



CASE REPORT **DERMATOLOGY**

Kimura's disease: A diagnostic dilemma

Asha Jyothi¹, Indira Bonthu¹, Anitha Vinnakoti¹, Suresh Kumar Behara¹

¹Department of Dermatology, Venerology and Leprosy, Rangaraya Medical College, Kakinada, India

***Corresponding author:**

Ch. Asha Jyothi,
Department of Dermatology,
Venereology and Leprosy,
Rangaraya Medical College,
Kakinada, India
ashachiguru5@gmail.com

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ABSTRACT

Kimura disease is benign, rare, chronic inflammatory condition seen predominantly in younger males. There is 3–6-fold male occurrence with a median age at onset of 32 years. The classical presentation is with bilateral cervical lymphadenopathy, occasionally fever, accompanied by peripheral blood eosinophilia and high levels of IgE. It usually presents as subcutaneous masses, particularly around the head and neck, with a variety of nonspecific findings, including itch, urticaria, and chronic eczema. 15% of patients with Kimura disease develop nephrotic syndrome.

Keywords: Angiod Hyperplasia, Biopsy, Eosinophilia, IgE, Kimura disease

INTRODUCTION

A rare, benign, chronic inflammatory¹ condition that mostly affects young males is identified as Kimura disease. It is more common in Asians. Occurs more commonly in the age group of 20–40 years. Classical presentations with high IgE levels, peripheral blood eosinophilia, bilateral cervical lymphadenopathy, and occasionally fever are recognized by subcutaneous lumps,² especially around the head and neck,³ as well as a range of nonspecific symptoms including itching, urticaria, and chronic eczema. With Kimura disease, nephrotic syndrome⁴ develops in about 15% of patients.

CASE REPORT

A 30-year-old male has been experiencing multiple nodules and papules over the left postauricular region for a year, which is gradually progressive. Not associated with itching, fever, and urticaria. Examination revealed multiple hyperpigmented subcutaneous nodules [Figures 1-3] over the left postauricular region [Figure 1]. No pus or bloody discharge was seen in Figure 2. Hematological examination revealed Hb 11 g/dl, TLC 9,000 cells/cumm (neutrophils 31%, lymphocytes 40%, eosinophils 25%, and monocytes 2%) and adequate platelets. Serum immunoglobulins IgE levels were elevated. FNAC of the regional lymph node showed a polymorphous population of lymphoid cells, histiocytes, and mixed inflammatory infiltrate. A biopsy from the lesion showed hyperplastic follicles with germinal centers in lymphoid cells with eosinophils, and eosinophil infiltrate with microabscesses in are present.

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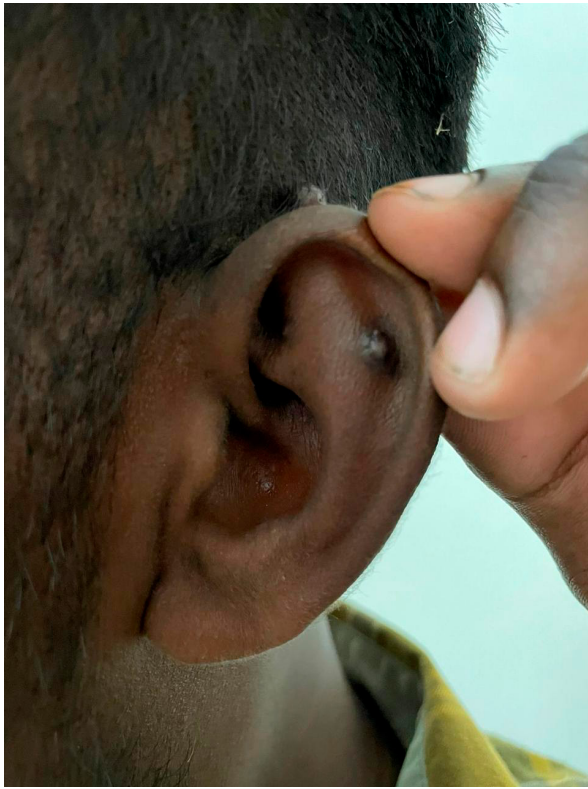


Figure 1: Subcutaneous nodules over the helix of the ear.

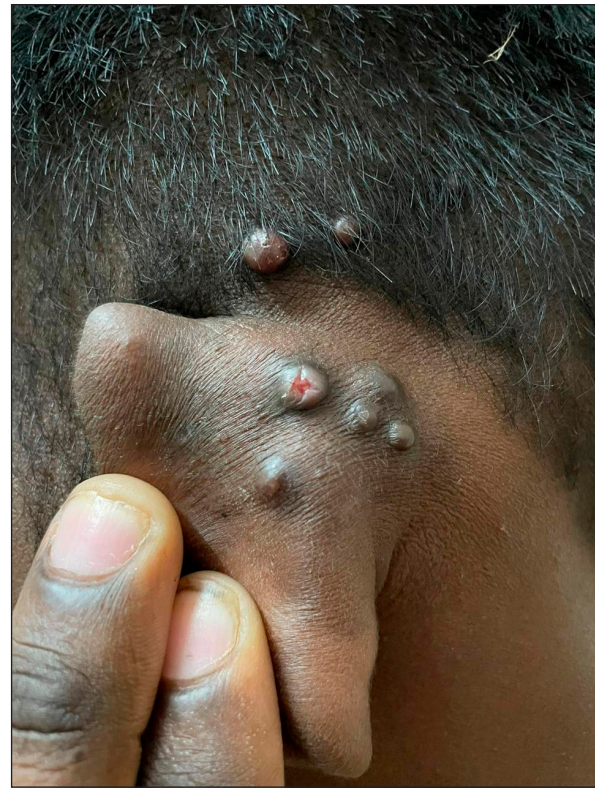


Figure 2: Subcutaneous nodules over retroauricular region.



