CURRENT ONCOLOGY
A Canadian Cancer Research Journal

Adult-onset Kaposiform hemangioendothelioma of the tongue: case report and review of the literature

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ABSTRACT

We present here a very rare clinical case of a 38-year-old man with Kaposiform hemangioendothelioma (KHE) of the tongue who presented to our institution with a growth under the left side of the tongue with no pain or discomfort. There were no enlarged lymph nodes and no significant neurologic findings. Diagnostic histopathology confirmed the lesion to be KHE. The tumour was removed surgically, and the surgical specimen confirmed the diagnosis. Follow-up at 3 months shows no clinical evidence of recurrence.

Key Words Kaposiform hemangioendothelioma, tongue, surgery

Curr Oncol. 2016 Oct;23(5):e517-e520 www.current-oncology.com

INTRODUCTION

Kaposiform hemangioendothelioma (KHE) is a very rare vascular neoplasm that was first described in 1993 by Zukerberg *et al.*[1]. It occurs mostly in children and is rarely observed in adults, with only approximately 20 adult KHE patients ever recorded[1,2]. Kaposiform hemangioendothelioma is locally aggressive, does not regress spontaneously, and has a tendency to metastasize locally as well as to regional lymph nodes. Cutaneous KHE typically appears as an erythematous violet-colour plaque with firm consistency, involving the full thickness of the skin and extending into the subcutaneous tissues. Sometimes, the skin surface shows hemorrhagic areas as the earliest sign of an accompanying Kasabach–Merritt phenomenon (KMP)[3], which is characterized by profound thrombocytopenia, life-threatening hemorrhage, and lymphangiomatosis[1,3].

The diagnosis of KHE is made on the basis of clinical presentation and standard pathologic examination, including immunohistochemical studies and imaging. Numerous treatment modalities are available for KHE, including surgery, radiation therapy, pharmacologic treatment, and angiography and embolization. Complete surgical removal with a wide margin is the mainstay of treatment; however, total excision might be either impossible or associated with unacceptable morbidity in some cases[1]. In such cases, combined medical treatments with corticosteroids and vincristine have been attempted with varying degrees of success[1,4].

Although any area of the body can be affected by KHE, the trunk and the proximal areas of the extremities are most commonly involved[3]. Only 1 adult case of KHE in the tongue has been reported[5]. To the best of our knowledge, ours is the second adult case of KHE in the tongue.

CASE DESCRIPTION

An otherwise healthy 38-year-old man with no medical problems and not taking any prescription medications noticed a small growth under the left side of his tongue. The growth increased in size slowly over a period of 6 months and was associated with some minor bleeding related to trauma and with tenderness. There were no associated symptoms of deviation of the tongue, change in taste, numbness, dysphagia, odynophagia, dysphonia, or shortness of breath. The patient did report a lifelong history of slight articulation problems with the letters T and S.

Physical exam was notable for a firm, tender 2.5-cm submucosal mass on the left ventral oral tongue. Magnetic resonance imaging showed an ill-defined enhancing lesion in the left sublingual area, measuring 1.2×2.9×3.4 cm. Involvement of the hypoglossus and the mylohyoid muscles was evident (Figure 1). Mass effect on the left genioglossus muscle was present. No lymph nodes were enlarged.

An incisional biopsy of the mass at our institution showed KHE. The slides were also sent to a tertiary care centre, where the pathology was confirmed. Tumour was

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composed of irregular vascular lobules that infiltrated soft tissue in glomeruloid structures (Figure 2). Slit-like vessels were present. Large lymphatic vessels were present adjacent to the tumour. In addition to the perineural invasion, tumour was found to be infiltrating into the striated muscle. No necrosis was seen. The tumour was composed of tightly coiled CD31-positive capillary vessels invested with actin-positive pericytes. Immunohistochemical staining for CD31 was focally positive, and CD34 was strongly positive (Figure 3). The platelet count in our patient was 188,000/mL, with no evidence of life-threatening hemorrhage and lymphangiomatosis, suggesting an absence of KMP.

