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# Kimura disease

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## Abstract

Kimura disease is a rare, benign, chronic, immune-mediated inflammatory disorder. We report a 46-year-old man who presented with a cutaneous nodule behind his left ear. Surgical removal of the growth confirmed the histological diagnosis of KD. There was no recurrence found after 3 years follow-up.

Keywords: Kimura disease

## Introduction

Kimura disease (KD) is a rare, benign, chronic immune-mediated inflammatory disorder, which was first described in 1938 by Kimura and Szeto [1]. The specific mechanism is unknown. However, allergic reaction, *Candida* infection, arthropod bite, deregulation of eosinophil dynamics and IgE synthesis, and altered systemic immune-mediated reaction have all been postulated as causative [2]. The condition is characterized by a painless mass, usually on the head and neck, with regional lymphadenopathy, blood and tissue eosinophilia, and elevated serum immunoglobulin E levels. There is still no definitive treatment for this disease; surgery, thalidomide, cyclosporine, interferon- $\alpha$ , omalizumab (an anti-IgE antibody), corticosteroids, and radiotherapy have been recommended [3, 4], but none are clearly standard of care. Rates of recurrence may be as high as 62% [5]. We present a case of Kimura disease treated with surgery.

## Case Synopsis

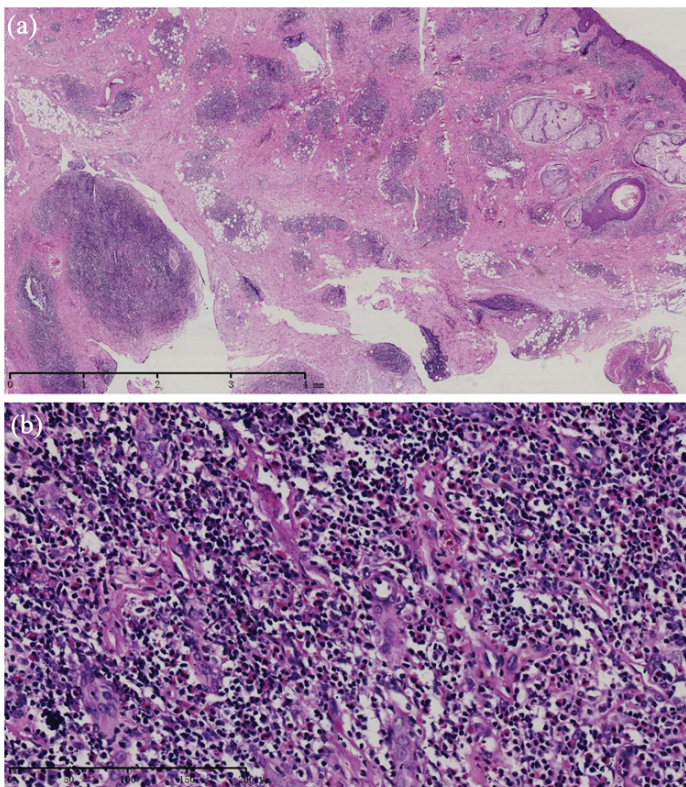
An otherwise healthy 46-year-old man was referred to our department. He exhibited a cutaneous



**Figure 1.** Cutaneous nodule behind the left ear.

nodule behind his left ear. The lesion appeared to be asymptomatic and had increased in size and erythema in the past 4 years. The patient had received a diagnosis of a lymphoma at another hospital before, but without pathologic confirmation and no treatment had been attempted.

Physical examination revealed a 3cm  $\times$  3cm erythematous cutaneous nodule behind the left ear (**Figure 1**). The lesion was soft and painless on palpation. Laboratory examination showed WBC of  $9.86 \times 10^9/L$ ; eosinophil count was  $1.78 \times 10^9/L$  (normal is  $0.02 \sim 0.50 \times 10^9/L$ ). The percentage of eosinophils was 18.1% (normal is 0.5-5%). Serum total IgE was elevated. An ultrasound examination showed a hypoechoic subcutaneous 22mm  $\times$  9mm nodule whose boundaries were unclear. The erythrocyte sedimentation rate, renal, liver, and thyroid function tests, creatine phosphokinase, and lactic dehydrogenase were normal. Treponema pallidum hemagglutination assay, venereal disease



