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Hemangiopericytoma/solitary fibrous tumor of the buccal mucosa: A case report

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ABSTRACT

Hemangiopericytoma/solitary fibrous tumor (HPC/SFT) is a perivascular mesenchymal tumor often found unexpectedly on histopathological examination, and occasionally shows malignant behavior. The incidence of intraoral HPC/SFT is extremely rare. We report a case of HPC/SFT located in the buccal mucosa. A 50-year-old woman presented with a 5-year history of a painless mass in the left buccal mucosa. Clinical findings showed the lesion was a well-defined tumor between the cheek skin and buccal mucosa with two feeding arteries, indicating hemangioma. The tumor was completely resected under general anesthesia. Histopathologically, immunostaining for STAT6 revealed results consistent with HPC/SFT, and no findings suggestive of malignancy, such as tumor size greater than 5 cm and high proliferative activity as shown by mitotic index and Ki-67 index. No other distinct primary lesion or distant metastasis was detected on whole-body computed tomography. Dealing with the lesion as a precancerous or potentially malignant tumor, follow-up was performed for 5 years after surgery, but neither recurrence nor metastasis was observed. As recurrence or metastasis may be delayed by many years, follow-up needs to be continued long-term according to risk factors of malignant behavior such as tumor size, cell characteristics and proliferative activity.

1. Introduction

Hemangiopericytoma/solitary fibrous tumor (HPC/SFT) is a rare perivascular tumor arising from capillary pericytes, and accounting for 1% of all vascular tumors. HPC/SFT is considered potentially malignant, because it occasionally shows malignant behavior [1,2]. HPC/SFT can occur in any part of the body where pericytes are present and is commonly seen in the inferior limbs and retroperitoneum. However, this pathology is also often found unexpectedly on histopathological examination because HPC/SFT has no specific characteristic clinical or imaging features [3]. The reported incidence of head and neck HPC/SFT is approximately 15% [1]. Most cases of head and neck HPC/SFT are observed in the nasal cavity [4], while intraoral HPC/SFT is relatively rare. We report herein a case of HPC/SFT of the buccal mucosa.

2. Case report

A 50-year-old woman presented with a 5-year history of a small painless mass in the right buccal mucosa. She was referred to our department due to gradual enlargement of the mass. No past or family medical history that could have contributed to the present illness was identified. Extraorally, slight swelling was evident in the right cheek region and no facial nerve palsy was evident. Intraorally, an elastic, hard, mobile mass was located under the right buccal mucosa (Fig. 1A). Contrast-enhanced computed tomography (CT) revealed a heterogeneously enhancing lesion measuring 24 × 20 mm in size between the cheek skin and buccal mucosa (Fig. 1B). Magnetic resonance imaging (MRI) revealed a spherical, well-defined tumor in front of the anterior border of the right masseter muscle. The tumor appeared hypointense on T1-weighted imaging, and hyperintense and heterogeneous on T2-weighted imaging (Fig. 1C). These MRI manifestations indicated the tumor was hemangioma. The facial and buccal arteries appeared to be feeders to the tumor on digital subtraction angiography (Fig. 1D). The clinical diagnosis was hemangioma of the right buccal mucosa, and complete tumor removal was performed via an intraoral approach under general anesthesia. The tumor was well demarcated from the

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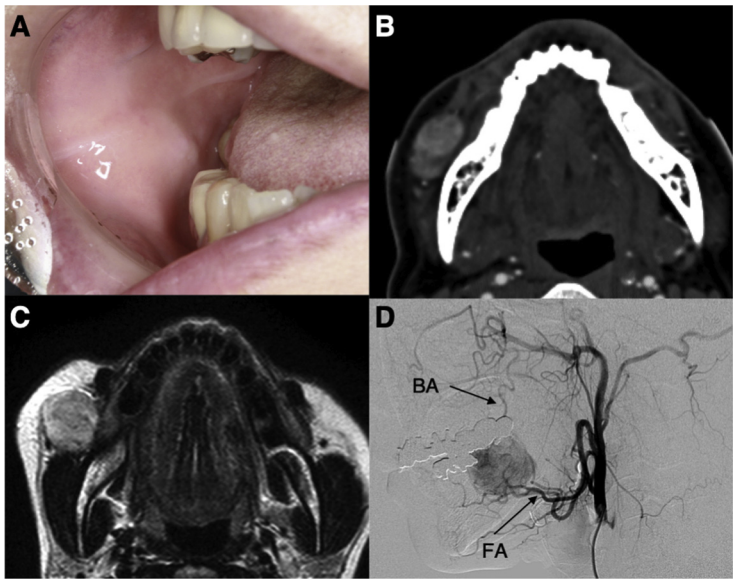

