

Case Report

Angiosarcoma of the mandibular gingiva

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Abstract: Angiosarcoma of the oral cavity is extremely rare. A 77-year-old woman consulted to our hospital because of polypoid mass of the mandibular gingival. Physical examination showed polypoid reddish mass measuring 1.5 x 1.5 x 1 cm in the mandibular gingival posterior to the front tooth. Enucleation of the tumor was performed. Grossly, the tumor was not encapsulated. Histologically, the tumor consisted of atypical spindle with hyperchromatic nuclei with nucleoli. Mitotic figures were scattered. Vasoformative channels were present in some areas. The surgical margins were positive. Immunohistochemically, the tumor cells were positive for factor VIII-related antigen, CD31, CD34, vimentin, p53 protein, but negative for pancytokeratin (AE1/3 and CAM5.2), S100 protein, α -smooth muscle antigen, and desmin. The Ki-67 labeling was 60%. A pathological diagnosis of angiosarcoma was made. Radical operation is planned now.

Keywords: Oral cavity, angiosarcoma, histopathology

Introduction

Angiosarcoma is a malignant mesenchymal tumor with a differentiation into vascular endothelium. Although angiosarcoma can occur in any location, the most common sites are soft tissue and skin [1]. Angiosarcoma of the oral cavity is extremely rare; there are a few case reports [2-4] and case series [5] in the literature. The author recently encountered a case of angiosarcoma of cheek mucosa, and this case was reported elsewhere [6]. The author very recently experienced angiosarcoma of mandibular gingival. The author herein reports this case of angiosarcoma of the mandibular gingiva.

Case Report

A 77-year-old Japanese woman consulted to our hospital because of oral polypoid reddish mass (1.5 x 1.5 x 1 cm). located in mandibular gingival posterior to the front tooth. Excision was performed. Grossly, the tumor was 1 cm in diameter and was not encapsulated, and showed a central cavity (**Figure 1**). Histologically, the tumor was composed of atypical spindle cells with hyperchromatic nuclei and nucleoli (**Figure 2A**). Mitotic figure were scattered. In some ar-

eas, vasoformative channels containing red blood cells were recognized (**Figure 2B**).

An immunohistochemical study was performed with the use of Dako Envision method (Dako, Glostrup, Denmark), as previously described [7,8]. The tumor cells were positive for positive for vimentin (Vim 3B4, Dako), factor VIII-related antigen (F-VIII-RA) (36B11, Novocastra, Newcastle upon type, UK) (**Figure 3A**), CD34 (NU-3A1,

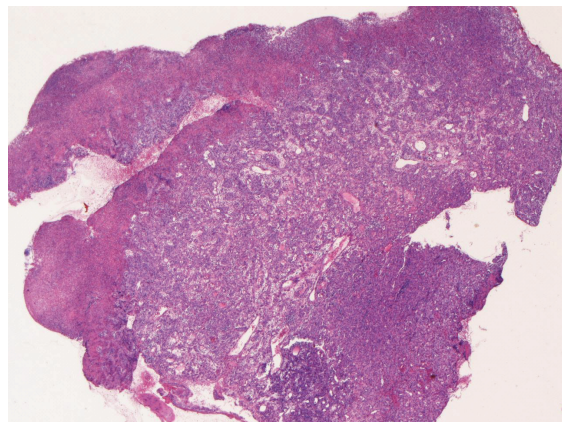


Figure 1. Very low power view of the tumor. The tumor is cellular solid tumor. Mucosa shows erosion. The surgical margins are positive. HE, x4

