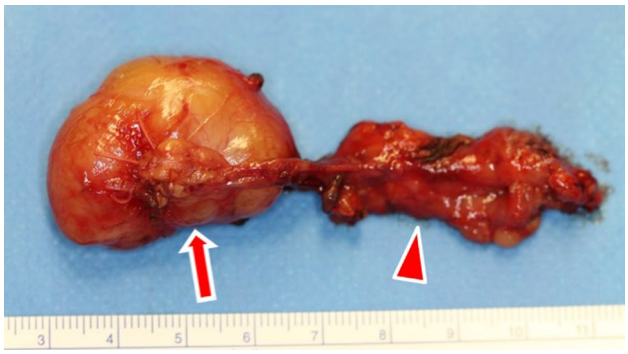

Fig. 5 Removed specimen. Arrow head indicates the sublingual gland. Arrow indicates the tumor mass

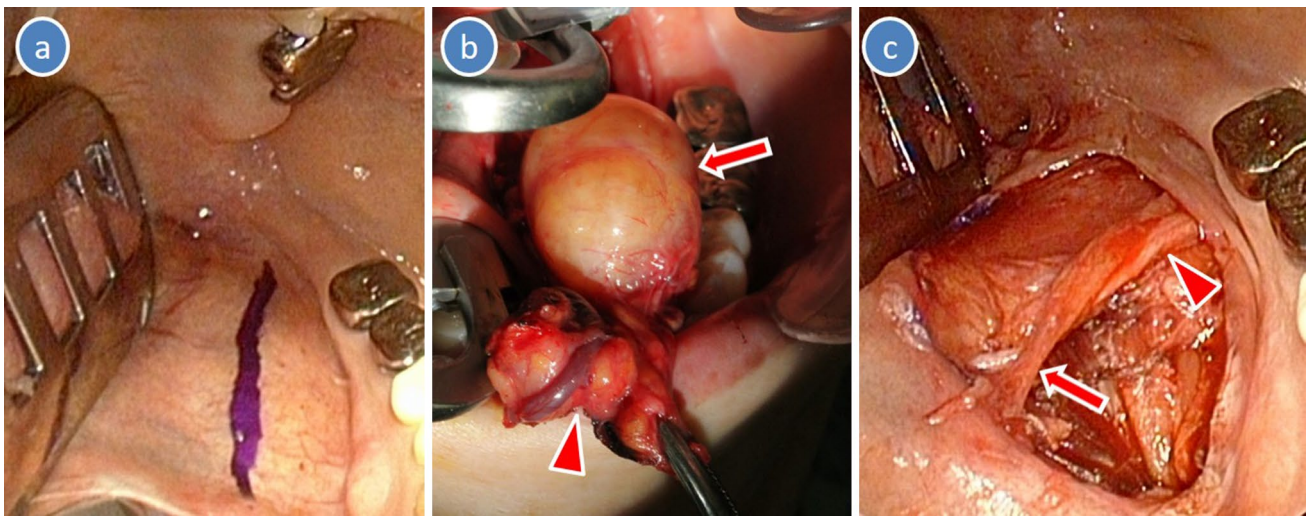

Fig. 4 Intraoperative view. **a** Intraoral view before mucosal incision of the oral floor without swelling. **b** Removal of tumor mass (arrow) with the sublingual gland (arrow head). **c** Surgical view after complete removal of the tumor. Arrow head indicates the lingual nerve. Arrow indicates the Wharton's duct

