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# Kimura disease –A rare case report

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**Abstract**

Kimura disease is a rare chronic inflammatory disorder of unknown etiology and mainly observed in young males. The disease is presented as multiple painless subcutaneous swelling, blood and tissue eosinophilia with raised IgE levels. Early diagnosis of Kimura's disease is important so that unnecessary invasive diagnostic procedures can be avoided. Here I have documented a case of Kimura disease in a 16-year-old male presenting with left pre auricular as well as external ear swelling.

**Keywords:** Kimura Disease, Eosinophilia, IgE

## Introduction

Kimura's disease is a rare benign chronic inflammatory disease of unknown etiology. It commonly presents as painless lymphadenopathy or multiple subcutaneous swellings in head and neck region. The first case report of this rare disease was from China in 1937.

Seven cases of a condition were described by Kimm and Szeto and termed as ''eosinophilic hyperplastic lymphogranuloma'' [1].

Further in 1948 the disease received its current name, when Kimura et al. noted the vascular component and referred to it as an "unusual granulation combined with hyperplastic changes in lymphoid tissue" [2]. The exact prevalence of Kimura's disease is still not known. Most cases of this rare disease are reported in East and Southeast Asia and small number of cases reported in Europe as well. Male to female ratio ranges from 3.5:1 to 9:1 in most of the cases reported. There have been case reports from various parts of India.

The commonly involved sites are periauricular region, groin, orbit, and eyelids. Peripheral blood eosinophilia and elevated serum immunoglobulin E (IgE) levels are the most constant features of Kimura's disease [3]. The diagnosis of Kimura's disease is usually difficult, and the biopsy or excision of the involved mass for a pathological study is needed. Here I have presented a

case which developed the swelling in front of the ear and over the external ear with no prior history of surgery.

**Case report**

A 16-year-old male patient presented to the Outpatient Department (OPD) with the complaint of swelling in front of the left ear for the past 3 years which was insidious in onset and gradually progressive in size. There was no associated neck swelling, puffiness or swelling around eyes or generalized body edema. Occasionally he had itching over the swelling. On examination, a 5x3cm lobulated, non-tender, firm bilobed mass was seen in left preauricular region involving the tragus as well. Skin overlying the swelling was normal and there was no local rise of temperature. The edges of the swelling were clearly defined with no visible pulsations seen over the surface. It was non fluctuant, non-reducible and compressible. Otoscopic examinations revealed normal findings. There was no other significant finding on systemic or local examination. The medical history of the patient did not reveal any significant history.

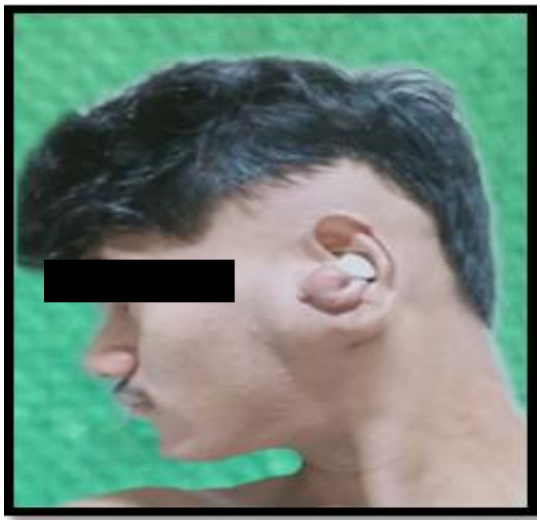

Figure 1: Clinical photo of patient showing swelling over left preauricular region and external ear.

On investigating, his Haemogram revealed mild leucocytosis with 12.5% eosinophils and an absolute eosinophil count of 1358 cells/mm3. The erythrocyte sedimentation rate was 28 mm 1st hour. Biochemical parameters including plasma glucose, kidney function test and liver function tests were found to be within normal range. Urine routine and microscopic examination also showed no abnormality. Chest radiograph findings were normal.

