

**CASE REPORT**

# An Unusual Clinical Presentation of Kimura's Disease Occurring in the Mesenteric Lymph Nodes

Sheikh Bilal Ahmad, Hamdani Shaista Mushtaq, Beigh Ambreen

**Abstract**

Kimura's disease is a rare inflammatory disorder of unknown aetiology primarily seen in young Asian males. The disease is characterized by a triad of painless subcutaneous masses in the head and neck region, blood and tissue eosinophilia, and markedly elevated serum immunoglobulin E (Ig E) levels. Clinically the subcutaneous nodules occur predominantly in the head and neck region of young males. However, we report the case of a 60 year old male presenting with mesenteric lymphadenopathy diagnosed with Kimura's disease.

**Key Words**

Kimura's Disease, Mesenteric Lymph Node

**Introduction**

Kimura's disease is a rare chronic inflammatory disorder of unknown cause, primarily seen in young Asian males(1). The typical clinical presentation is characterized by a triad of painless unilateral cervical adenopathy or subcutaneous masses predominantly in the head or neck region, blood and tissue eosinophilia, and markedly elevated serum immunoglobulin E (Ig E) levels (2). Descriptions of Kimura's disease are limited with hardly over 120 cases reported worldwide (3). Although the disease can present at any age, most cases have been reported in the second or third decades of life and sites such as the oral cavity, axilla, groin, limbs, and trunk may also be involved (4). We report a case of Kimura's disease in the mesenteric lymph nodes of a 60 year old Asian male.

**Case Report**

A 60 year old male presented to surgical casualty with history of pain whole abdomen, colicky in nature, off and on for a duration of 4 days which was associated with 2-3 episodes of vomiting. There was no history of fever, night sweats, weight loss, blood with stools, hematemesis, jaundice, or anorexia. There was a history of similar episodes of abdominal pain in the past. On physical examination, he seemed well except mild distention of the abdomen with mild diffuse tenderness all over the

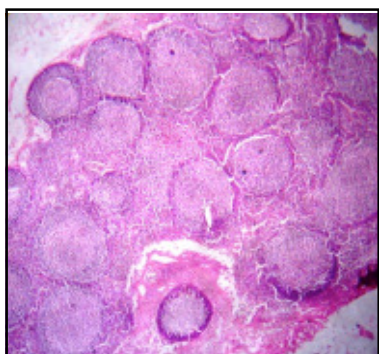
abdomen. Laboratory data included a hemoglobin of 14.2 g/dl, platelet count of 138x10<sup>9</sup>/L, and white cell count of 7.9x10<sup>9</sup>/L:differential showed 56% segmented neutrophils, 17% lymphocytes, and 26% eosinophils, and 1% monocytes. The erythrocyte sedimentation rate was 25mm/hr. Results of serum electrolytes, liver function tests, albumin, blood urea nitrogen and creatinine were normal. Mantoux test for tuberculosis was mildly positive. X ray abdomen showed multiple air fluid levels and ultrasonography revealed mildly distended gut loops with no interloop fluid or ascitis.

Sub acute intestinal obstruction (SAIO) was the first diagnosis made and patient was put on conservative treatment. While the patients general condition improved, abdominal pain persisted. In view of pain abdomen not responding to conservative treatment and positive Mantoux test a decision of diagnostic laparotomy was taken with the indication of Recurrent SAIO ( Abdominal Tuberculosis). Intra operative findings revealed multiple enlarged lymph nodes in the mesentery. Whole of the small and large gut was normal, there was no evidence of abdominal tuberculosis or malignancy. Multiple mesenteric lymph nodes were taken for biopsy.

The histopathology of the lymph nodes revealed follicular lymphoid hyperplasia with progressively

From the Department of Pathology, Govt.Medical College Srinagar- J&K, Kashmir- India

Correspondence to : Dr. Shaista M Hamdani, Senior Resident, Department of Pathology, Government Medical College, Srinagar 190010, India



