

## Unusual Presentation of Rare Kimura's Disease - A Case Report

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**Abstract:** Facial lymphadenopathy due to rare kimura's disease diagnosed in young Indian female, with solitary, long standing, nontender, slow growing, freely mobile swelling over right cheek in mandibular region. Provisional diagnosis was tubercular lymphadenopathy based on initial cytology (FNAC) report, as features suggestive of early granulomatous lymphadenitis. So, patient started on Anti tubercular drugs, but not responded, till two months. Then excisional biopsy performed revealed enlarged lymph node and finally diagnosed as Kimura's disease based on histological features. Blood investigations showed eosinophilia and raised Immunoglobulin E levels. She is on cetirizine 10 mg once daily, no evidence of recurrence till last follow up. Kimura disease has to be considered as differential diagnosis in facial lymphadenopathy. In our knowledge of references with literatures published, this may be the first case report of kimura's disease only with unilateral facial lymphadenopathy.

**Keywords:** kimura's disease, eosinophilia, facial lymphadenopathy, lymphoid follicular hyperplasia, Immunoglobulin E.

### I. Introduction

Kimura's disease is rare, locoregional, benign, chronic inflammatory disorder of subcutaneous deep soft tissues, predilection for head and neck regions with unknown etiopathogenesis, for this aberrant immunological response to endogenous antigen is postulated. Most of patients are male oriental young adults usually presents as clinical triad of pain less subcutaneous masses, blood and tissue eosinophilia, and marked elevated immunoglobulin E levels. It may involve regional lymph nodes, salivary glands, kidneys and may be associated with asthma, rhinitis, and eczema. Diagnosis is done by histological picture consist triad of florid follicular hyperplasia with reactive germinal centers (cellular), eosinophilic infiltrates and microabscesses, sclerosis(fibro collagenous) and proliferation of post capillary venules with well formed endothelium(vascular).Surgery is main stay of treatment, other modalities include steroids, immunomodulators, antihistaminics, radiation but none proved curative. But recurrences after surgery and after stopping the medical line of treatment are present. Prognosis is good, there is no evidence till date of this disease turning into malignant.

### II. Case Report

A 22 years old lady, resident of Mysore presented to surgical outpatient with history of single, painless, slow growing swelling over right cheek since one year. There is no history of constitutional symptoms or trauma. On examination she is moderately built and nourished, swelling was 3x3 centimeters, round, nontender, smooth, firm, subcutaneous, well defined, over the right lower mandibular region near anterior border of masseter muscle [Fig 1]. Skin over the swelling, both parotids and oral cavity are normal. Right facial artery pulsations normally felt. No other head and neck nodes felt. Systemic examination normal. On investigation hemoglobin-13.7g/dl, total leucocytes count  $9.6 \times 10^9/L$ , ESR 30mm/hour, blood urea 24mg/dl, serum creatinine 0.9mg/dl, urine routine examination normal. Sputum AFB negative. Initial cytology (FNAC) showed early granulomatous lymphadenitis. With provisional diagnosis of tubercular lymphadenopathy, she started on anti tubercular drugs on daily basis. But after two months, on follow up swelling was increasing in size, so after admission we performed per oral excision biopsy [Fig 2] by keeping lymphoma in mind. That swelling was enlarged lymph node retained its architecture with fleshy pale appearance on section [Fig 3]. Finally histopathological examination revealed rare kimura's disease as lymphoid follicles surrounded by fibroblastic tissue with many capillaries and numerous eosinophils with many plasma cells, some of the germinal centers are also vascularised and show eosinophilic proteinaceous deposits [Fig 4,5,6]. On retrospective investigations absolute eosinophils count was  $1.1 \times 10^9/L$  and immunoglobulin E level was 650 (normal 0-380IU/ml). So, patient discharged with cetirizine 10 mg once daily for six months .She is on regular follow up once in a month without any recurrence or de novo swelling till now.