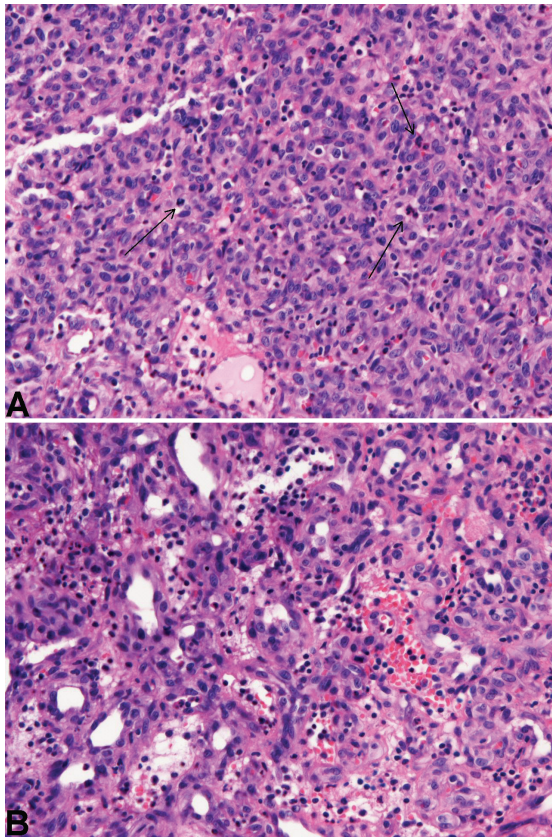


Figure 2. Histologic features of the tumor. A: The tumor is composed of atypical spindle cells with hyperchromatic nuclei arranged in solid nests. Nucleoli are present. Mitotic figures are present (arrows). No apparent differentiation is recognized in this area. HE, x200. B: This area shows apparent vasoformative channels, some of which contain red blood cells. HE, x200.

Dako) (**Figure 3B**), and CD31 (1A1, Novocastra) (**Figure 3C**) and p53 protein (DO-7, Dako) (**Figure 3D**). In contrast, they were negative for pancytokeratin (AE1/3, Dako; CAM5.2 Beckton-Dickinson, CA, USA), S100-protein (polyclonal, Dako), desmin (D33, Dako), α -smooth muscle antigen (1A4, Dako). The Ki-67 (MIB-1, Dako) labeling was 60% (**Figure 3E**). A pathologic diagnosis of angiosarcoma of the oral cavity was made. Radical operation or chemoradiation was planned now.

Discussion

The present tumor showed nuclear atypia and mitotic figures. The tumor cells were positive for p53 and Ki67 labeling was very high (60%). These findings show that the present tumor is

malignant, though the tumor is very small.

The present tumor appeared mesenchymal tumor on HE histology. The positive reaction to vimentin and negative reaction to pancytokeratins indicate the mesenchymal characters of the present tumor. The present tumor showed vasoformative channels, indicating that the present tumor is an angiogenic tumor. The positive reaction to F-VIII-RA, CD34 and CD31, which are endothelial markers, indicates that the present tumor have endothelial characteristics. The presence of red blood cells in the vasoformative channels suggests that the tumor is not a lymphatic tumor but a vascular tumor. Taken together, the present tumor is angiosarcoma histologically and immunohistochemically.

Differential diagnosis includes exuberant granulation tissue, malignant vascular tumors, such as epithelioid hemangioendothelioma, perivascular myoid tumor (malignant Glomus tumor and malignant myopericytoma), perivascular epithelioid tumor (malignant Pecoma), Kaposi's sarcoma, spindle cell carcinoma, intravascular endothelial hyperplasia, epithelioid angiosarcoma. The present tumor is different from exuberant granulation tissue, because the present tumor showed malignant nature on HE and immunohistochemical findings. The current tumor is different from epithelioid hemangioendothelioma which shows more little atypia, no vasoformative channels, and collagenization. The present case is different from malignant Glomus tumor and malignant myopericytoma histologically and negative smooth muscle markers. This case is different from Pecoma histologically and immunohistochemically, and is different from Kaposi's sarcoma in histological features and absence of multiple lesions and red cell extravasation. This tumor is apparently different from spindle cell carcinoma with regards to histology and immunohistochemistry. The tumor is obviously different from intravascular endothelial hyperplasia in histology and atypical features. The current tumor is different from epithelioid angiosarcoma histologically and absence of cytokeratin expression.

Generally, angiosarcoma is a very aggressive tumor. Angiosarcoma of the oral cavity may occur in various tissues, such as oral soft tissue, minor salivary glands, and bones [2-6]. The present case occurred in the cheek soft tissue in the oral cavity. Fanburg-Smith et al [5] reported 22 cases of primary angiosarcoma of the oral