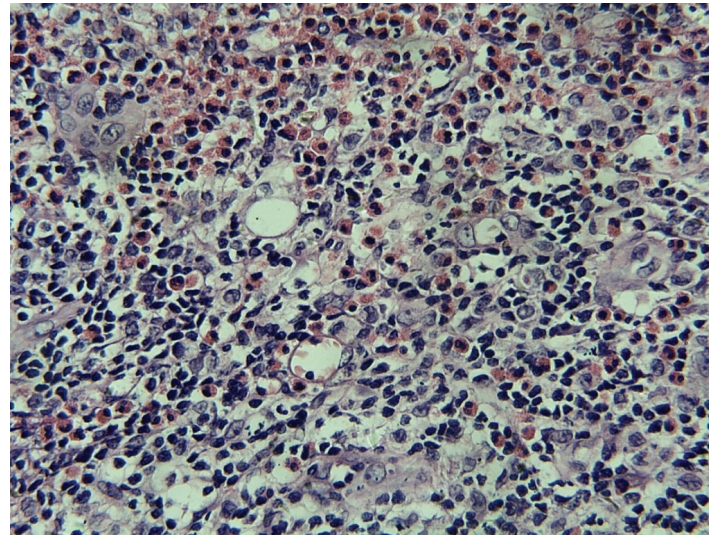
**Figure 2.** Biopsy was taken from the lesion showing dense lymphocytes and eosinophil infiltration at the dermis and subcutaneous tissue with visible lymphoid follicles formation.

research laboratory test, human immunodeficiency virus serology, hepatitis A, B, C tests, antinuclear antibodies, rheumatoid factor, complement serum C3 and C4 levels, antimitochondrial and anti-smooth muscle antibodies, toxoplasmosis titer were all normal or negative. Chest radiograph was normal.

Histopathological examination demonstrated dense lymphocyte and eosinophil infiltration of the dermis and subcutaneous tissue with visible lymphoid follicles and histiocytosis (**Figure 2**). The infiltrate consisted of plasma cells, lymphocytes, and numerous eosinophils, which formed occasional eosinophilic abscesses. The background showed vascular proliferation with plentiful thin-walled postcapillary venules (**Figure 3**). Based on the clinical and histologic findings, the diagnosis of KD was made. After surgical removal, no recurrence was found during the follow-up of 3 years.

### Case Discussion

This quite rare condition is primarily seen in Asian males in their 2nd to 4th decade of life (70-80%). The histological characteristics of Kimura disease



**Figure 3.** Lymphocytes, plasma cells and numerous eosinophils showing vascular proliferation. (H&E, 400x)

are eosinophilic tissue infiltration, lymphocyte follicular hyperplasia, fibro-collagenous deposition, and vascular proliferation. Blood eosinophilia and elevated serum IgE level are important markers in Kimura disease. Owing to frequent mass localization in the parotid gland region and accompanying lymphadenopathy, Kimura disease may be easily mistaken for a malignant disorder, including T-cell lymphoma, Kaposi sarcoma, Hodgkin disease, or parotid tumor.

Kimura disease is often confused with angiolymphoid hyperplasia with eosinophilia (ALHE), which is now considered as an inflammatory vascular proliferation of epithelioid morphology, whereas KD is considered to be a lymphoproliferative disease with a longer duration [6, 7]. Other conditions in the differential diagnosis of KD include vascular lymphoid hyperplasia with eosinophilia, pyogenic granuloma, epithelioid hemangioendothelioma, hamartomas, lymphomas, insect bites, tuberculosis, nodal metastasis, Warthin tumor, and low-grade angiosarcoma [8, 9]. Surgical treatment is currently the preferred and most widely used treatment for KD. Surgery and postoperative corticosteroid therapy is commonly used as a treatment method. In addition, radiation therapy, cryotherapy, and cytotoxic therapy are utilized in KD. KD often relapses after surgery but other adjuvant therapies have more side effects. Recent reports show that smoking, additional systemic diseases, surgery alone, and surgery followed by oral corticosteroids were associated with a poor prognosis of Kimura

disease [10].

## Conclusion

KD is a rare condition; the diagnosis of KD is based on both clinical and histologic findings. Rates of recurrence of KD following treatment may be up to 62% [5]. In our case, although no recurrence has occurred over 3 years, further follow-up is required.

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