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# HIDDEN IN PLAIN SIGHT: RECOGNISING KIMURA DISEASE AMONG COMMON CONDITIONS

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## ABSTRACT

Kimura disease is a rare chronic inflammatory disorder of unknown etiology, predominantly seen in young Asian males. It is characterized by painless subcutaneous swelling, serum and tissue eosinophilia with raised IgE levels.

## KEYWORDS

submandibular gland, lymphocytes, eosinophils, germinal centres

## INTRODUCTION

Kimura disease is a rare chronic inflammatory disorder of unknown etiology. It presents as a subcutaneous mass in the head and neck region and is frequently associated with regional lymphadenopathy or salivary gland involvement.[1] It is generally seen in young Asian adults, within the age group of 20 and 40 years; men are affected more commonly than women, in the ratio of 3:1. Sites commonly involved are periauricular, groin, orbit, and eyelids. Peripheral blood eosinophilia and elevated serum immunoglobulin E (IgE) levels are characteristic features of Kimura's disease.[2] 10% to 60% of the patients have a coexistent renal pathology. Diagnosis includes biopsy or excision of the involved mass for a pathological examination.

### Case Study

A 42 year male came with complaints of left sided neck swelling since 4 years, initially small in size gradually progressive to current size, not associated with pain. No history of throat pain, difficulty in swallowing, other constitutional symptoms. On examination: A 3X3 cm ovoid swelling noted in the left submandibular region extending superiorly till angle of mandible ,1 cm from the midline medially 2 cm from the left sternocleidomastoid laterally. On palpation inspectory findings confirmed, firm in consistency, mobile, no warmth, no tenderness.

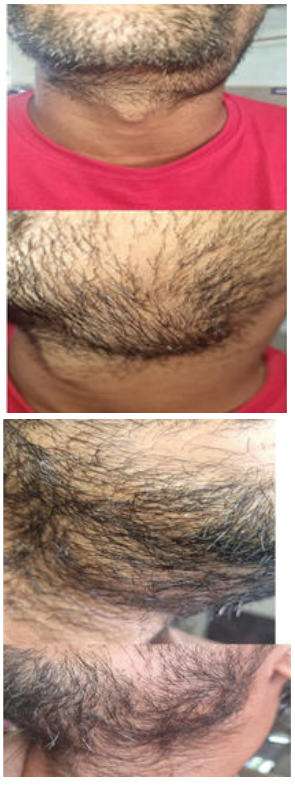

**Fig.1** Clinical images of patient presenting with left sided neck swelling

**CECT neck:** Bilateral submandibular gland enlargement suggestive of acute sialadenitis. FNAC of submandibular swelling showed features of inflammatory process. Patient underwent excision of left submandibular gland under general anesthesia and specimen sent for histopathological analysis.

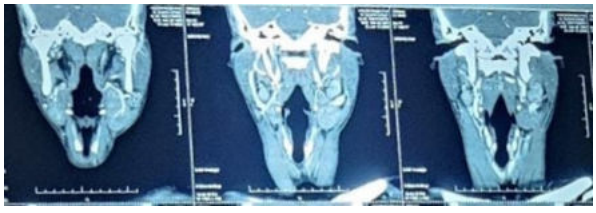

**Fig.2** CT neck with contrast images showing enlarged submandibular glands

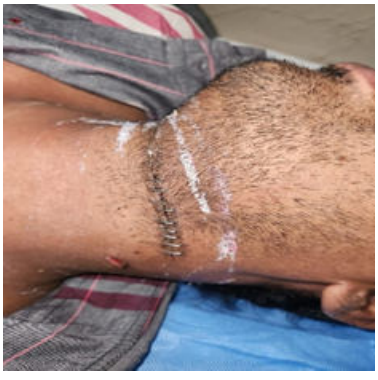

**Fig.3** Postoperative image

Histopathological examination revealed salivary gland tissue infiltrated by dense chronic inflammatory infiltrate composed of lymphocytes, prominent germinal centres, multiple blood vessels, dense eosinophilic infiltrates with abscess formation. Features suggestive of angiolymphoid hyperplasia with eosinophilia - KIMURA'S DISEASE

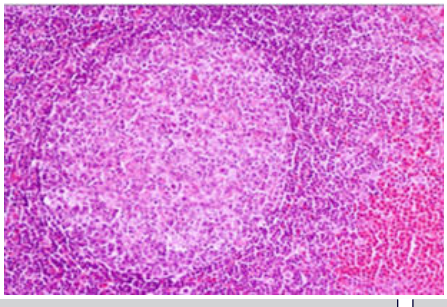

**Fig.4** Lymphocyte aggregates with germinal centres with eosinophilic infiltration Postoperatively patient was discharged in a stable state

## DISCUSSION:

Kimura Disease is a rare chronic inflammatory disorder, first described in 1937 by Kim and Szeto as "eosinophilic hyperplastic lymphogranuloma" and has been known as Kimura's disease since its description by Kimura et al. in the Japanese literature in 1948. This disease is endemic in middle-aged Asian males and rarely seen sporadically in non-Asian population. The 2nd and 3rd decades of life are the most common. It presents predominantly as subcutaneous nodules in the head and neck, often unilateral, and frequently associated with regional lymphadenopathy with or without the involvement of salivary glands. [3,4] Orbit, eyelid, palate, and pharynx have also been reported to be involved, in the head and neck. The clinical course of the disease is generally benign and self-limiting. Most patients have a prolonged course with gradual increase in the swelling. Occasional spontaneous resolution is seen.