Figure 3: Subcutaneous nodules over the ear pinna and occipital area.

DISCUSSION

Kimura disease, originally described by Kimura, is an inflammatory condition of soft tissue of unknown cause. It was originally called Eosinophilic Hyperplastic Lymphogranuloma. It is characterized by lymphatic and angiomatous proliferation and related to regional lymphadenopathy. Disease appears as large, frequent, unilateral nodules in under the skin on the head and neck, along with eyes, axilla, pharynx, palate, groin, and arms. The disease is self-limiting and benign. Exact cause is not known; it may be due to an autoimmune response triggered by some antigenic stimulus. Studies show CD4⁺ Proliferation, CD4-T helper 2 (Th2) cells, and overproduction of cytokines, interleukins, and TNF-alpha. Kimura disease should be included in the differential diagnosis of any lymph node presenting with eosinophilic infiltrate and follicular hyperplasia [Figures 4 and 5]. Differential diagnosis includes Hodgkin's disease, ALHE (Angiolymphoid hyperplasia with Eosinophilia), Castleman's disease, infections like tuberculosis, dermatophytes, parasitic origin, and eosinophilic granuloma. Histopathologically, Kimura shows inflammatory infiltrate,⁵ which includes follicular hyperplasia, increased eosinophils, and fibrocollagenous and vascular proliferation of endothelial cells. The diagnosis in our case is through cutaneous biopsy

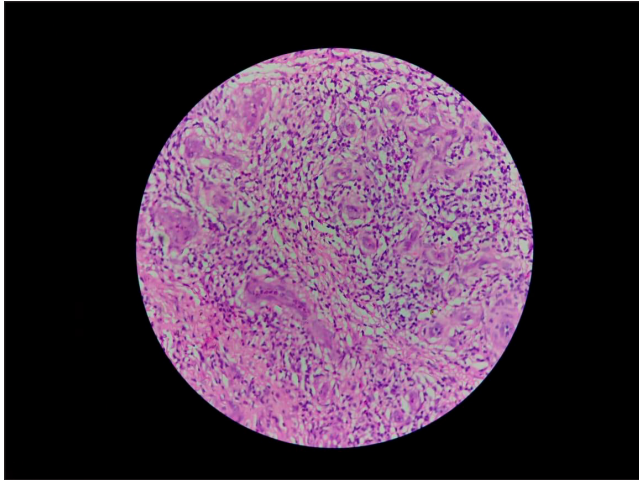


Figure 4: Follicular hyperplasia (Hematoxylin and eosin stain, 40X).

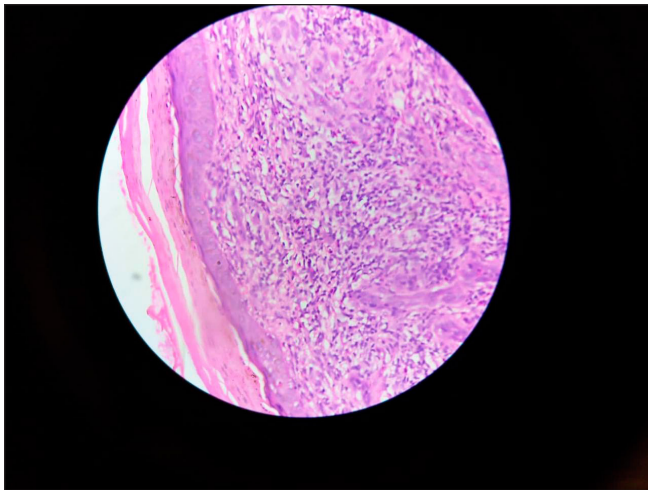


Figure 5: Inflammatory infiltrate with increased eosinophils (Hematoxylin and eosin stain, 40X).

and histopathological correlation. The closest differential is ALHE, which is characterized by more female prevalence and regional lymphadenopathy, peripheral eosinophilia, and elevated IgE levels that have been rarely observed.

Kimura disease can be treated with surgical removal of the localized lesions. Systemic steroids decrease the disease

progression. Radiation for steroid-resistant lesions. In our case, the patient was kept on topical steroids and is under follow-up.

CONCLUSION

Kimura disease is a rare condition that mimics neoplastic origin. Patients with regional lymphadenopathy along with head and neck masses should be evaluated for this condition. A proper knowledge of the disease helps us to narrow down the diagnosis.

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