

## Case Report

### A case report of Kimura disease

Dr. Smita Pathak, Dr. Sneha Joshi. Dr. Pooja Banwaskar. Dr. Dhanashree Muley

Name of the Institute/college: MIMER Medical College, Talegaon Dabhade Pune

Corresponding author : Dr. Smita Pathak

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#### Abstract

Kimura disease is a rare chronic inflammatory disorder of unknown etiology primarily seen in young Asian males. It is a benign disease with an indolent course, gradually increasing in size over months or years. It usually presents as a mass lesion in the subcutaneous tissue of the head and neck region or major salivary glands area often associated with regional lymphadenopathy. In some cases lymph node enlargement is the only manifestation of the disease. It has no potential for malignant transformation. Spontaneous involution is rare. The main concern in this disease is its capacity to grow and cause disfigurement.

We report a case in a 40yr old male who presented with bilateral neck swellings diagnosed clinically as lymphadenopathy . The gross specimen received in histopathology department measured 2.5cm X 2cm. The histopathological examination of excised mass was diagnostic of Kimura disease.

**Key words** : benign, bilateral, subcutaneous tissue , kimura

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#### Introduction

Kimura disease is a rare chronic inflammatory disorder of unknown aetiology which is commonly seen in Asian population.<sup>1</sup> It usually presents as a mass lesion in the subcutaneous tissue of head and neck region or major salivary glands.<sup>2</sup> For years Kimura disease was believed to be or part of the same disease spectrum as Angiolymphoid hyperplasia with eosinophilia (ALHE). Recent reports however have confirmed that the two are in fact separate entities.<sup>1</sup>

The case is reported for it's rarity.

#### Case report

A 40 year old male patient presented with bilateral painless neck swellings since 4 months. He had no history of cough, low grade fever, fatigue or loss of weight.

On examination right & left side cervical swellings measured 3x2x2 cm & 2x1x1 cm respectively. Both the swellings were mobile & non tender.

Systemic examination showed no significant abnormality. His blood counts revealed eosinophilia (10%) with an absolute eosinophil count of 800/cmm.

Clinically the diagnosis of bilateral cervical Lymphadenopathy was made.

Both the masses were excised surgically. The specimens were received in the department of Pathology. Gross examination showed specimens measuring 2.5x2x0.5cm and 1.5x1x1 cm, & irregular in shape. Cut section appeared grayish white & lobulated.

Histopathological study demonstrated capsule beneath which were seen hyperplastic lymphoid follicles with prominent germinal centres. ( Fig No 1) Extensive infiltration by mature eosinophils was seen forming eosinophilic microabscesses at places. (Fig No 2 & 3) Numerous thin walled blood vessels with flattened endothelial lining

were noted. (Fig No 4) Some of the blood vessels appear hyalinised.

Considering above histological findings the diagnosis of Kimura disease was offered.

#### **Discussion**

Kimura disease was first reported by Chinese authors Kimm & Szeto in 1937 with the term eosinophilic hyperplastic lymphogranuloma.<sup>3</sup> The disorder received its current name in 1948 when Kimura et al noted the vascular component & referred to it as an unusual granulation tissue combined with hyperplastic changes in lymphoid tissue.<sup>4</sup> The other synonyms of the disorder include eosinophilic granuloma of soft tissue, eosinophilic hyperplastic lymphogranuloma, eosinophilic lymphofolliculosis, eosinophilic lymphofollicular granuloma & eosinophilic lymphoid granuloma.<sup>3</sup>

Most cases of Kimura disease are found in Asians & it is endemic in the far East. Young males are commonly affected, the median age being 28-32 years.<sup>3,4,5</sup>

It usually presents as a mass lesion in the subcutaneous tissue of head & neck region or the major salivary glands, often associated with regional lymphadenopathy. Sometimes lymph node enlargement is the only manifestation of the disease.<sup>2</sup> The patient may present with characteristic triad of painless slowly enlarging soft tissue mass, blood & tissue eosinophilia and markedly increased serum Immunoglobulin E.<sup>5</sup> Periauricular, parotid & submandibular regions are commonly affected. Rare sites of involvement include kidneys, orbits, ears, spermatic cord & nerves. Involvement of kidneys can cause nephrotic syndrome.<sup>3,6</sup> Pathophysiology of Kimura disease is unknown, although an allergic reaction, trauma & an autoimmune process have all been implicated as the possible causes.<sup>4</sup>

Histopathological study is recommended to confirm the diagnosis. Lymphoid follicles with

prominent germinal centres characterize Kimura disease. Marked eosinophilic infiltration sometimes forming eosinophilic microabscesses along with capillary proliferation forming canalized vessels with flat endothelium and fibrosis surrounding the lesion are the hallmarks of the disease. Hyalinised vessels are often seen in the paracortical region.<sup>2,3</sup>

