Kimura's disease of the tongue: a new case report

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Kimura's disease (KD), a chronic inflammatory disease of uncertain etiology, manifests as a painless subcutaneous swelling mostly in the head and neck region that involves major salivary glands and regional lymph nodes. To date, the majority of cases of KD have been documented in Asian men aged 20–40 years. However, the number of reported cases of KD involving the oral and maxillofacial area is limited, and since the masses appear similar to cysts or benign tumors, the establishment of an accurate preoperative diagnosis is challenging. The accurate diagnosis of KD is considered to require surgical excision followed by histopathological examination. In this case report, we present a case of KD affecting the ventral surface of the tongue, a unique site of the disease that is not reported earlier according to our knowledge.

Keywords:
angiolymphoid hyperplasia, kimura, rare tongue lesion

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Introduction

Kimura's disease (KD) is a benign, rare, chronic inflammatory disorder of unknown etiology affecting mainly the middle-aged Asian people. However, sporadic cases have been also found in the non-Asian population. It was first described in 1937 by Kim and Szeto (China) as 'Eosinophilic Hyperplastic Lymphogranuloma.' Kimura *et al*. [1] reported it in the literature titled 'On the unusual granulation combined with hyperplastic changes of lymphatic tissue' after which this entity became widely known as KD. The pathophysiology of KD remains unknown. It has been hypothesized that an infection or toxin may trigger an autoimmune phenomenon or lead to a type I [immunoglobulin E (IgE)-mediated] hypersensitivity reaction. Some evidence has suggested a predominance of Th2 cells in patients with KD. Additional studies have shown elevated granulocyte-macrophage colony-stimulating- actor, tumor necrosis factor-α, soluble interleukin (IL)-2 receptor (sIL-2R), IL-4, IL-5, IL-10, and IL-13. Another study indicated that the activation of the IL-21/pERK1/2 pathway is a component of KD immunopathogenesis and that pERK1/2 could be a potential prognostic indicator of the disease [2].

KD usually involves deep subcutaneous tissue and lymph nodes of the head and neck region. The clinical presentation is characterized by a triad of painless unilateral cervical lymphadenopathy, eosinophilia, and markedly elevated serum IgE levels. It presents with painless subcutaneous masses in the head and neck region. It usually involves preauricular, axillary and inguinal lymph nodes, parotid and submandibular glands with unusual sites being auricle, scalp, orbit, nerves, and the spermatic cord. Few cases of palate and cheek with eyelid affliction have been described [3,4]. Besides a patient presenting as lymphadenopathy and painful oral ulcerations was reported [5].

The common finding is within the subcutaneous tissues and lymph nodes. It can represent itself either as single or multiple lesions, with the latter being less frequent. Other lymph nodes of the face and neck area can be affected, as well as distant subcutaneous lymph nodes either as solitary or multiple lesions. Histopathologically, the disease is characterized with hyperplasia of the lymphoid tissue with well-developed lymphoid follicles, marked lymphocyte (eosinophil) infiltration, proliferation of thin-walled capillary venules, and varying degrees of fibrosis [6]. Renal involvement in the disease characterized by proteinuria (12–16%) may be seen [7].

KD is usually seen in young adults, with most patients being aged between 20 and 40 years of age; men are affected more commonly than women, with a 3 : 1 ratio [8].

Most cases of KD have been reported in Asians, and the prevalence among persons of other races is thought to be low. A retrospective review of 21 histopathologic

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specimens diagnosed as KD at the US Armed Forces Institute of Pathology found the following racial distribution: seven Whites, six African Americans, six Asians, one Hispanic, and one Arab [9]. This illustrates that if clinically suspected, KD should be included on the differential diagnosis for persons of any racial group.

Case report

A 46 year old fit and healthy male presented to the Department of Oral and Cranio-maxillofacial Surgery in April 2016 after being referred from his primary health general dentist for a nonhealing lesion of the ventral surface of the tongue; the lesion was discovered by chance about 40 days before presentation, and was followed up by his dentist for 3 weeks which he smoothened some sharp teeth thinking it was a traumatic ulcer.

