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# KIMURA'S DISEASE: A COMMON PRESENTATION OF A RARE DISEASE.

## Abstract

Kimura's disease is a rare chronic inflammatory disorder of unknown origin involving the subcutaneous tissue and lymph nodes of head and neck region. Clinically the swelling may mimic a neoplastic condition or tubercular infection which are more common in this region. Fine needle aspiration cytology is most often inconclusive and routine blood investigations provides little clue which leads to the delay in the diagnosis. The recurrent nature of the disease further adds to the physical and mental distress of the patient.

**Keywords:** Kimura's disease, lymphadenopathy eosinophilic lymphadenitis.

## INTRODUCTION

Kimura's disease is a form of chronic inflammatory disorder characterised by subcutaneous mass and painless lymphadenopathy primarily in the head and neck region associated with blood and tissue eosinophilia and raised serum Ig E level. The exact prevalence of this rare disease not known but according to a report published in 1997 , less than 120 cases had been reported worldwide.[1] This disease has been reported frequently in the Chinese , Japanese and South Asians however sporadic cases have been seen in Europeans and Americans. Here we report one such case.

## CASE REPORT

A twelve years old boy from Sindhupalchok, Nepal came to ENT OPD with right preauricular swelling for five months (figure 1). It was painless but gradually
increasing in size. On examination the patient had diffuse swelling of 8x5 cm in the right parotid region, overlying skin was normal and was soft ,non tender on palpation with some firm nodular structures within it and the facial nerve was intact. On routine investigation his total WBC count was 8800/cumm, neutrophil count was to 20% however eosinophil count was to 34%. ESR was 20mm. Ultrasonogram of the preauricular
region showed multiple enlarged lymph nodes in the right parotid and cervical region. FNAC was done them the lesion which showed scattered lymphoid cells predominantly small mature lymphocytes in some haemorrhagic background suggesting of
reactive lymphadenitis. A course of broad spectrum antibiotic was prescribed for seven days but the lesion did not subside so an incisional biopsy
including the skin and subcutaneous tissue was carried out under local anesthesia. Sections from the specimen showed dense mixed inflammatory cell infiltration consisting of lymphocytes, plasma cells, neutrophils and eosinophils in an oedematous stroma suggesting of inflammatory lesion. The biopsy was repeated to include deeper tissue and two large lymph nodes each measuring about 3x2x2 cm each. Histopathology report showed lymphoid follicles with marked hyperplasia of germinal centers.Germinal centers were well vascularised and containeosinophilic deposits with extensive infiltration of the interfollicular area by mature eosinophils occasionally forming eosinophilicmicroabscess (figure 2). He was diagnosed as Kimura's disease and was given intralesional steroid. His lesion had subsided in size and is on regular follow up.

## DISCUSSION

Though this disease has predilection for the people of Asian origin, it is rarely seen in Nepali population. The first report of Kimura disease was from China in 1937, in which the authorsdescribed seven cases of a condition they termed "eosinophilic hyperplastic lymphogranuloma.[2] The disorder received its current name in 1948, when Kimura et alnoted the vascular component and referred to it as an "unusual granulation combined with hyperplastic changes in lymphoid tissue".[3] Kimura's disease is more common in male patients (male/female
ratio 6:1) with with mean age of 32 years.[4] It almost exclusively involves the regions of in head and neck: such as postauricular, cervical and epitrochlear, howeveringuinal lymph nodes involvement have also been reported.[4] Kimura's disease in our patient was involving the preauricular region which was similar in location to the one

reported by Shah et al.[5] Eosinophilia and elevated serum Ig E level is a common finding in this disease. Chen et al noted eosinophilia in 16 cases out of 21.4 Histological feature of the involved subcutaneous tissue and lymph node reveal marked hyperplasia of the follicle with proliferating germinal centre. The follicles and the interfollicular area are extensively infiltrated with eosinophils. Occasionally eosinophilicmicroabscesses are seen in the interfollicular areas. Perinodal soft tissue and the subcutaneous tissue are also infiltrated with eosinophils. Kimura disease is sometimes confused with angiolymphoid hyperplasia with eosinophilia ALHE) which occurs in the superficial skin while Kimura's disease involve deeper tissue of the head and neck region. ALHE does not present with increase in Ig E level and, lymphadenopathy is rare but has prominent vascular proliferation.[6] Treatment primarily consists of surgical excision of the lesion however relapse is common. Intralesional and systemic corticosteroids are another viable option wherecosmesis is desired and in cases of relapsing form of the disease. Cyclosporine has been reported to induce remission in patients with Kimura disease.[7] Local radiation therapy (30-40 Gy) is also prescribed in lesions refractory to corticosteroid therapy or where surgery is not possible.[8] Pranlukast ( leukotriene receptor ntagonist) and cetirizine (H-1 receptor blocker ) have been shown to be effective in small number of patients.[9] Omalizumab, a humanisedmonoclonal antibody which binds to free Ig E, has recently been shown to be effective in decreasing the size of the growth as well as peripheral eosinophilia.[10]

## CONCLUSION

Kimura's disease is a chronic inflammatory lesion of the subcutaneous tissue and the lymph node of the head and neck region. The exact aetiology of this disease is unknown. Affects middle aged male patients and histopathological examination is diagnostic. Though it has a benign course relapses are seen occasionally.

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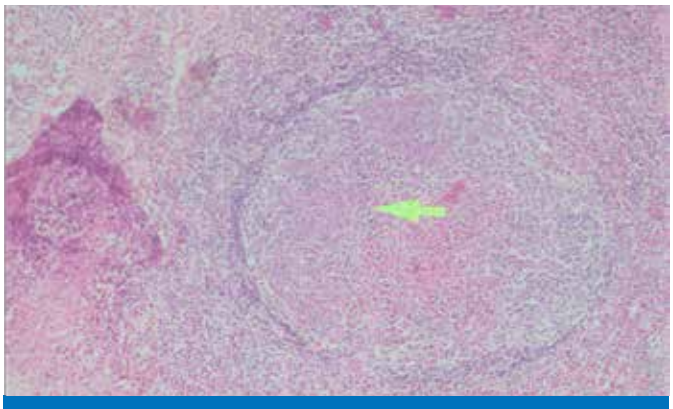

Figure 2 Follicular hyperplasia with eosinophilic infiltrates