Differentiating Kimura disease from ALHE requires a strict analysis of clinical & histologic features because the diseases are similar & were once thought to be same disorder. Both the diseases usually present with soft tissue masses in head & neck region, but in ALHE the lesions are mostly dermal or subcutaneous & not found in lymph nodes, which is a common location for Kimura disease.<sup>6</sup> Clinically Kimura disease is believed to be a disease of the Asians & ALHE one of the western world. ALHE occurs in older, predominantly female population. Kimura disease is primarily a disease of young males.<sup>3</sup> Regional lymphadenopathy, peripheral blood eosinophilia & raised serum Ig E levels which are characteristic of Kimura disease are rarely seen in ALHE.<sup>1</sup> The histology of ALHE is typified by an exuberant proliferation of capillary vessels some of which may not be canalized. These are lined by epithelioid or histiocytoid endothelial cells which are not seen in Kimura disease. While there is an inflammatory infiltrate, the associated lymphocytes are not arranged in germinal centres as they are in Kimura disease. Tendency towards renal involvement & nephrotic syndrome is restricted to Kimura disease & not observed in ALHE.<sup>1</sup>

Spontaneous resolution, though rare, has been reported. Most patients have a prolonged course with slow enlargement of the masses.<sup>6</sup> Surgical excision is the treatment of choice to preserve cosmesis & function. However recurrence rate can be as high as 25%. Other treatment modalities include conservative treatment with the aim of

spontaneous involution, intra lesional or oral steroids, radiotherapy, cryotherapy, laser & cytotoxic agents.

However the overall prognosis of Kimura disease is reported to be good.<sup>3,6</sup> Our patient was an Asian male with bilateral cervical lymphadenopathy. There was peripheral blood eosinophilia. Histopathologically the lymph nodes showed hyperplastic lymphoid follicles with prominent germinal centers, dense eosinophilic infiltration & proliferation of post capillary venules. These features guided us towards a definitive diagnosis of Kimura disease. The patient is on regular follow up and has not shown any evidence of recurrence till now.

#### Conclusion

Kimura disease is a locally disfiguring disease with indolent course, whose true importance lies in it's gradually increasing in size over months or years. A high index of suspicion is needed to diagnose this condition. .

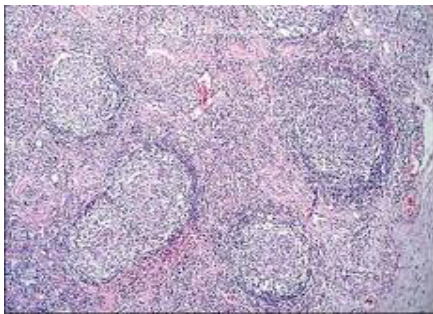


Fig No 1 Photomicrograph showing hyperplastic lymphoid follicles with prominent germinal centres (H & E,5 X)

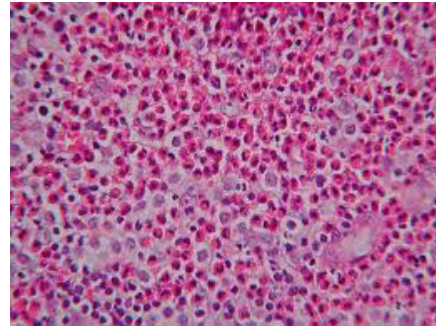


Fig No 2 Photomicrograph showing dense eosinophilic infiltration (H & E,5 X)

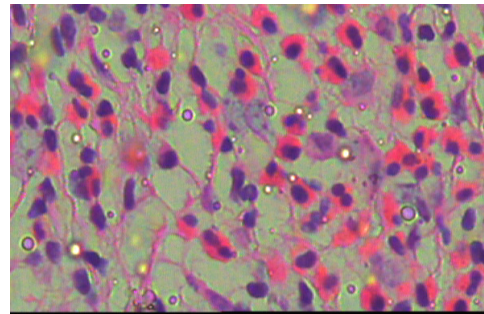


Fig No 3 Photomicrograph showing dense eosinophilic infiltration (H & E,10 X)

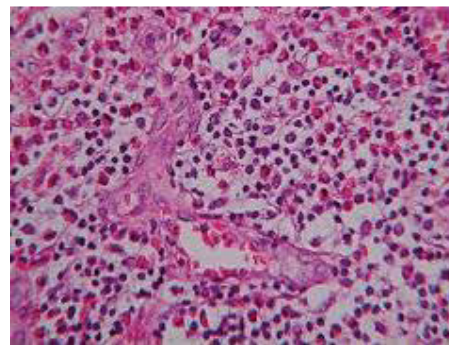


Fig No 4 Photomicrograph showing proliferation of post capillary venules (H & E,5 X)

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