The exact etiology and pathogenesis of Kimura's disease are unclear, although it might be a self-limited allergic or autoimmune response triggered by an unknown persistent antigenic stimulus. Studies show that the proliferation of CD4+ T cells, specifically the CD4 T-helper2 (Th2) cells and resultant overproduction of their cytokines, such as granulocyte macrophage colony-stimulating factor, tumour necrosis factor-α, IL-4, IL-5, eotaxin, and RANTES trigger the production of lymphoid follicle and high IgE.[5] Clonal T-cell population attributes to the disease development and recurrence. Upto 60% of these patients exhibit renal involvement manifesting as extra membranous glomerulonephritis and nephrotic syndrome. Final needle aspiration show significant numbers of eosinophils in a background of lymphoid cells with occasional fragments of collagenous tissue and Warthin-Finkeldey polykaryocytes.[6] In our case of FNA smears revealed salivary gland tissue infiltrated by dense chronic inflammatory infiltrate composed of lymphocytes, prominent germinal centres, multiple blood vessels, dense eosinophilic infiltrates with abscess formation.

Hui et al. classified the histological features of Kimura's disease as constant, frequent and rare. The constant features include preserved nodal architecture, florid germinal centre hyperplasia, eosinophilic infiltration and postcapillary venule proliferation. Frequent features comprise sclerosis, polykaryocytes, vascularization , proteinaceous deposits and necrosis of the germinal centres , eosinophilic abscesses and reticular IgE deposition within germinal centres. Nodal architecture is largely preserved in most cases, however, capsular fibrosis with subcapsular sinusoid obliteration and perinodal soft tissue involvement is frequently present.[7,8]

The differential diagnosis includes angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin's disease, Kaposi sarcoma, eosinophilic granuloma, epithelioid hemangioma, Castleman's disease, tuberculosis, dermatopathic lymphadenopathy, lymphadenopathy of drug reactions, parasitic lymphadenitis, eosinophilic granuloma, epithelioid hemangioma.[9]

Histologically, kimura's disease has three components: cellular (inflammatory infiltrate including increased eosinophils and follicular hyperplasia), fibrocollagenous and vascular (arborizing vascular proliferation of the post capillary venule, endothelial cells are usually flat and lack cytologic atypia or vacuolization).

Imaging studies can help in staging the extent and progression of the disease as well as the lymph node involvement. The diagnosis in our case was through the histopathological examination of the excised tissue.

Management methods include surgical excision, steroids and radiation. Surgical excision may be considered first especially for localized lesions. Systemically administered steroids show good effects on disease progression; however, withdrawal of steroids can often result in relapse.The rate of recurrence of disease is about 25%. [10,11] Steroid resistant lesions can be treated with radiation

## CONCLUSIONS

Kimura's disease should be considered as a differential diagnosis in patients presenting with a mass in the head & neck region along with lymphadenopathy and investigated accordingly if FNAC shows lymphoid aggregates. Early diagnosis can help in prompt management of the condition and symptomatic relief to the patient.

## REFERENCES:


1. Ioachim H, Ratech H. Kimura lymphadenopathy. In: Ioachim H, Ratech H, editors. Ioachim's Lymph Node Pathology. 3rd ed. Philadelphia: Lippincott-Raven; 2002. pp. 209–11.
2. Abhange RS, Jadhav RP, Jain NK. Kimura disease: A rare case report. Ann Pathol Lab Med. 2018;5:53–55.
3. Rajesh A, Prasanth T, Naga Sirisha VC, Azmi M. Kimura's disease: A case presentation of postauricular swelling. Niger J Clin Pract. 2016;19:827–830.
4. Tseng CF, Lin HC, Huang SC, Su YC. Kimura's disease presenting as bilateral parotid masses. Eur Arch of Otorhinolaryngol. 2005;262:8–10.
5. Fouda MA, Gheith O, Refaie A, El-Saeed M, Bakr A, Wafa E, et al. Kimura disease: a case report and review of the literature with a new management protocol. Int J Nephrol. 2011;2010 673908.
6. Shetty AK, Beaty MW, McGuirt WF, Jr, Woods CR, Givner LB. Kimura's disease: a diagnostic challenge. Pediatrics. 2002;110:e39.
7. Chen H, Thompson LD, Aguilera NS, Abbondanzo SL. Kimura disease: a clinicopathologic study of 21 cases. Am J Surg Pathol. 2004;28:505–513.
8. Meningaud JP, Pitak-Arnnop P, Fouret P, Bertrand JC. Kimura's disease of the parotid region: report of 2 cases and review of the literature. J Oral Maxillofac Surg. 2007;65:134–140.
9. Chong WS, Thomas A, Goh CL. Kimura's disease and angiolymphoid hyperplasia with eosinophilia: two disease entities in the same patient: case report and review of the literature. Int J Dermatol. 2006;45:139–145.
10. Deshpande AH, Nayak S, Munshi MM, Bobhate SK. Kimura's disease. Diagnosis by aspiration cytology. Acta Cytol. 2002;46:357–363.
11. Hui PK, Chan JK, Ng CS, Kung IT, Gwi E. Lymphadenopathy of Kimura's disease. Am J Surg Pathol. 1989;13:177–186.