

A rare case of inguinal kimura disease

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SUMMARY

Kimura's disease is a rare chronic inflammatory disease of unknown etiology, commonly presenting with painless lymphadenopathy and subcutaneous masses in the head and neck regions.¹ However, presentations with inguinal lymphadenopathy are rare and mimics other differentials, may pose a diagnostic challenge.

We present a case of a 50-year-old male, with right inguinal swelling for one month duration that was finally diagnosed with Kimura's Disease after a multitude of investigations to rule out differentials of lymphadenopathy, delaying conclusive treatment.

Specialized test had been done resonated with the histopathological findings only. We report this case to increase awareness of Kimura's Disease.

INTRODUCTION

Kimura's disease (KD) is a benign, chronic inflammatory soft tissue disorder of unknown origin. It predominantly affects young Asian men, uncommon among Caucasians and rare among Africans. There is a marked male predominance. The peak age of onset is during the third decade of life.¹ The disease usually presents with swelling and lesions in the head and neck regions involving the subcutaneous soft tissue, major salivary glands and lymph nodes. Presentations with inguinal lymphadenopathy however are rare, hence it may pose a diagnostic challenge to the assessing physician, especially in an area where tuberculosis is endemic.

It was first described in 1937 in Chinese literature by Kimm and Szeto and initially was recognized as "eosinophilic hyperplastic lymphogranuloma". The definitive histological description was published by Kimura et al. in Japan in 1948, henceforth the name. Ever since there has been a gradual increase in the number of reports of the disease.²

CASE REPORT

A fifty-year-old male with no co-morbidity, presented with right inguinal swelling for one month, gradually increasing in size. He was having on and off fever despite completed courses of antibiotics. History was otherwise unremarkable.

Clinical examination revealed a 4x4cm swelling over right inguinal region which was firm and mobile. The overlying skin was normal. He was hemodynamically stable

throughout and the remaining physical examinations were normal.

His laboratory assessment showed raised total white cell count ($16.3 \times 10^3/\text{UL}$) with normal hemoglobin (15.1 g/dL) but had raised eosinophilic count of 30%. Liver and renal functions and coagulation profile were normal. Test for HIV, Syphilis and gonorrhoea were negative. A chest radiograph excluded any suspicion of pulmonary tuberculosis features. Mantoux test and sputum samples for direct smear were also negative.

Ultrasound of bilateral inguinal regions noted several other lymph node enlargements, with the largest measuring 2.3 x 1.2 cm on right side. There was no focal mass or collection seen (Figure 1). Excisional biopsy of right inguinal lymph nodes was performed. The pathological examination of excised sample showed extensive infiltration of eosinophils in the paracortical and interfollicular areas without atypical mononuclear cells or Reed-Sternberg cells as confirmed by the negative CD30 stain (Figure 2). These findings were in keeping with KD. For confirmation of diagnosis, histopathological report was correlated with the results of specialized investigation such as raised Eosinophil Cationic Protein Test, 23.3 ug/L; Serum Ig E, 2090 kU/L and Absolute Eosinophils Count, 3300/UL. Specific IgE test was also carried out directed towards d1, d201, f24, d2, f23, f58; which all showed very low results.

Conservative management was opted with active surveillance for disease progression. He had been followed up for the last 6 months and is still relatively well without any medications and disease progression.

DISCUSSION

The etiology of KD still remains unknown although the presence of eosinophilia and elevated serum IgE levels suggest an allergic or hypersensitivity process.³ It may be easily mistaken initially for a malignant hematological disorder such as chronic lymphocytic leukemia or Hodgkin's disease until further investigations are carried out. However differentiation between KD and Angio-Lymphoid Hyperplasia with Eosinophilia (ALHE) instead has been a challenge for decades among histopathologist. They were once considered to be variations of the same disease. Nevertheless, differentiation between these two diseases carries little significance clinically as it does not alter the treatment strategies eventually.