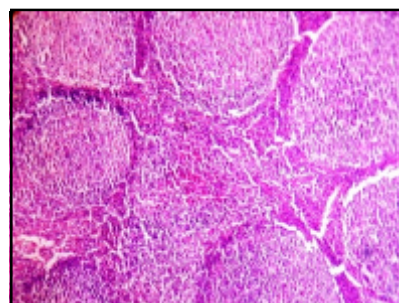
**Fig.1 Scanner View Photomicrograph of Lymph Node Showing Follicular Lymphoid Hyperplasia with Progressively Transformed Germinal Centers (H & E, 10X)**

transformed germinal centres (Fig 1). There were intranodal and perinodal eosinophilic infiltrates (Fig 2) with occasional areas of eosinophilic microabscesses (Fig 3). The interfollicular region showed fibrosis and proliferating vessels. The endothelial cells of these vessels were not epithelioid or histiocytoid, as is usually seen in angiolymphoid hyperplasia with eosinophilia but were plump endothelial cells resembling high endothelial or post capillary venules of lymph nodes. A diagnosis of Kimura's disease was made based on the marked peripheral eosinophilia and histopathologic findings. A serum Ig E was obtained, which was markedly elevated at 982 IU/ml (normal 0-87 IU/ml), further supporting the diagnosis of Kimura's disease.

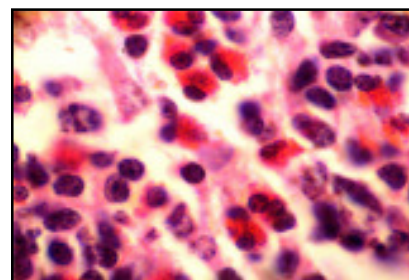
### Discussion

Kimura's disease was first described in China in 1937 by Kim and Szeto (5). However, the entity became more widely known as Kimura's disease after a systematic description in 1948 by Kimura *et al.* (1) Young and middle-aged Asian males of Chinese and Japanese origin are primarily affected (1,3). The disease typically presents with insidious onset of painless subcutaneous masses with adenopathy in the head and neck region (2). The disease usually involves subcutaneous tissues, lymph nodes (periauricular, axillary, and inguinal), parotid and submandibular salivary glands, (6) and rarely, oral mucosa (7). Other unusual sites of involvement include the auricle, (8) scalp, and orbit (9). This case presented here has two unusual clinical features: the age at onset (sixty years old) and the location of the lesion (mesenteric lymph node).

The clinical course of Kimura's disease is generally benign and self-limited. Kimura's disease may be complicated by renal involvement. Proteinuria may occur in 12% to 16% of cases (10). Nephrotic syndrome is the



**Fig 2. Prominent Reactive Appearing Germinal Centers are seen within this Hyperplastic Lymph Node with a Focus of Dense Intranodal Eosinophilic Infiltrate (H & E, 40X)**



**Fig 3. Photomicrograph Showing Eosinophilic Microabscess in the Intranodal Location (H & E, 100X)**

most common presentation (10); a wide spectrum of histologic lesions such as minimal change disease or mesangioproliferative glomerulonephritis, focal segmental glomerulosclerosis, membranous nephropathy, IgM nephropathy, and IgA nephropathy have been described (11). Our patient has normal renal function and no evidence of proteinuria. The lesions of Kimura's disease usually precede or coincide with the development of renal disease; occasionally, Kimura's disease may present with renal involvement before the appearance of subcutaneous lesions leading to delayed diagnosis (11). Widespread disseminated intravascular thrombosis is also reported in literature, affecting mesenteric and renal veins (thrombotic storm) (12).

The cause and pathogenesis of Kimura's disease is unclear, although it might be a self-limited allergic or autoimmune response triggered by an unknown stimulus. It has been speculated that a viral or parasitic trigger may alter T-cell immunoregulation or induce an IgE-mediated type 1 hypersensitivity resulting in the release of eosinophil-trophic cytokines (3,11). Abundant expression of eosinophil-trophic cytokines such as IL-4, IL-5, and IL-13 in peripheral blood mononuclear cells has been reported recently in a patient with Kimura's disease (13). This suggests that these cytokines may have a role in pathogenesis. High levels of circulating



eosinophilic cationic protein and major basic protein and high tissue IgE concentrations also have been found in the active stage of Kimura's disease (13).

The constant histologic features which are seen in this disease 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 paracortex, vascularization of the germinal centers, proteinaceous deposits in germinal centers, necrosis of germinal centers, eosinophilic abscesses, and atrophic venules in sclerotic areas (3).