### **III. Discussion**

Kimura's disease is a rare inflammatory disease of unknown etiopathogenesis, first reported from China in 1937 which was then termed as "eosinophilic hyperplastic lymphogranuloma". It shows higher predilection of male patients in the third decade of life. More reported in Asian populations, usually present one or more subcutaneous masses in the head and neck, accompanied by satellite adenomegalies and/or increased volume of salivary glands, especially the parotid and submaxillary glands. Itchiness, urticaria, and chronic eczema, along with renal disease, are occasionally associated. Kimura disease is sometimes confused with angiolymphoid hyperplasia with eosinophilia, Hodgkin lymphoma, angioimmunoblastic T-cell lymphoma, florid follicular hyperplasia, Castleman disease, and lymphadenopathy secondary to drug reactions and parasitic infections. Appropriate cultures, histochemical stains, and immunophenotypic studies will help separate these unique and distinct lesions.

A closely related entity is angiolymphoid hyperplasia with eosinophilia (ALHE). These two diseases are distinguished on the basis of clinical and histopathological features. Lymphadenopathy and eosinophilia are more commonly seen in the case of Kimura's disease as compare to ALHE and in histopathology, Kimura's disease shows sparse vascular component and more of lymphoid proliferation with prominent eosinophilic cell infiltrate. [1,2,3]

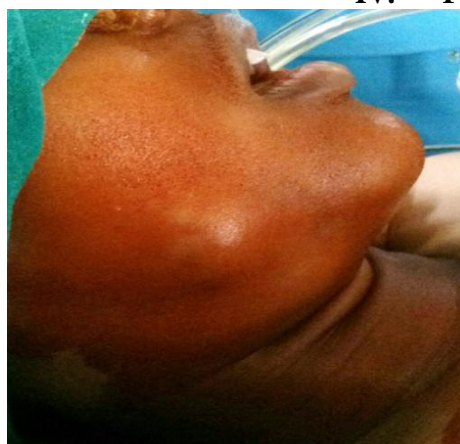
The pathophysiology of Kimura's disease remains unknown. It has been hypothesized that an infection or toxin may trigger an autoimmune phenomenon or leads to a type I (immunoglobulin E-mediated) hypersensitivity reaction. Predominance of TH2 cells which produce eosinophilic cytokines, including interleukin (IL-4 and IL-5) in patients with Kimura's disease. Additional studies have shown elevated granulocyte macrophage-stimulating factor, tumor necrosis factor- $\alpha$ , soluble IL-2 receptor, IL-5, IL-4 and IL-13. [4].

The anatomic site of lymph node involvement can be posterior auricular, cervical, and inguinal and epitrochlear lymph nodes, salivary gland involvement has also been reported. Patients almost always have marked peripheral eosinophilia, and elevated serum IgE levels. [5]

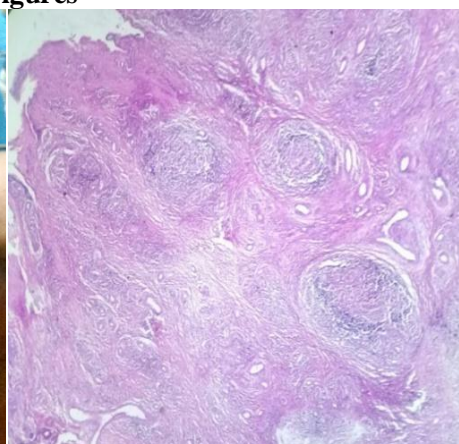
Kimura's disease diagnosed histologically, characterized by dense fibrosis, lymphoid infiltration with reactive follicles, and a mixed inflammatory cell infiltrate with numerous eosinophils all of which can develop in subcutaneous tissue, salivary glands, and lymph nodes. Some authors view it as having three components- cellular, fibrocollagenous, and vascular. The conspicuous feature of the cellular component is distinct lymphoid follicles, consisting mainly of lymphocytes. The fibrocollagenous component is formed by the infiltrate with numerous eosinophils and eosinophilic microabscesses are common; mast cells and plasma cells also abound; fibrosis is a constant, even in young lesions. The vascular component consists of proliferating and swollen endothelial cells, but these do not have atypical nuclei or abundant eosinophilic cytoplasm; salivary glands are frequently involved (they experience parenchymal atrophy and fibrosis); regional lymph nodes, usually enlarged, experience follicular hyperplasia with increases in eosinophils with or without fibrosis. [6,7]