On presentation the patient looks well-built obese, well nourished, and does not seem in distress, his vitals upon arrival were the following:

(1) Blood pressure: 166/89 mmHg.
(2) Heart rate 82 bpm.
(3) Temperature: 36.8°C.
(4) Respiratory rate: 18 breaths/min.
(5) SpO_2: 99%.

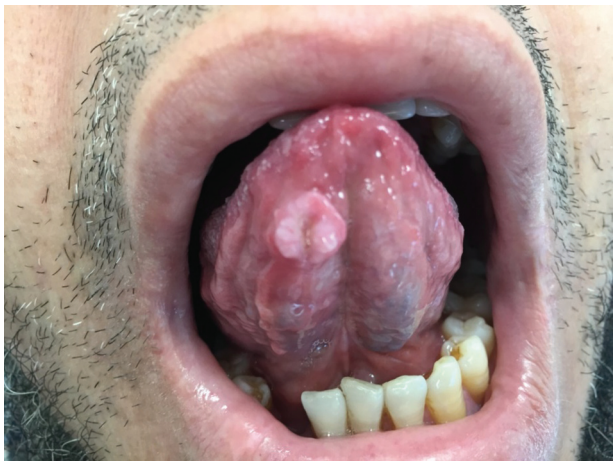
The patient is generally healthy with no chronic medical illness or previous surgeries, nonsmoker, nonalcoholic, no history of trauma, and no family history of malignancy.He complained of a painless, slowly growing mass on his ventral surface of the tongue since discovered by chance over a month, nonbleeding, and not associated with any discomfort.

Upon oral examination, a small mass of about 1 cm on the right side of the ventral surface more toward the anteromedial part of the tongue, pale in color (white-pinkish color), noninflamed, nonulcerated, nonbleeding, fixed to the underlying tissue, sessile, nontender, central groove seen dividing the mass into two halves (Fig. 1).

Oral mucosa looks normal in color and texture, no signs of dehydration and/or other mucosal lesion noted, no sharp restorations or teeth that were noted to contact the lesion, normal salivary flow, and general oral hygiene seems to be fair.

Facial examination showed no skin lesions or any functional deficits; cervical examination was unremarkable for any palpable lymphnode.

Figure 1



Ventral surface of the tongue mass on presentation.

The patient was consented for an excisional biopsy which was performed under local anesthesia and the obtained mass was sent for histopathological examination.

Microscopic description

Stratified squamous mucosa revealing hyperplasia

The submucosal connective tissue shows dense fibrous stroma heavily infilterated by mixed chronic inflammatory infiltrate comprising lymphocytes, histiocytes, and scattered eosinophils. Occasional lymphoid aggregates are also identified together with prominence of small blood vessels. The proliferative blood vessels are lined by plump-appearing (epithelioid) endothelial cells. No fungal elements, viral inclusions, dysplasia, or malignancy were seen (Figs 2–5).

Diagnosis

Vasculoproliferative lymphohystiocytic-rich lesion, consistent with KD.

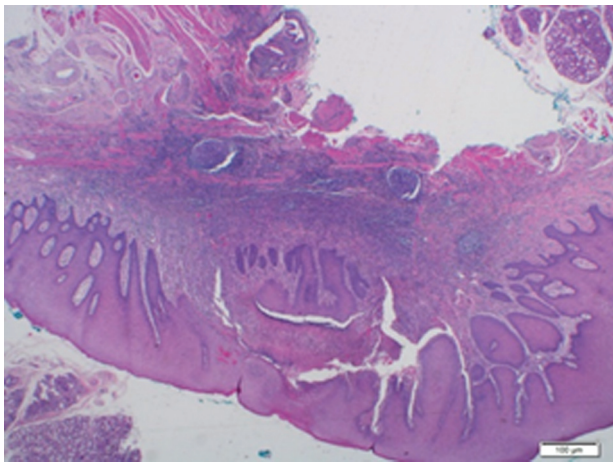
A follow-up was arranged after 10 days for reviewing the surgical site and sutures removal, in addition informing the patient of the histopathology findings.

