

# Kimura Disease: Report of a Rare Case

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## Abstract

Kimura disease is a rare entity causing subcutaneous swellings and lymphadenopathy, with hardly 120 cases reported worldwide. It is mainly seen in Asian men. Herein, we present a typical case that presents with submandibular mass and persistent eosinophilia. The patient was a 38-year-old man suffering from left submandibular mass for the last 5 years. It has happened upon a dental infection. Treatment with prednisolone resulted temporary decrease in the size of the lesion, but it wouldn't disappear totally. Since the swelling did not get improved, an excisional biopsy was conducted. The high rate of the eosinophil and histopathology results conducted us to diagnose him with Kimura.

Histologically, Kimura disease presents as lymphadenopathy with preserved lymph node architecture and reactive and prominent germinal centers. Dense eosinophilic infiltration of the interfollicular zones, lysis of the follicles, and occasionally microabscesses are seen. Granuloma formations contain infiltration of eosinophils, lymphocytes, plasma cells, and histiocytes. Tissue fibrosis, sclerosis, and vascular proliferation are also present. Vessels remain thin-walled with cubical endothelial cells. This was in line with our patient symptoms.

**Keywords:** Kimura disease, Neoplasms, Lymphadenopathy, Case report

## Introduction

Kimura disease (KD) is a chronic inflammatory disorder typically manifesting as large subcutaneous masses on the head or neck of Asian males.<sup>1</sup> The first report of KD was from China in 1937, in which Kimm and Szeto described seven cases of a condition they termed "eosinophilic

hyperplastic lymphogranuloma".<sup>2</sup> The disorder received its current name in 1948, when Kimura et al. noted the vascular component and referred to it as an 'unusual granulation combined with hyperplastic changes in lymphoid tissue'.<sup>3</sup>

The pathophysiology of KD has

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still remained unclear. It has been hypothesized that toxins and infections can trigger autoimmune diseases or can result in type I hypersensitivity reaction. Some evidence has suggested a predominance of TH 2 cells in patients with KD.<sup>4</sup> Additional studies have shown elevated granulocyte macrophage-stimulating factor (GM-CSF), tumor necrosis factor- $\alpha$  (TNF- $\alpha$ ), soluble IL-2 receptor (sIL-2R), IL-4, IL-5, IL-10, and IL-13.<sup>5</sup>

Patients with KD usually demonstrate an enlarged lymph node with no pain or general lymphadenopathy. It typically presents a subcutaneous swelling in the head and neck region, particularly in the preauricular and submandibular area. It is occasionally associated with local/distal lymphadenopathy, peripheral eosinophilia, and an increased IgE level.<sup>6</sup>

### Case Presentation

A 38-year-old man suffering from left submandibular mass visited the ward and declared that from about 5 years ago (2015) upon a dental infection he has been experiencing swelling in the aforesaid location as it can be seen in figure 1.

As a result of prednisolone treatment, the size of the lesion had temporarily decreased but it didn't disappear completely. Since the swelling never ameliorates totally, an excisional biopsy was conducted and KD was diagnosed. The percentage of the eosinophil and eosinophil count had been high in the last 5 years and still was high (about 30%) when the patient visited the ward. After receiving treatment with prednisolone, the count was 4% and the size of the mass decreased substantially. Histopathology revealed extensive eosinophilic infiltration consisting of mature eosinophils in follicular and interfollicular areas and marked hyperplasia of the germinal centers. These histological features were consistent with KD.

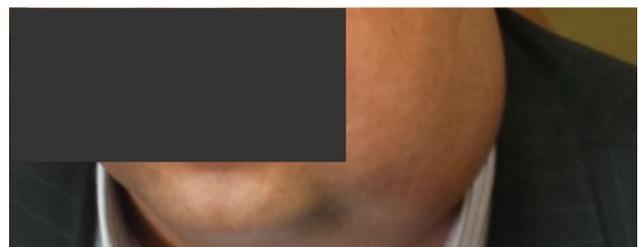
### Discussion

KD is a rare chronic inflammatory disorder typically presenting as a painless mass or masses

in the head and neck region, with occasional pruritus of the overlying skin. Renal disease, nephrotic syndrome, in particular, is present in up to 20% of patients with KD.<sup>10</sup> Less commonly, several reports in the literature have linked KD with a hypercoagulable state in patients without associated nephrotic syndrome.<sup>7</sup>

Our case presented with painless submandibular mass, without any renal involvement or coagulopathy. Histologically KD presents as lymphadenopathy with preserved lymph node architecture and reactive and prominent germinal centers. Dense eosinophilic infiltration of the interfollicular zones, lysis of the follicles, and occasionally micro abscesses are seen. Granuloma formations contain infiltration of eosinophils, lymphocytes, plasma cells, and histiocytes. Tissue fibrosis, sclerosis, and vascular proliferation are also present. Vessels remain thin-walled with cubical endothelial cells. Rarely the features include progressive destruction of germinal centers, presence of poly karyocytes (which are not pathognomonic for the disease). Immunofluorescence tests show germinal centers containing heavy IgE deposits and variable amounts of IgG, IgM, and fibrinogen.<sup>8</sup>

There is no consensus on the management aspects of KD so far. However, primary prophylactic surgery is performed as a therapeutic and/or diagnostic procedure. Conservative treatment includes oral steroids which are reported to be responsible for decreasing the size of the enlarged lymph nodes but there is no evidence of reduction of the affected salivary gland size. Furthermore, the lesions usually get enlarged again when steroid treatment is terminated. Thus, successful treatment is mainly reassured by a constant low dose of steroids.<sup>9</sup> Another positive



**Figure 1.** The patient presented with left submandibular mass for about 5 years.

effect of steroid treatment is that it decreases renal symptoms as well.

Remissions reach 25% in groups of patients treated surgically. Surgery and following subsequent steroid treatment are proposed as an alternative treatment. Radiation therapy is useful to control lesions that are not responsive to steroids or with relapse after surgery. The effective total dose of radiation is proved to be 20-30 Gy.<sup>10</sup> We treated this patient with prednisolone and conducted a gradual tapering. We continue administering a low dose of prednisolone to him.

### Informed Consent

The patient signed a written informed consent agreement to publish the data.

### Conflict of Interest

None declared.

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