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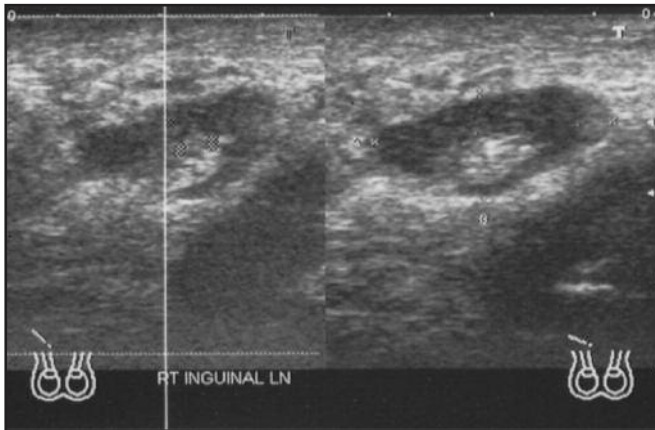


Fig. 1: Ultrasound images showing enlarged lymph nodes bilaterally.

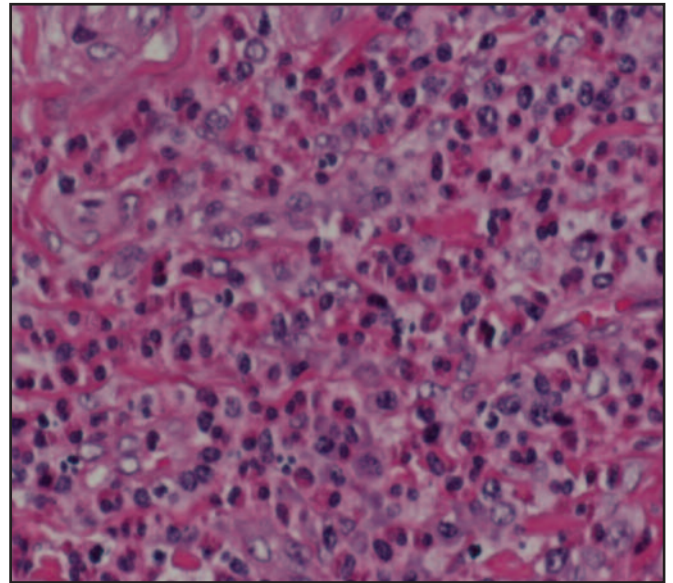


Fig. 2: Histological features from right inguinal lymph nodes demonstrating eosinophilic infiltration.

For the clinician, it is of paramount importance to exclude lymphoma in cases of lymphadenopathy. To add more complexity to the diagnosis, tissue eosinophilia is commonly seen in Hodgkin's disease and non-Hodgkin's lymphomas, especially of T-cell lineage. CD30 staining, which is also known as Ki-1 or Ber-H2, can be adopted by histopathologists to confirm diagnosis of anaplastic large cell lymphoma, classic Hodgkin's lymphoma, and embryonal carcinoma before confirming KD. In addition histological immunofluorescence tests would show germinal centers containing heavy IgE deposits and variable amounts of IgG, IgM, and fibrinogen.⁴

Ultrasound imaging should be the first foremost test performed in cases of lymphadenopathy. Lymph nodes in KD are hypoechoic, solid, and round or oval in shape with normal surrounding soft tissues. On radiological examination, KD may mimic other chronic and malignant diseases such as tuberculosis or lymphoma. It is almost impossible to exclude malignancy with imaging modality alone, therefore a histological confirmation is necessary.⁵ Although imaging studies are not diagnostic, they may help delineate the extent of disease.³

Treatment options for KD are divided into surgical, medical and radiotherapy. Primary prophylactic surgery is done as therapeutic or diagnostic purpose. It is recommended in those young patients with localized mass, primary or localized recurrence. Conservative treatment which includes steroid therapy is indicated in patient with renal involvement, localized mass and recurrent disease. Steroid therapy shows good outcome in decreasing the lymph node size besides reducing the renal symptoms. The last treatment choice in KD is radiotherapy, which is indicated in patients who fail

surgical or medical treatment or unresectable mass.⁵ Nevertheless, the optimal treatment regimens and long-term prognosis in patients with KD, especially when renal symptoms and signs are present, are largely unknown due to the rarity of these lesions and the lack of long-term follow-up data in the literature.¹ Watchful management was adopted for this patient due to the localized disease, absence of renal involvement and halted disease progression.

CONCLUSION

We report this case to increase awareness of this unusual manifestation of Kimura's disease, which may mimic other pathologies. However, the other more sinister etiologies of inguinal lymphadenopathy have to be excluded first. The continuum of cytological features and appropriate specialized test with a lead from peripheral eosinophilia helps in confident diagnosis of Kimura's disease.

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