On later follow-up, the patient was seen after a month, then seen after 9 months with no new complains, site was healing well, no discomfort, or any sign of recurrence (Fig. 6).

Discussion

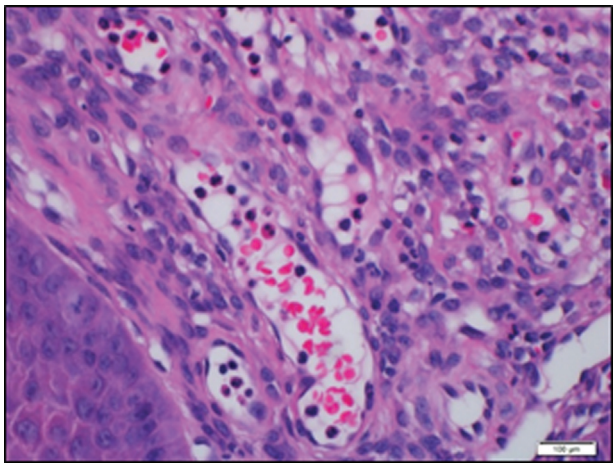
KD presents as a benign tumor-like lesion, rare chronic inflammatory disorder with angiolymphatic proliferation. The diagnosis of KD can be based on

Figure 2



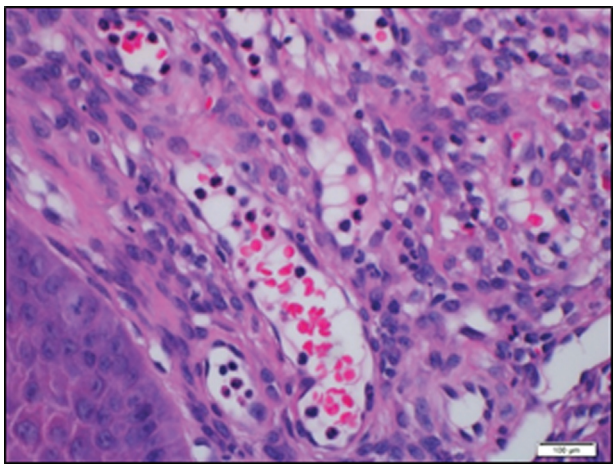
Tongue biopsy shows hyperplastic stratified squamous mucosa with underlying dense chronic inflammatory infiltrate with occasional lymphoid aggregates (H&E, ×2).

Figure 3



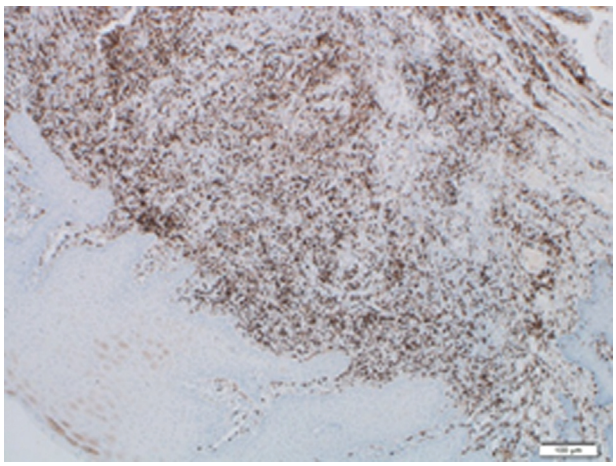
Submucosal lymphovascular proliferative lesion composed of channels of small blood vessels surrounded by dense chronic inflammatory.

Figure 4



The small vascular spaces are lined plump-appearing endothelial cells (H&E, ×40).

Figure 5



CD45 immunostaining that highlights the submucosal dense lymphocytic infiltrate.

