

# Kimura's disease: a cheek presentation



Submitted: 16-05-2017

Revised: 02-06-2017

Published: 01-07-2017

Sir,

Kimura's disease, also known as eosinophilic lymphogranuloma, is a chronic inflammatory disease of unknown aetiology. Though the disease is endemic in Asia, it is considered as a rare entity.<sup>1</sup>

A 20 year old male, without any comorbidities, presented with complaints of swelling over right cheek for the past 1 month. It has been progressive and not associated with pain or fever. He did not have any history of allergies. Local examination revealed a soft non-tender swelling of about 5 cm × 4 cm over the right zygomatic region (Figure 1). The skin over the swelling was normal, with no local rise in temperature. His vitals and systemic examinations were normal. His blood investigations showed normal leukocyte count but eosinophils 42%. Erythrocyte sedimentation rate was elevated (51 mm in the first hour). His serum IgE levels were 482 UI/ml (normal 1-87 UI/ml). Other blood investigations like renal and liver functions, urine examination and chest Xray were normal. Excision biopsy was done and histopathological examination revealed lymphoid hyperplasia with dense eosinophilic infiltration and proliferation of post-capillary venules, suggestive of Kimura's disease. He was given 1 month course of prednisolone (1 mg/kg/day with tapering), along with montelukast (10 mg once daily) and cetirizine (10 mg once daily). After 2 weeks, there was mild regression of the swelling; but patient was lost for follow up thereafter.

Kimura's disease was first described by Kimm and Szeto in 1937. However, the actual histological description was explained by Kimura in 1948; and thus bearing his



**Figure 1:** Soft non-tender swelling over the right zygomatic region

name. An immune reaction towards an antigenic stimulus is considered to be the probable mechanism; with prolonged immunogenic stimulation leading to lymphoid proliferation. The mast cells play a major role by regulating IgE synthesis and promoting eosinophilic infiltration. The disease has a male predominance; with peak age of onset during the third decade. These patients present with one or more subcutaneous nodules over the head and neck region, which are indolent and progressive in nature. The nodules can be painful and pruritic. The skin over the lesion is normal. Some patients may have an associated extramembranous glomerulonephritis.<sup>2</sup> Laboratory investigations show peripheral blood eosinophilia and elevated IgE levels. On histological examination, the excision specimen shows hyperplastic lymphoid tissue with proliferating germinal centers, eosinophilic infiltration and proliferation of post-capillary venules. Surgical excision of the lesion is the mainstay of therapy. Corticosteroids are indicated in refractory cases and in those with renal involvement.<sup>1</sup> Pranlukast and cetirizine have also shown positive results.<sup>3,4</sup> Local radiation therapy has been indicated for corticosteroid failure cases and in those where surgery is not possible.<sup>1</sup> Cyclosporine and levamisole have been used in the treatment of Kimura's disease associated with steroid dependant nephrotic syndrome.<sup>5,6</sup>

Because of its benign nature, Kimura's disease may go unrecognised. Since the condition has an association with nephrotic syndrome, an early recognition and diagnosis of this disease may prove useful.

**Key words:** Kimura's disease, Eosinophilic lymphogranuloma, Eosinophilia

**Manoj Gopalakrishnan<sup>1</sup>, Robin George Manappallil<sup>2</sup>,  
Dipu Ramdas<sup>3</sup>, Neena Mampilly<sup>4</sup>**

**Access this article online****Website:**<http://nepjol.info/index.php/AJMS>**DOI: 10.3126/ajms.v8i4.17318****E-ISSN: 2091-0576****P-ISSN: 2467-9100**

<sup>1</sup>Senior Consultant- Physician, <sup>2,3</sup> Consultant- Physician, Department of Internal Medicine, Baby Memorial Hospital, Calicut, Kerala, India, <sup>4</sup>Senior Consultant-Pathologist, Department of Pathology, Baby Memorial Hospital, Calicut, Kerala, India

**Address for correspondence:**

Dr. Robin George Manappallil,  
Consultant- Physician, Department of Internal Medicine,  
Baby Memorial Hospital, Calicut, Kerala 673004, India.  
**E-mail:** drrobingeorgempl@gmail.com  
**Tel:** 0091-8547753396

## REFERENCES

1. Larroche C and Blétry O. (2005) Kimura's Disease. Orphanet Encyclopedia
2. Chauvel C, Valeyrie-Allanore L, Essig M, Vrtovsnik F, Walker F, Marinho E, et al. Kimura's disease associated with extracapillary glomerulonephritis. *Ann Dermatol Venereol* 2006; 133:689-692.
3. Ohtsuka Y, Shimizu T, Fujii T, Sato T, Saito M, Nagata S, et al. Pranlukast regulates tumour growth by attenuating IL-4 production in Kimura disease. *Eur J Pediatr* 2004; 163(7):416-417.
4. Ben-Chetrit E, Amir G and Shalit M. Cétirizine: An Effective Agent in Kimura's Disease. *Arthritis Rheumatology* 53; 117-118.
5. Do YS, Kim HY and Namgoong MK. A case about cyclosporine treatment of Kimura's disease associated with steroid dependant nephrotic syndrome. *J Korean Soc Pediatr Nephrol* 2009; 13:271-277.
6. Jaesung Yu, Hyunju Jin and Hoseok Kang. A Case of Levamisole Treatment for Kimura's disease -Associated Nephrotic Syndrome. *The Korean Journal of Nephrology* 2011; 30: 315-320.

**Authors Contribution:**

**MG**-Concept and design of case report and treating Physician; **RGM**-Concept and design of case report, reviewed the literature, manuscript preparation and treating Physician; **DR**-Critical revision of manuscript and treating Physician; **NM**-Critical revision of manuscript and Pathologist in-charge.

Dr. Robin George Manappallil:  <http://orcid.org/0000-0003-3973-6800>

**Source of Support:** Nil, **Conflict of Interest:** None declared.