The patient underwent surgical resection of the mass at our institution. After the mucosa had been incised, electrocautery was used to resect the tumour from the surrounding tongue musculature. Given the benign nature of the lesion, no attempt was made to secure wide surgical margins. Although the lesion had no discrete capsule, the feeling was, based on visual inspection and palpation, that it had been completely excised. The mass was noted to be very vascular, requiring distal ligation of the lingual artery. The final pathology confirmed the diagnosis of KHE.

Postoperatively, the patient has recovered completely, with minimal residual post-operative dysarthria, which has been improving with time. Plans are to obtain magnetic resonance imaging at 6 months after surgery for a baseline and to follow this patient with serial examinations.

LITERATURE REVIEW

The PubMed database was used to identify all published case studies of KHE in adults as of 2 April 2016. The search was performed using the main keyword "Kaposiform hemangioendothelioma." Studies were selected for inclusion if they reported data on KHE in adults (≥18 years of age). We also searched the bibliographies of selected papers to identify relevant case studies that might have been missed during the primary search.

DISCUSSION

Kaposiform hemangioendothelioma occurs predominantly in infants and adolescents; only 26 cases of KHE in adults have been reported to date, as shown in Table I. Of those 26 patients, 13 were men, and 11 were women (in 2 cases the sex of the patient was not reported), with ages ranging from 19 to 81 years. The most common tumour locations were the extremities, the torso, and the head–neck region. None of the 26 cases showed evidence of KMP. Recurrence was reported in 3 patients after their respective treatments. Only 2 patients developed lymphangiomatosis.

The unusual age at presentation and the involvement of tongue as the primary tumour site are the two aspects of our case that make it unique. Only 1 case of KHE of the tongue—White *et al.*[5]—has been reported in the literature. However, the patient described by those authors was 21 years of age and had associated comorbidities of hypertension and systemic lupus erythematosus with chronic renal failure. In contrast, our patient was 38 years of age and had no associated medical problems.

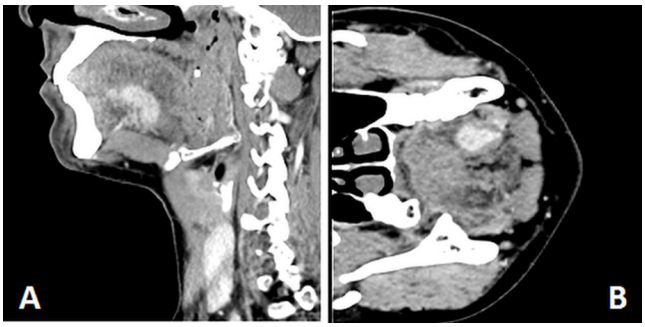

FIGURE 1 Images from T2-weighted magnetic resonance examination show an ill-defined enhancing mass in the left sublingual region, involving (A) the left hypoglossus and (B) the mylohyoid muscles.

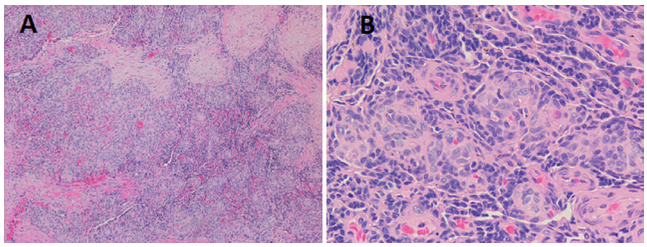

FIGURE 2 (A) Low-power microscopy illustrating large, irregularly shaped tumour nodules infiltrating into soft tissue, with striking desmoplasia. Hematoxylin and eosin stain, 20× original magnification. (B) High-power view of glomeruloid clusters of vessels in Kaposiform hemangioendothelioma, a signature feature of the lesion. 20× original magnification.

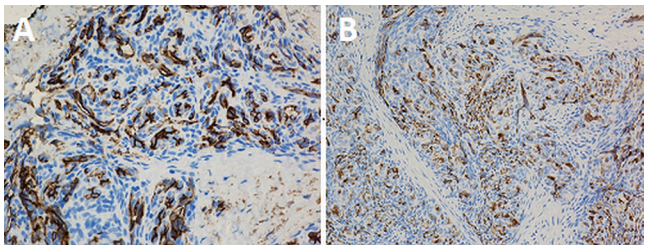

FIGURE 3 (A) Low-power microscopy shows tumour expression of CD34, a vascular marker. Immunohistochemical stain, 20× original magnification. (B) Lesion is composed of tightly coiled, CD31-positive capillary vessels surrounded by pericytes. 20× original magnification.