There is no gold standard for treatment for kimura's disease. Indications include cosmesis and pressure effects. Surgical excision is the most frequently used procedure but recurrence (25%) is common. Other treatment options including antihistaminic, local radiation, systemic corticosteroids, cyclophosphamide, and pentoxifylline, have all been used with varying responses. Antiallergic agent, such as cetirizine, be tried prior to corticosteroids or for sparing the effect of corticosteroids in this disease. Systemic steroids may be used in frequent relapses or cases complicated with nephrotic syndrome. [8,9,10]

### **IV. Figures**



**Fig 1:** Pre operative



**Fig 4:** Microscopic view – low power (10X)



Fig 2: Per operative

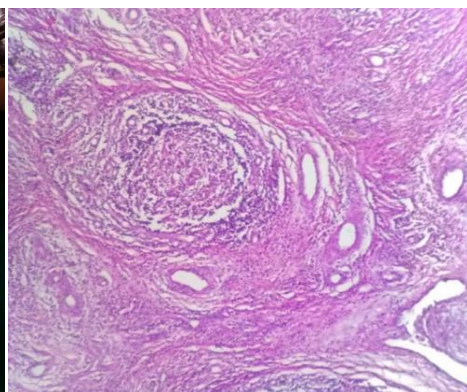


Fig 5: Microscopic view of lymphoid follicle

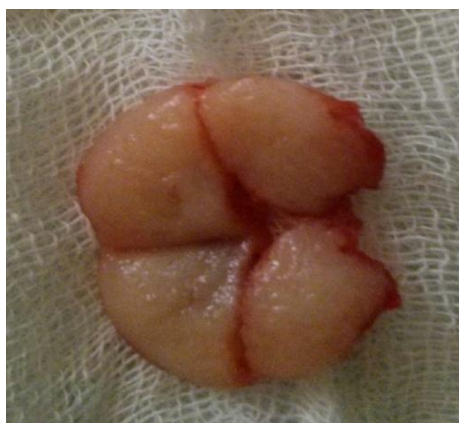


Fig 3: Cut section of lymph node

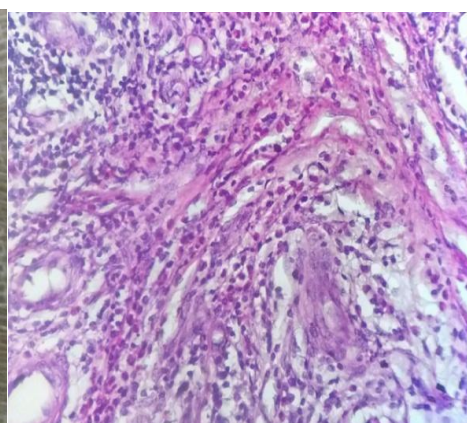


Fig 6: Microscopic view – high power (40X)

## V. Conclusion

Kimura's disease is a rare but an important case of lymphadenopathy. In our case it is facial lymphadenopathy. Kimura disease should be considered in the differential diagnosis of any patient presenting unexplained lymphadenopathy associated with non-specific symptoms. Consideration of the diagnosis is particularly important before prescribing potentially inappropriate drug therapy. We prescribed antihistaminic cetirizine in this case. Knowledge of Kimura's disease, its clinical appearance, course and histopathology puts the practitioner in a better position to answer questions from concerned patients and to optimize management strategies.

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