Fig. 1. A) The mass is located under the right buccal mucosa, and covered with normal mucosa. B) Contrast-enhanced CT showing a heterogeneously enhancing lesion between the cheek skin and buccal mucosa. C) MRI reveals a well-defined tumor in front of the anterior border of the right masseter muscle. The tumor shows heterogeneous hyperintensity on T2-weighted image. D) The facial and buccal arteries are considered as feeders to the tumor.
Abbreviations: BA: buccal artery; FA: facial artery.

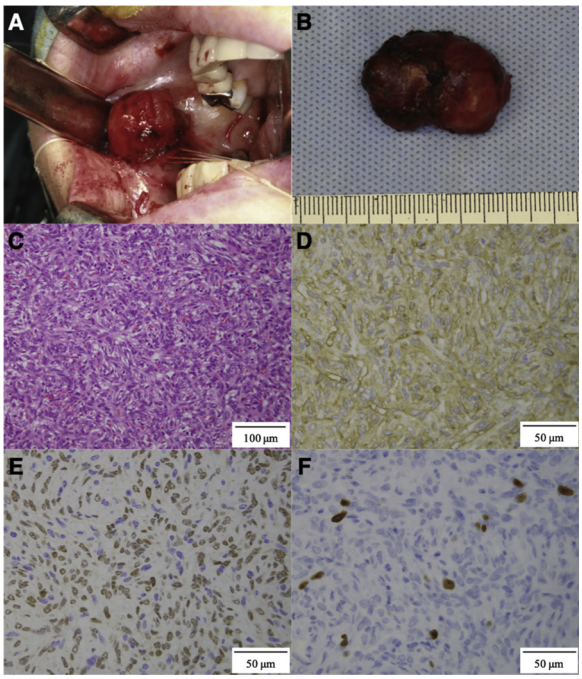

Fig. 2. A) Complete tumor removal is performed via an intraoral approach. B) The tumor is located between the cheek skin and buccal mucosa, approximately 30 mm in diameter, and well encapsulated with reddish fibrous tissue. C) Hematoxylin-eosin stain reveals prominent vasculature and increasing stromal cellularity. Spindle cells with round or oval nuclei are seen. D–F) Immunohistochemistry reveals high expression of CD34 (D) and STAT6 (E), and Ki-67 index is low (2–3%) (F).

surrounding tissue and the feeding arteries were ligated (Fig. 2A). The tumor was approximately 30 × 20 mm in size and was well encapsulated (Fig. 2B). No postoperative complications were seen. Microscopic examination showed the tumor comprised spindle cells with round or oval nuclei. Immunohistochemistry revealed high expressions of CD34 and STAT6. On the other hand, CD31, SMA, desmin, and S-100 staining were negative. Expression of Ki-67 was poor and the staining index was 2–3% (Fig. 2C-F). Histopathological diagnosis revealed the tumor was hemangiopericytoma, with no malignant characteristics. No other tumor such as a distinct primary lesion or distant metastasis was detected on whole-body CT. Dealing with the lesion as precancerous or a potentially malignant tumor, follow-up contrast-enhanced CT was performed annually for 5 years after surgery, neither recurrence nor metastasis was observed.

