

# Rare and Commonly Missed Benign Medical Disease - Kimura

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Received: 01 April 2023

Accepted: 01 May 2023

Published: 08 May 2023

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**Keywords:** Kimura disease, angiolymphoid hyperplasia with eosinophilia, IgE

## 1. Introduction

Kimura's disease is a rare chronic inflammatory disorder characterized by multiple painless solitary head and neck lymphadenopathy often accompanied by peripheral eosinophilia and elevated serum IgE. It is a benign condition with unknown etiology, usually affecting young men of the Asian race. Affected Caucasians are very rare.

This quite rare condition is found almost exclusively in Asian individuals in their 2nd to 4th decade of life mostly in males (70–80%) [1, 2]. The etiology is unknown, however, an allergic reaction or an alteration of the immune system is taken into consideration. Persistent antigenic stimulation

following arthropod bites and parasitic or *Candida* infection are also suspected [3].

Initially, the lesion was described as a neoplasm. Very rare cases of Kimura's disease are reported in Caucasian individuals in Europe, the United States of America, and Australia. The diagnosis of Kimura's disease can be very difficult and misleading; it is important not to ignore histopathological features.

## 2. Case Presentation

A 30-year-old male was admitted to GBH General Hospital in February 2023 with chief complains of swelling (painless) in the neck and retroauricular area and itching since one and a half years (**Figure 1**).



**Figure 1:** A 30-year-old male with swelling (painless) in the neck and retroauricular area and itching since one and a half years.

On examination, the swelling was painless, non-tender, and not relieved by taking medication localized to the neck. H/O similar complaints one year back in the postauricular area for which FNAC was done S/O atypical lymphoproliferative disorder and it was operated then.

FNAC from the swelling in the right neck was done which was suggestive of reactive lymphadenitis, suggestive of Kimura's disease.

The patient also complained of itching all over the body which was gradual in onset and progressive in nature and it is associated with redness.

Blood examination on first presentation on 16th June 2021 showed Hb - 13.60 g/dl, TLC - 12.40 (eosinophilic -14.50 raised), platelet count - 299, PBF: normocytic

normochromic RBCs anisocytosis seen. There is leucocytosis with neutrophilia.

SGPT - 51, SGOT - 37, ESR - 14, LDH - 372, USG of the neck - few subcentimetric to enlarged right intraparotid submandibular, upper and mid cervical lymph nodes with fatty hilum are seen.

MRI neck: features suggestive of right posterior auricular soft tissue lesion with neck lymph nodes as described likely malignant/infective etiology.

8th February 2023: total IgE - 1585, absolute eosinophilic count - 1333, BMI - 38 (obese) (Table 1).

Date	Investigation	Treatment
16/6/2021	Hb-13, TLC-12.40/cumm, DLC-lymphocytes-22.80, monocytes-5.10, eosinophils-14.50, basophils-0.30, PC-299	Tab celecoxib 200mg 1BD Tab multivitamin 1OD
8/2/2023	Hb-14, TLC-11/cumm, DLC-lym-20, monocytes-6, eosinophils-16.50, basophila-0.40, PC-300, total IgE-1585, absolute eosinophilic count- 1333, S. triglyceride-210	Tab leflunomide 10mg OD, Tab Vit C 500mg OD, Tab deflazacort 18mg OD, Tab Vit D3 once a week

Table 1: Blood examination reports.

### 3. Discussion

Kimura's disease is a chronic inflammatory condition presented as painless solitary or multiple subcutaneous nodules, asymmetric, mostly in the head and neck region, with coexisting lymphadenopathy in 30–40% of the cases [1].

Typical areas for the nodules are the preauricular, submandibular, and popliteal regions, oral cavity, larynx, and parotid glands [4, 5]. They are rarely reported in other localizations like eyelids, lacrimal glands, orbit, axilla, groin, forearm, and kidneys [1, 2, 6–8].

Kimura's disease may affect the kidneys in up to 60% of patients. In those cases, it may present itself as almost all types of glomerulonephritis or as a nephritic syndrome (12%) [2, 5].

Hypereosinophilia and elevated serum IgE are found in Kimura's disease as well.

Kimura disease may be easily mistaken for a malignant disorder (acute lymphocytic leukemia, T-cell lymphoma, Kaposi sarcoma, Hodgkin's disease, or parotid tumor) because of a mass localized in the parotid gland and accompanied by lymphadenopathy.

That is why differential diagnosis should be performed very carefully taking into account all clinical and histological findings [2, 3, 9]. Differential

diagnosis between Kimura's disease and angiolymphoid hyperplasia with eosinophilia (ALHE) has been a challenge for a long time. They were considered to be variations of the same disease making the diagnostic process very complicated.

Histologically Kimura's disease presents as preserved lymph node architecture with 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 present. Rarely, the features include progressive destruction of germinal centers and the presence of polykaryocytes (which are not pathognomonic for that disease). Immunofluorescence tests show germinal centers containing heavy IgE deposits and variable amounts of IgG, IgM, and fibrinogen [1–3].

As previously mentioned, Kimura's disease (nodules localized in the head and neck, accompanied by lymphadenopathy) is often confused with angiolymphoid hyperplasia with eosinophilia (ALHE) that predominantly affects middle-aged Caucasian females.

Histological differences are not distinct. Blood vessel walls containing hypertrophied and sometimes

vacuolated endothelial cells with eosinophilic cytoplasm and rarely atypical nuclei never appear in Kimura's disease. ALHE is now considered a type of histiocytoid hemangioma.

Other differences between those two diseases are presented in **Table 1** [3, 10].

Ultrasound imaging of the salivary glands and neck should be the first test that is performed in case of lymphadenopathy. Lymph nodes in Kimura's disease are hypoechoic, solid, and round or oval in the parotid and submandibular areas with normal surrounding soft tissues.

On radiological examination, Kimura's disease mimics other chronic and malignant diseases such as tuberculosis or lymphoma. In case of a mass in the major salivary gland, the differential diagnosis should include the following: adenocarcinoma, adenoma, and metastatic lesions. Despite the similar radiological characteristics, in the case of Kimura's disease, it is not possible to exclude malignancy and thus diagnosis cannot be based exclusively on imaging.

Histological confirmation is necessary. Preoperative USG, CT, and MRI are useful in demonstrating salivary gland involvement and localization of abnormal lymph nodes and to assist in guided biopsy [11, 12].

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