Kimura's disease can also be confused with angiolymphoid hyperplasia with eosinophilia (ALHE) (4,15). ALHE is a rare but distinctive vascular tumor typically presenting in women during early to mid-adult life. Lymphadenopathy is uncommon, and blood eosinophilia is noted in <10% of cases (14). Histologically, the presence of inflammation around medium-sized arteries or veins and evidence of vascular damage (florid fibrointimal proliferation and cuboidal to dome-shaped endothelial cells) are key features in differentiating ALHE from Kimura's disease (14,15). The presence of reactive lymphadenopathy with similar-appearing inflammatory infiltrates is very common in Kimura's disease. By contrast, ALHE is generally a well-circumscribed subcutaneous vascular neoplasm in which the vessel proliferation is more florid, and has very plump, epithelioid-appearing endothelial cells, which may even mimic glandular structures. Although the vascular tumor is associated with inflammatory infiltrates, the presence of reactive germinal centers and eosinophilic microabscess formation are uncommon features of ALHE, as is an associated reactive lymphadenopathy (15).

The treatment of choice is said to be surgical excision (16). Although many alternative treatments, including radiation, high-dose intralesional steroids, and vinblastine have been attempted with good response; however, the tumors usually recur after discontinuing treatment or surgery. Some investigators have reported satisfactory results with cyclosporine (17) and pentoxifylline (16).

### Conclusion

The present report highlights the need for increased awareness by all clinicians about the unusual clinical presentation of Kimura's disease. The early diagnosis of this disease may spare the patient from potentially harmful and unnecessary invasive diagnostic procedures.

### References

1. Kimura T, Yoshimura S, Ishikawa E. Unusual granulation combined with hyperplastic changes of lymphatic tissue. *Trans Soc Pathol Jpn* 1948;37:179-80
2. Armstrong WB, Allison G, Pena F, Kim JK. Kimura's disease: two cases and a literature review. *Ann Otol Rhinol Laryngol* 1998;107:166-71
3. Jani A, Coulson M. Kimura's disease: Atypical case of a rare disorder. *West J Med* 1997;166:142-44
4. Thomas J, Jayachandran NV, Chandrasekhara PK, Rajasekhar L, Narsimulu G. Kimura's disease-an unusual cause of lymphadenopathy in children. *Clin Rheumatol* 2008;27:675-77
5. Kim HT, Szeto C. Eosinophilic hyperplastic lymphogranuloma, comparison with Mikulicz's disease. *Chin Med J* 1937;23:699-700
6. Tham KT, Leung PC, Saw D, Gwi E. Kimura's disease with salivary gland involvement. *Br J Surg* 1981;68 :495- 97
7. Hongcharu W, Baldassano M, Taylor CR. Kimura's disease with oral ulcers: response to pentoxifylline. *J Am Acad Dermatol* 2000;43 :905- 07
8. Chan KM, Mok JS, Ng SK, Abdullah V. Kimura's disease of the auricle. *Otolaryngol Head Neck Surg* 2001;124 : 598- 99
9. Buggage RR, Spraul CW, Wojno TH, Grossniklaus HE. Kimura disease of the orbit and ocular adnexa. *Surv Ophthalmol* 1999;44 :79- 91
10. Atar S, Oberman AS, Ben-Izhak O, Flatau E. Recurrent nephrotic syndrome associated with Kimura's disease in a young non-Oriental male. *Nephron* 1994;68 :259- 61
11. Rajpoot DK, Pahl M, Clark J. Nephrotic syndrome associated with Kimura disease. *Pediatr Nephrol* 2000;14:486-88.
12. Saini A, Singh V, Chandra J, Dutta AK. Kimura's disease: An unusual glandular involvement with blood and tissue eosinophilia. *Indian J Pediatr* 2009;76:647-48
13. Katagiri K, Itami S, Hatono Y, Yamaguchi T, Takayasu S. In vivo expression of IL-4, IL-5, IL-13 and IFN-gamma mRNAs in peripheral blood mononuclear cells and effects of cyclosporin A in a patient with Kimura's disease. *Br J Dermatol* 1997;137:972- 77
14. Chun S, Ji HG. Kimura's disease and angiolymphoid hyperplasia with eosinophilia: clinical and histopathological differences. *J Am Acad Dermatol* 1992;27 :954- 58
15. Seregard S. Angiolymphoid hyperplasia with eosinophilia should not be confused with Kimura disease. *Acta Ophthalmol Scand* 2001;79 :91- 93
16. Hongcharu W, Baldassano M, Taylor CR. Kimura's disease with oral ulcers: response to pentoxifylline. *J Am Acad Dermatol* 2000;43:905-907
17. Wang YS, Tay YK, Tan E, Poh WT. Treatment of Kimura's disease with cyclosporine. *J Dermatolog Treat* 2005;16: 242-44