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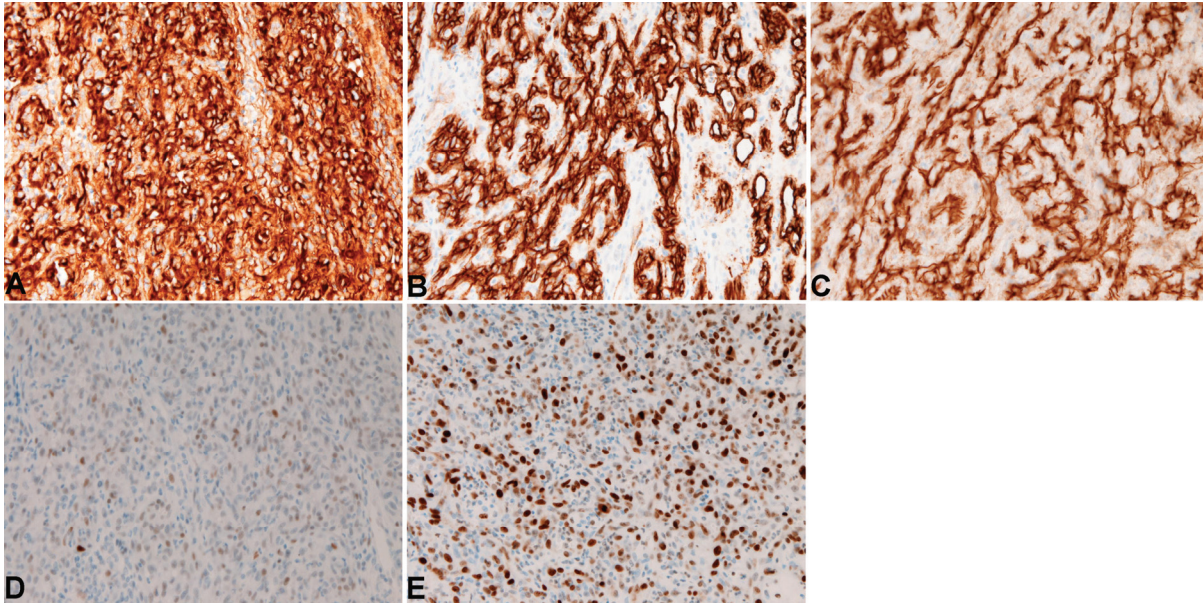


Figure 3. Immunohistochemical features of the tumor. A: The tumor cells are strongly positive for factor VIII-related antigen. x200. B: The tumor cells were positive for CD34. x200. C: The tumor cells were positive for CD31. x200. D: The tumor cells are positive for p53 in about 5%. E: The Ki-67 labeling is 60%. X200.

and salivary glands. The sites of the 22 cases were tongue in 9, parotid gland in 4, lip in 4, submandibular gland in 3, and palate in 1 [5]. Male and female were equally affected [5]. The symptoms are mass or mass bleeding. The size ranged from 0.8 -7.0 cm. Histologically, all tumors were vasoformative, 86 % had solid areas, and 17% had papillary areas. Immunohistochemically, F-VIII-RA was positive in 19/21, CD31 in 16/19, CD34 in 7/12, and Ulex lectin in 1/1 [5]. The survival differed depending on locations, and ranged from 1 year to 20 years with an average of 7.3 years. The survival is longer in low grade angiosarcoma than high grade angiosarcoma [5].

Conflict of interest

The author has no conflict of interest.

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References

[1] Fisher C. Soft tissue tumor. In: LeBoit PM, Burg G, Weedon D, Sarasin A eds. World

Health Organization Classification of Tumours. Pathology and genetics of skin tumours. IARC Press, Lyon, 2006. pp230-262.

- [2] Abdullah BH, Yahya HI, Talabani NA, Alash NI, Mirza KB. Gingival and cutaneous angiosarcoma. *J Oral Pathol Med* 2000; 29: 410-412.
- [3] Loudon JA, Billy ML, DeYoung BR, Allen CM. Angiosarcoma of the mandible: a case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000; 89: 471-476.
- [4] Favia G, Lo Muzio L, Serpico R, Maiorano E. Angiosarcoma of the head and neck with intra-oral presentation: a clinico-pathological study of four cases. *Oral Oncol* 2002; 38: 757-762.
- [5] Fanburg-Smith JC, Furlong MA, Childers EL. Oral and salivary gland angiosarcoma: a clinico-pathologic study of 29 cases. *Mod Pathol* 2003; 16: 263-271.
- [6] Terada T. Angiosarcoma of the oral cavity. *Head Neck Pathol* (in press).
- [7] Terada T, Kawaguchi M, Furukawa K, Sekido Y, Osamura Y. Minute mixed ductal-endocrine carcinoma of the pancreas with predominant intraductal growth. *Pathol Int* 2002; 52:740-746.
- [8] Terada T, Kawaguchi M. Primary clear cell adenocarcinoma of the peritoneum. *Tohoku J Exp Med* 2005; 206:271-275.