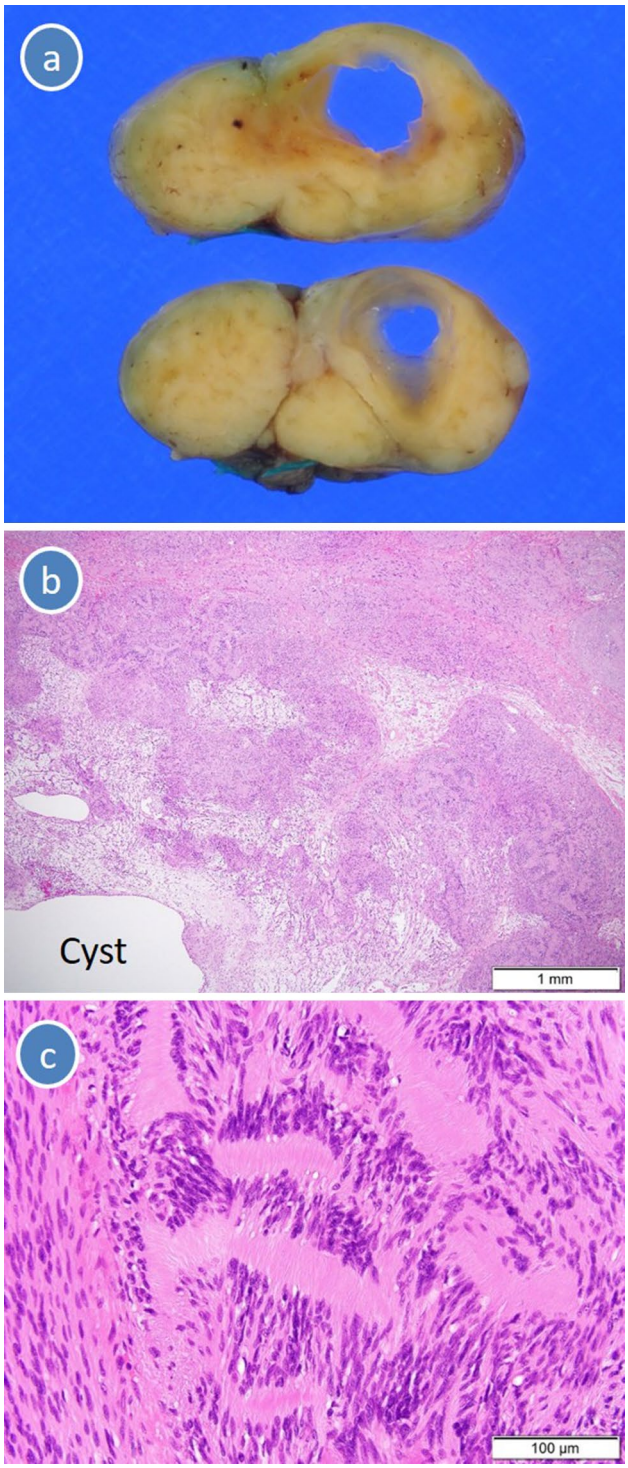

Fig. 6 Macroscopic and microscopic pathology of the specimen. **a** Macroscopic pathology. Sectioning of tumor including cystic formation. **b** Microscopic pathology. Mixing highly cellular areas (Antoni A) and hypocellular areas (Antoni B) (hematoxylin and eosin staining). **c** Microscopic pathology (Antoni A area). Spindle-shaped cells are proliferating in bundle-like arrangement. The palisade arrangement of the nuclear is clear (hematoxylin and eosin staining)

nerve fibers including the present case, FNAC was not useful for the diagnosis [1, 5–8]. Preoperative or intraoperative incisional biopsy is necessary for the diagnosis of schwannoma and exclusion of malignancy.

Several imaging modalities also are often ordered for the diagnosis of schwannoma. US images typically show a well-defined, ovoid or round, hypoechoic, primary homogenous solid mass with or without a moderate posterior acoustic enhancement [12]. King et al. [13] reported that schwannoma is highly vascular tumor with an abundance of vessels and blood flow that is easily obliterated with light pressure from the probe, and cystic change may be seen. These US findings were seen in the present case. The CT features of schwannoma are well-circumscribed tumor with low or soft-tissue attenuation and homogeneous enhancement or heterogenous enhancement reflecting its histological features such as smooth margin and tissue homogeneity [1]. According to Kami et al. [12], the low-enhanced area is considered to be Antoni B area and the slightly more enhanced component is considered as corresponding to Antoni A area. Schwannoma sometimes includes cystic changes [12], as the present case. On MRI, schwannoma appears low-signal (isointense signal to muscle) intensity on T1-weighted images, and high-signal intensity on T2-images [10, 12]. Target sign (biphasic pattern of peripheral hyperintensity and homogeneous central hypointensity on T2-weighted images) and reverse target sign (biphasic pattern central high intensity and peripheral low intensity on gadolinium-enhanced T1-weighted images) are specific to the schwannoma [12, 14]. On diffusion-weighted image, schwannoma is reported to show high apparent diffusion coefficient (ADC) values [15], which probably arise through the less dense cellular areas of the tumors. MRI is most sensitive and specific in the diagnosis of schwannoma in the head and neck region, and the accuracy is 80% [11]. The present case had similar MRI features with cystic degeneration, but target sign was not showed. Although the relationship between schwannoma and its nerve origin can be better appreciated with MRI than CT [11], the diagnosis of sublingual schwannoma or the identification of the nerve origin is very difficult even in MRI. Especially, the differential diagnosis is very important, and it is challenging to differentiating sublingual schwannoma from sublingual gland tumor.

Sublingual gland tumor is very rare, accounting for only 0.3–1% of all epithelial salivary gland tumors, and most (70–90%) of salivary gland tumors are malignant, with the most common histologic types being adenoid cystic carcinoma and mucoepidermoid carcinoma [7, 16]. In high-grade carcinoma, MRI shows that the internal signal is characteristically low-to-intermediate on both T1-weighted and T2-weighted images [16]. In contrast, low-grade carcinoma is defined with low T1-weighted and high T2-weighted internal signal, simulating its benign counterpart [16]. The

most common benign sublingual gland tumor is pleomorphic adenoma, which is mixed salivary gland neoplasm comprising the combination of epithelium and myoepithelial cells. On MRI, pleomorphic adenoma shows low T1-weighted and high T2-weighted internal signal [16]. The MRI findings of low-grade carcinoma and benign tumor in sublingual gland are similar with schwannoma. In the present case, clinical and radiological diagnosis was benign sublingual gland tumor such as pleomorphic adenoma, but the final pathological diagnosis was schwannoma. Although the diagnosis of schwannoma is very difficult even in MRI and is decided by only pathological examination, surgeons and radiologists should consider the possibility of schwannoma in radiological diagnosis of sublingual tumor.

There are advantages and limitations in several imaging modalities for radiological diagnosis of sublingual tumor. US provides excellent tissue characterization, multi-planner information, and vascular pattern with Doppler technique. Furthermore, there are advantages of low cost and no radiation exposure. Because CT and MRI have metallic artifacts, good images are not always acquired. Although MRI without radiation exposure is superior in its soft-tissue differentiation than CT, the disadvantage of MRI is the relative high cost, susceptibility to motion artifacts, and poor cortical bone delineation compared to CT. In conclusion, surgeons and radiologists should take schwannoma into consideration in radiological differential diagnosis of sublingual tumor.

Compliance with ethical standards

Conflict of interest Shuhei Minamiyama, Toshinori Iwai, Satomi Sugiyama, Yuichiro Hayashi, Makoto Hirota, and Kenji Mitsudo declare that they have no conflict of interest.

Human rights statement All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Informed consent Informed consent was obtained from the patient for being included in this study.

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