3. Discussion

HPC/SFT, as a rare mesenchymal tumor arising from capillary pericytes, was first described by Stout and Murray in 1942 [5]. The peak prevalence of HPC/SFT is in the sixth to seventh decade of life with no sex predilection [2]. Even though manifestations in the head and neck region are reported in 15% of all HPC/SFTs [1], intraoral HPC/SFT is extremely rare. The local recurrence rate has been reported as approximately 25–40% of head and neck cases, so complete tumor resection is recommended as the primary therapy for HPC/SFT [4]. The efficacy of radiation therapy is controversial, because HPC/SFT is considered to be radiation-resistant. Wushou et al. [4] reported that surgery with adjuvant radiotherapy was not superior to surgery alone in 116 cases of head and neck HPC/SFT. Neoadjuvant and adjuvant chemotherapy is not administered due to poor response to traditional chemotherapy in the metastatic setting. Recently, pazopanib or similar multi-targeted tyrosine kinase inhibitors such as bevacizumab and temozolomide have been used in the treatment of metastasis cases [1].

Consistent diagnosis of HPC/SFT prior to starting treatment is difficult because of the poor clinical findings. MRI has advantages in detecting highly vascularized tumors, while angiography is advantageous for confirming the diagnosis of vascular neoplasm and achieving preoperative definition of the blood supply. While, HPC/SFT is shown basically homogenous hypoechoic, but occasionally heterogenous hypo-hyperechoic lesion at ultrasound imaging. In addition, lack of color flow at Doppler imaging despite its hypervascularity is reported to be a useful feature of HPC/SFT [6]. However, these modalities are still inappropriate to distinguish HPC/SFT from other tumors, because HPC/SFT often has feeding arteries like hemangioma [7]. Both fine needle aspiration biopsy and incisional biopsy may be able to reach consistent diagnosis, but these techniques show a risk of bleeding after treatment. This is particularly relevant for tumors with multiple feeding arteries. Microscopic examination was thus avoided in the present case. Most cases of HPC/SFT are diagnosed after complete resection, even when incisional biopsy has been performed [3]. Immunohistochemical staining is useful for diagnosis, and HPC/SFT shows immunoreactivity for STAT6 and CD34, and often positive results for BCL-2 and CD99 [1]. Positive CD34 expression has traditionally been used to confirm the diagnosis, but is non-specific for HPC/SFT. At present, nuclear

expression of STAT6 remains the 'gold standard' for identifying this pathology. Previously, HPC was considered a controversial tumor and its differentiation from SFT has been debated. However, recent studies have revealed that HPC and SFT share inversions at 12q13, fusing the NAB2 and STAT6 genes [8]. Thus, it has become clear that HPC and SFT overlap and have thus been integrated into the new entity of "solitary fibrous tumor/hemangiopericytoma" in the 2016 World Health Organization classification [9].

In the present case, tumor resection was performed with preoperative diagnosis of hemangioma, but the postoperative histopathological diagnosis was HPC/SFT, unexpectedly. HPC/SFT is classified as a potentially malignant tumor according to the clinical and histological features. Tumor size greater than 5.0 cm in diameter, presence of > 4 mitotic figures per 10 high-powered fields, and areas of tumor necrosis and nuclear pleomorphism have been reported as risk factors for malignant behavior [2]. Kowalski et al. [10] evaluated the prognostic significance of the proliferation index using Ki-67 staining in HPC/SFT of the head and neck, and reported that a proliferation index of $\geq 10\%$ may be indicative of a malignant subset. Espat et al. [11] reported that all cases treated with complete resection showed a 100% survival rate at 5 years after surgery, suggesting radicality of surgery is required to control malignant behavior of HPC/SFT. Although there is no clearly established safety margin to resect head and neck HPC/SFT, complete resection should be planned in consideration of the clinical and histological malignant risk factor as mentioned above in the case which obtained consistent diagnosis ahead of surgery.

The present tumor was completely removed and did not correspond to any of the above factors. No recurrence or metastasis has been seen for 5 years. The appropriate duration of follow-up for HPC/SFT is under debate because of the malignant potential. The duration to recurrence or distant metastasis was reported as approximately 17 months and 40–60 months, respectively [4,12]. Subject to complete resection of the tumor, the above-mentioned histopathological characteristics of malignant behavior and mean duration until recurrence or distant metastasis should be considered when deciding the duration of follow-up.

Ethical approval

Informed consent was obtained from the patient.

Conflict of interest

None.

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