CASE REPORT

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

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Abstract
Kimura's disease is a rare chronic inflammatory disorder of unknown cause primarily seen in young Asian males. The disease 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. 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 138x10^9/L, and white cell count of 7.9x10^9/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 mesentry. 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...
transformed germinal centres (Fig 1). There were
intranodal and perinodal eosinophilic infiltrates (Fig 2)
with occasional areas of eosinophilic microabcesses (Fig 3).
The interfollicular region showed fibrosis and
proliferating vessels. The endothelial cells of these vessels
were not epitheloid or histiocytoid, as is usually seen in
angioymphoid 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-
age 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

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
eosinophilotropic cytokines (3,11). Abundant
expression of eosinophilotropic 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**