Pathologically, the two important differential considerations in KHE are capillary (cellular) hemangioma and Kaposi sarcoma. Capillary hemangioma usually does not display spindling of the cells, nor does it contain the signature glomeruloid structures of KHE. Moreover, unlike hemangioma, KHE has to be followed very closely for local recurrence and growth. Kaposi sarcoma, on the other hand, is characterized by uniform spindling of the cells and often a striking inflammatory infiltrate peripherally. In contrast, KHE shows much greater variation from area to area. Human papillomavirus 8 has not been associated with KHE, and staining in the present case was negative as well.

TABLE I Clinical characteristics and outcomes for adult patients with Kaposiform hemangioendothelioma reported in the literature

Reference	Age	Sex	Tumour location	KMP	Treatment	Recurrence
Mentzel *et al.,* 1997[6]	64	Male	Chest wall	No	Not reported	Not reported
	35	Male	Groin	No	Not reported	Lymphangiomatosis
	48	Female	Neck	No	Not reported	Not reported
Zamecnik *et al.,* 2000[7]	37	Male	Abdominal wall	No	Excision	Not reported
Mac-Moune Lai *et al.,* 2001[8]	33	Female	Arm	No	Excision	Yes
Cooper *et al.,* 2002[9]	26	Female	Thigh	No	Excision	No
Hardisson *et al.,* 2002[10]	27	Male	External auditory canal	No	Excision, radiation	Yes
El-Sayed and Ramadan, 2004[11]	22	Male	Cervical lymph node	NR	Not reported	Not reported
	35	Female	Chest wall	NR	Not reported	Not reported
Lyons *et al.,* 2004[12]	19	Female	Forearm	No	Excision	No
	20	Female	Leg	No	No treatment	No
Bienaime *et al.,* 2006[13]	72	Male	Shoulder	No	Not reported	Not reported
Cho and Na, 2006[14]	62	Male	Conjunctiva, upper eyelid	No	Prednisolone	No
Senturk *et al.,* 2006[15]	58	Male	Shoulder	No	Excision	No
Vetter-Kauczok *et al.,* 2008[16]	36	Female	Chest	No	Excision	Lymphangiomatosis
Karnes *et al.,* 2009[17]	49	Male	Upper arm	No	Excision, chemotherapy	No
White *et al.,* 2009[5]	21	Male	Tongue	No	Excision	No
Kim *et al.,* 2011[18]	32	Female	Breast	No	Excision, mastectomy	Yes
Yu and Yang, 2011[19]	36	Female	Spleen	No	Excision, chemotherapy	No
Costa and Folpe, 2013[20]	39	Male	Testis	No	Excision	No
	81	Male	Testis	No	Excision	No
Croteau *et al.,* 2013[21]	48	NR	Upper extremity	No	Yes	Not reported
	25	NR	Upper extremity	No	Yes	Not reported
Wu *et al.,* 2013[22]	21	Male	Pleura	No	Excision	No
Wang *et al.,* 2014[2]	51	Female	Ilium	No	Radiotherapy	No
Wong *et al.,* 2014[23]	46	Female	Paranasal sinus	No	Excision	No

KMP = Kasabach–Merritt phenomenon; NR = not reported.

The optimal management of KHE is governed by several factors: accessibility to surgical excision, location (cutaneous vs. visceral), size of the tumour, and whether the patient has lymphangiomatosis and KMP. In our case, surgical excision was the treatment of choice because of a well-defined mass that was surgically accessible in a patient with no associated comorbidities and an absence of KMP.

SUMMARY

This very rare case of KHE of the tongue occurring in an otherwise healthy man was managed successfully by surgical excision. To the best of our knowledge, the patient described in the present report represents the 2nd confirmed case of KHE of the tongue in an adult.

ACKNOWLEDGMENTS

This study was funded by Cancer Treatment Centers of America.

CONFLICT OF INTEREST DISCLOSURES

We have read and understood *Current Oncology*'s policy on disclosing conflicts of interest, and we declare that we have none.

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