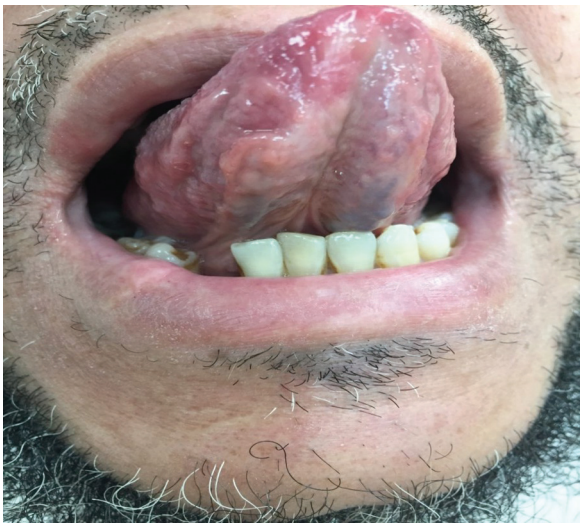
characteristic findings following surgical excision in conjunction with peripheral eosinophilia and elevated serum IgE levels [10].

Differentiating KD from angiolymphoid hyperplasia with eosinophilia requires a strict analysis of clinical and histologic features because the diseases are similar and were once thought to be of the same disorder. Both the diseases usually present with soft tissue masses in the head and neck region, but in angiolymphoid hyperplasia with eosinophilia, the lesions are mostly dermal or subcutaneous and not often in lymph nodes, which is a common location for Kimura's lesions. Angiolymphoid hyperplasia with eosinophilia is more typically seen in middle-aged women and KD in younger men.

The etiology and the pathogenesis of KD are still unknown but various suggestions have been put forward by many investigators. The disease is classified as a benign reactive process. Allergic reactions, infections, and autoimmune reactions with an aberrant immune reaction have been suggested in its etiopathogenesis [11]. A viral or parasitic trigger that may alter T-cell immunoregulation or induce a type I hypersensitivity may result in the release of eosinophiliotrophic cytokines.

The usual methods of treatment include observation, drug administration, immunosuppressive therapy including tacrolimus, radiotherapy, surgical excision, and photodynamic therapy. Other treatment modalities are intralesional administration of

Figure 6



Patient after 9 months postexcision of the lesion.

steroids, cytotoxic agents, and electrodesiccation. All-transretinoic acid with low-dose prednisone induced remission in one case. Pranlukast, a leukotrine receptor antagonist, and cetrizine, an H1 receptor blocker, were also effective in inducing clinical remission in a few cases.

Among them, surgical excision is considered the most effective as it provides not only a treatment for KD, but also a sample for histopathological investigations in order to facilitate an accurate diagnosis. However, treatment plan should aim to preserve at preservation of vital structures associated with the lesion and cosmetic rehabilitation, while preventing recurrences. If the lesion recurred after an excision and shows systemic involvement then use of medication like corticosteroids and immunosuppressive agents have been shown to reduce the size of lesions; irradiation should be considered in patients resistant to steroid or to prevent patients from deleterious effect of long-term use of steroids.

In the present case, a histopathological examination of the diseased tissue was conducted in order to investigate the potential diagnosis of KD affecting the oral and maxillofacial areas and to rule out early malignancy. The examination showed vigorous proliferation of vasculature and lymphocytic infiltration enriched with eosinophils, which may be considered as the gold standard for KD diagnosis.

The recurrence of disease is affected by various factors, including disease duration time, lesion diameter, blood eosinophil count, well-defined lesion boundaries, serum IgE levels, and single or multiple lesions.

In the current case, a single well-defined lesion was seen, which was less than 2 cm in diameter, less than 2 months duration, and was totally excised. However, no sign of recurrence of the mass has yet been observed on the basis of the follow-up interviews.

Conclusion

The diagnosis of KD was based on histopathological report though not initially thought of while planning surgery. The treatment mainly aims to preserve function and cosmetics while keeping in view the recurrence and the sequel. This case is reported to make the surgeons aware of such rare occurrence which may be borne in mind for a complete cure. The site of such case was not documented in the literature to our knowledge.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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