# A Rare Case of Kimura's Disease

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## ABSTRACT

A rare case of Kimura's disease was seen in a 42-year-old female patient who presented with swelling in left pre-auricular region. Initial cytology (FNAC) showed angio-follicular hyperplasia suggestive of early lymphoma or Castleman's disease. However only after biopsy, final diagnosis of this rare disease could be made. This emphasizes the need of considering Kimura's disease among differential diagnosis for swelling over parotid region.

**Key words:** FNAC, Lymphadenopathy, Castleman's disease

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## Introduction

Kimura's disease is a rare condition, first described in 1937, as painless, sometimes disfiguring, subcutaneous nodule in head & neck region. It is a chronic inflammatory disease of unknown etiology, having eosinophilia with adenopathy. It is endemic in Asia. It is mainly a disease of middle aged men (M:F= 3.5:1) [1]. There are fewer than 120 cases reported worldwide. Few cases of palate & cheek with eyelid affliction have been described [2, 3], Besides a patient presenting as lymphadenopathy and painful oral ulcerations was reported [4]. It presents with painless soft tissue nodules and lymphadenopathy in the head and neck region. Common sites of involvement are the parotid glands and the epitrochlear, axillary, and inguinal nodes. Although the masses enlarge slowly, patients remain otherwise asymptomatic. Pruritus and dermatitis may occur, and skin lesions can present as reddish brown papules or as subcutaneous nodules. Rare sites of involvement include the kidneys, orbits, ears, spermatic cord, and nerves [5]. Laboratory findings include peripheral eosinophilia and elevated serum IgE levels, and the diagnosis is made histologically, preferably from a lymph node biopsy.

## Case Report

A 42 years old Female, presented with swelling in front of left pinna for last 5 years at the Department of ENT, Chirayu Medical College & Hospital, Bairagarh, Bhopal, M.P. Patient had undergone treatment from various practitioner including 2 weeks of ATT, with no relief in symptoms, before presenting to us. On examination, a 3cm x 2 cm mobile, non-tender, fluctuant swelling was seen in Left pre-auricular region.(Figure.1) Overlying skin was dark red in colour, but temperature was normal. Few lesions were seen extending to root of helix & superior & anterior wall of EAC. Otomycosis was seen in left EAC, which on cleaning showed intact & retracted tympanic membrane. On examination of neck, multiple lymph nodes at level Ia, II & III on left side were seen, that were firm, mobile, non-tender, non-fluctuant on palpation. Rest of the ENT examination was within normal limit. CBP was done that showed Hb: 11.6gm%, TLC: 6600/mm3 DLC: N78, L16, E04, M02, ESR: 13mm/1hr. LFT, KFT, urine examination were within normal limits. HIV, HBsAg & HCV was negative. Mantoux test was negative with 5TU PPD. X-ray chest was also within normal limit. USG neck revealed multiple discrete & confluent lymph nodes at left intra-parotid and sub mandibular (Ib), largest measuring 2.3 x 0.9 cm. Few lymph nodes were also seen at left level II & III with short axis diameter of less than 10mm. On aspiration, frank blood came out. FNAC from pre-auricular swelling & left level II lymph node was suggestive of Angiofollicular hyperplasia (Castleman's disease) or early follicular lymphoma. Based on this an excision biopsy of level II lymph node was done which on HPE showed findings of florid follicular hyperplasia with eosinophilia, possi-
bly Kimura disease.

Discussion

Kimura's disease was first described in China in 1937, but it was not referred to as Kimura's disease until its description in the Japanese-language literature in 1948 [6,7]. The cause of this rare disease is thought to be an immune-mediated disorder and not neoplastic.

The characteristic histologic features of Kimura's disease were classified in 1989 as being constant, frequent, or rare. The constant features are preserved lymph node architecture, florid germinal centers, eosinophilic infiltration, and an increased amount of postcapillary venules. The frequent features include sclerosis, karyocytosis in both the germinal centers and the para-cortex, vascularization of the germinal centers, proteinaceous deposits in germinal centers, necrosis of germinal centers, eosinophilic abscesses, and atrophic venules in sclerotic areas. The single rare feature is the progressive transformation of germinal centers. The immunochemistry findings were also described, which are IgE reticular network in germinal centers and IgE-coated non degranulated mast cells [8]. Due consideration to above mentioned histologic features differentiates this rare condition from other more common masses. Malignancy should be ruled out first, and the absence of Hodgkin's disease helps exclude Hodgkin's disease. Unfortunately, T-cell lymphomas can present with polymorphonuclear lymphocytes and eosinophilia, making the distinction difficult. Although atypical, histiocytosis X can present with subcutaneous masses; the histologic diagnosis is made by finding characteristic Langerhans' cells. Peripheral eosinophilia suggests a parasitic infection or allergic reaction as a cause of soft tissue swelling. Further data to support either are lacking. Differentiating Kimura's disease from angiolymphoid hyperplasia with eosinophilia requires a strict analysis of clinical and histologic features because the diseases are similar and were once thought to be the same disorder. Both diseases usually present with soft tissue masses in the head and neck region, but in angiolymphoid hyperplasia with eosinophilia, the lesions are mostly dermal or subcutaneous and not often in lymph nodes, which is a common location for Kimura's lesions. Angiolymphoid hyperplasia with eosinophilia is more typically seen in middle-aged women and Kimura's disease in younger men. In both cases, histologic examination of the lesion shows lymphocyte infiltration, numerous eosinophils, germinal center formation, and proliferative blood vessels. But most important, the vascular endothelial cells in angiolymphoid hyperplasia have nuclei of varied size and shape and hemosiderin deposits. Also, the endothelial lining usually is more than one cell thick. These changes are not seen in Kimura's disease. These histologic changes suggest that angiolymphoid hyperplasia with eosinophilia is neoplastic in origin, whereas Kimura's disease is an immune-mediated disorder [9,10].

The treatment of Kimura's disease has mainly involved the use of oral corticosteroids. Radiation treatment is usually used for the local control of lesions not responsive to steroids, and total doses of 20 to 30 Gy have proved effective. Irradiation should be considered not only in patients resistant to steroids, but also in young patients in whom the long-term side effects of steroids may be more deleterious than a limited course of irradiation that may prevent relapse [11]. Other treatments have included complete surgical excision, the intralesional administration of steroids, cytotoxic agents, and electrodesiccation. Although spontaneous resolution has been reported, most patients have a prolonged course with slow enlargement of the masses. There is no potential that the lesions will become malignant.

Conclusion

Kimura’s disease is a rare condition with similar clinicopathologic presentations with many diseases of the head and neck region. However only after biopsy, final diagnosis of this rare disease can be made. This emphasizes the need of considering Kimura’s disease among differential diagnosis for swelling over parotid region.

References


9. Chow LTC, Yuen RWS, Tsui WMS, Ma TKF, Chow WB, Chan SK. Cyto- logical features of Kimura’s disease in fine needle


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