For pathological investigations fine needle aspiration (FNA) of the mass was performed. Smears were found to be more of a cellular type and composed of polymorphous population of lymphoid cells, histiocytes and many endothelial cell clusters. It also showed abundant eosinophils, lymph histiocytes, mixed inflammatory cell infiltrate, fibrous stroma. (Figure 2). Reports were suggestive of chronic non-specific lymphadenitis with eosinophilia. Further excisional biopsy of the mass was advised which showed reactive lymphoid hyperplasia with interfollicular vascular proliferation. Dense infiltrates of eosinophils were noted in these areas. There was no granulomas or caseous necrosis. Also, there was no evidence of parasite or malignancy. The above features were suggestive of the histopathological diagnosis of Kimura's Disease. Further correlation was advised with serum IgE levels. When investigated, serum IgE level was found to be 4250 IU/ml.

Patient was started on oral steroids and mycophenolate. The dose of steroid was then slowly tapered for 6 weeks. The swelling was significantly reduced after 8 weeks of treatment.

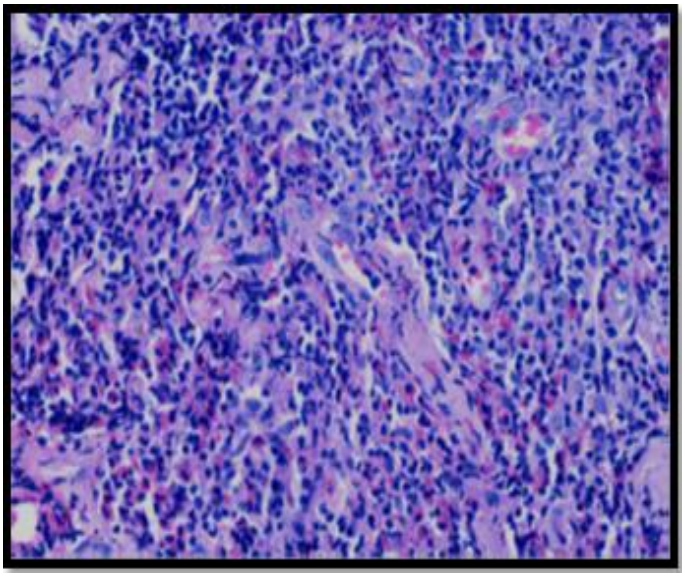

Figure 2: Interfollicular vascular proliferation with dense areas of eosinophils.

**Discussion**

Kimura's Disease is a rare chronic inflammatory disorder of unknown etiology. This disease is endemic in Asian males of middle age group. It is rarely seen sporadically in non-Asian population [4].

The exact cause and pathogenesis of Kimura's disease are still not known, although it can be considered as a self-limited allergic or autoimmune response triggered by an unknown persistent antigenic stimulus.

There are studies showing the proliferation of CD4+ T cells, specifically the CD4 T-helper2 cells and overproduction of their cytokines, like granulocyte macrophage colony-stimulating factor, tumor necrosis factor-α, IL-4, IL-5, eotaxin, and RANTES may trigger the production of lymphoid follicle and high level of IgE [5].

Nearly almost all patients with Kimura's disease demonstrate peripheral blood eosinophilia and markedly elevated levels of serum IgE.

Blood urea nitrogen, creatinine, and urinary protein levels should be obtained to exclude concomitant renal dysfunction. [6] The clinical course of the disease is benign and usually self-limiting. However, the diagnosis of Kimura's Disease can be difficult and misleading as patients with this disease are often evaluated using avoidable procedures by just not being aware of the disease.

Hence an attempt is made to put forward the clinical and investigative features of Kimura's disease, which usually pose challenge in diagnosing this rare condition. Though it is rare, one has to be aware of it because the recurrence rate is very high following any kind of treatment. The steroid resistant cases are usually treated with radiation.

**Conclusion**

The relevance of above case is due to the rare occurrence of the disease which can be commonly confused with malignancy.

Though rare, it should always be considered as one of the differential diagnosis in all the patients presenting with multiple swelling in the head & neck region and lymphadenopathy and investigated properly.

Knowledge of Kimura's disease to ENT surgeons will certainly help to diagnose and treat the disease in a better way.

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