# Kimura's Disease

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**Citation**

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**Abstract**

Kimura's disease is a benign disease with an indolent course, gradually increasing in size over months or years. It usually presents as head and neck masses, cervical lymphadenopathy and peripheral eosinophilia. Histopathological study shows lymphoid follicles with prominent germinal centers, marked eosinophilic infiltration along with capillary proliferation and fibrosis. Surgical excision is the treatment of choice to preserve cosmesis and function.

## INTRODUCTION

Kimura's disease is a rare benign chronic inflammatory disease of unknown etiology. Kimm and Szetol first described this condition in 1937 when they reported 7 cases with the term “eosinophilic hyperplastic lymphogranuloma”. The present nomenclature was given by Kimura et al. in 1948, who noted the vascular component and described it as an “unusual granulation combined with hyperplastic changes in lymphoid tissue”. Eosinophilic granuloma of soft tissue, eosinophilic hyperplastic lymphogranuloma, eosinophilic lymphofolliculosis, eosinophilic lymphofollicular granuloma and eosinophilic lymphoid granuloma are some of the other synonyms of this rare disease.[1, 2]

## CASE REPORT

A 34-year-old female was admitted with the history of a slowly progressive recurrent swelling in the left preauricular region since 12 years. She had undergone excision 10 years ago and had lost the histopathology report. She had also noticed development of another swelling over the medial canthus of the left eye since 6 months.

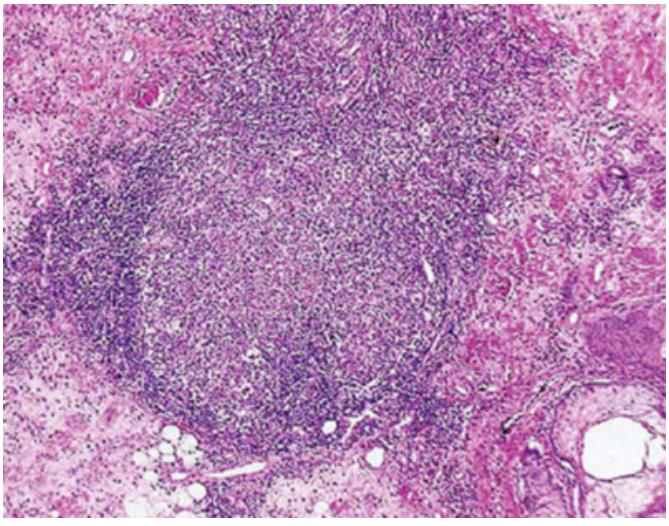
Examination revealed a non-tender oval swelling over the left preauricular area of 6x4cm with a well defined margin and not fixed to skin or any underlying structure. Another well-defined non-tender swelling of 1x1cm was found over the medial canthus of the left eye.

Laboratory investigations revealed: hemoglobin 9.7gm/dl, total leukocyte count 6510/cu.mm and differential leukocyte count: neutrophils 55%, lymphocytes 26%, eosinophils 18%, monocytes 1%. Absolute eosinophil count was 1350 cells/mcL (normal value: below 350 cells/mcL). Serum immunoglobin E level was not done in our patient.

Fine needle aspiration cytology revealed features of reactive hyperplasia. Excision of the two swellings was done under general anaesthesia. The histopathological study showed lymphoid follicles having prominent germinal centers containing marked eosinophilic infiltration along with capillary proliferation. There was also presence of eosinophils, plasma cells and histiocytes along with fibrosis in between the follicles suggestive of Kimura's disease (Fig. 1).

**Figure 1**

Figure 1



Histopathological study showing lymphoid follicles with prominent germinal centers containing marked eosinophilic infiltration along with capillary proliferation suggestive of Kimura's disease.

## DISCUSSION

Most of the cases of Kimura's disease are found in Asians and it is endemic in the Far East. Males are more commonly affected. The median age of presentation is 28 years.[3]

The pathophysiology of Kimura's disease still remains an enigma. Several theories such as an allergic reaction to repeated arthropod bites and parasitic or candidial infection; trauma and an autoimmune process have been postulated for the triggering factor in the pathogenesis. The disease is manifested by an abnormal proliferation of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and the presence of eosinophilic infiltrates in the lesion suggest a hypersensitive reaction. Systemic diseases such as connective tissue disorders, nephrotic syndrome, ulcerative colitis and asthma have been reported to be associated with Kimura's disease.[3, 4, 5, 18]

Kimura's disease is generally limited to skin, lymph nodes and salivary glands. Patients may present with single or multiple non-tender subcutaneous nodules, usually located in the region of head and neck. Periauricular, parotid and submandibular regions are commonly affected. Kimura's disease of the eyelids, orbits and lacrimal glands has been reported. It may also present as a solitary or generalized painless lymphadenopathy. Rarely, Kimura's disease may affect the extremities, inguinal and axillary lymph nodes, spermatic cord, and peripheral nerves. Spontaneous involution of the lesion is rarely seen and malignant transformation is not reported till now.[3, 5, 6, 15, 16]

Laboratory investigation almost always reveals peripheral eosinophilia along with elevation of immunoglobulin E. Serum eosinophilic cationic proteins usually follow the course of the disease. Imaging can help in determining the extent of the disease. Computed tomography may show an irregularly shaped subcutaneous mass along with salivary gland swelling. On contrast-enhanced computed tomography, lymph nodes appear homogeneous whereas salivary glands are enhanced heterogeneously. The masses are iso- or hypointense on T1-weighted magnetic resonance images and are hyperintense on T2-weighted images, compared to the parotid gland.[6, 7, 8, 14]

Histopathological study is recommended to confirm the diagnosis. Lymphoid follicles with prominent germinal centers characterize Kimura's disease. Marked eosinophilic infiltration sometimes forming eosinophilic microabscesses along with capillary proliferation forming canalized vessels with flat endothelium and fibrosis surrounding the lesion are the hallmark of this disease.[3, 5]

Fine needle aspiration cytology can be performed in recurrent lesions to obviate the need for repeated open biopsies.[9]

Surgical excision is the treatment of choice to preserve cosmesis and function. However, the recurrence rate can be as high as 25%. Other treatment options include conservative treatment with the aim of spontaneous involution, intralesional or oral steroids, radiotherapy, cryotherapy, laser, fulguration and drugs such as pentoxifylline and tretinoin (retinoids).[6, 8, 10, 11, 12, 13]

However, the overall prognosis of Kimura's disease is reported to be good.

## CONCLUSION

Kimura's disease is a benign disease with an indolent course, gradually increasing in size over months or years. A high index of suspicion is required to diagnose this condition in patients with head and neck masses, cervical lymphadenopathy and peripheral eosinophilia.

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