

Kimura's disease presenting with inguinal lymphadenopathy in an 11-year old girl: A case report

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Abstract

Kimura's disease (KD), mostly seen in young Asian boys, typically manifests with subcutaneous lesions and cervical lymphadenopathy. It has a good prognosis and has no risk of malignancy. Inguinal lymphadenopathy is rarely reported as a presenting feature of KD. Here we report such a case of KD in an 11-year old girl. The diagnosis of KD was confirmed in this case by characteristic histopathological findings of the excised lymph node.

Keywords

Kimura disease, inguinal lymphadenopathy, children

Introduction

Kimura's disease (KD) is a rare chronic inflammatory disorder causing lymphadenopathy and subcutaneous lesions typically involving the head and neck region. Only about 200 cases of KD have been reported worldwide.¹ These patients mostly have blood and tissue eosinophilia, and raised IgE levels. Although the prognosis of KD is generally good, rarely there may be renal and vascular complications, described as a thrombotic storm.² Therefore, proper follow-up is required in these cases.

Case

An 11-year old girl presented with swelling in the left inguinal region for 1 year. She neither had any history of fever, weight loss, bleeding manifestations, nor a family history of tuberculosis.

On examination, she was afebrile, with normal growth and blood pressure. There was no pallor, clubbing and oedema. Systemic, general examination was normal, apart from a mobile, firm, non-tender, lymph node of 4 × 4 cm with normal overlying skin in the left groin. Other groups of lymph nodes were not enlarged. There was specifically no hepatosplenomegaly.

Blood counts revealed eosinophilia (15%), and an erythrocyte sedimentation rate of 10 mm/h. Peripheral blood smear examination was within normal limits

except for an eosinophilia. Renal and liver function tests were normal. Serum IgE was 580 IU/mL (normal, approximately 23.6 IU/mL). HIV Elisa and filarial serology were negative. Urine and stool examinations were normal. Skin tuberculin test and gastric aspirate for AFB were negative. The chest radiograph was normal.

Histopathological examination of the excised inguinal lymph node showed multiple follicles of variable sizes throughout the cortex and medulla with prominent germinal centres. Capsule and pericapsular tissue showed infiltration by eosinophils and lymphocytes (Figure 1). Many hyalinised proliferating vessels were also seen. The interfollicular region showed focal collection of eosinophils and lymphocytic proliferation and plasma cells (Figure 2).

After approximately 1 month following excision of inguinal node, she developed a firm, non-tender and

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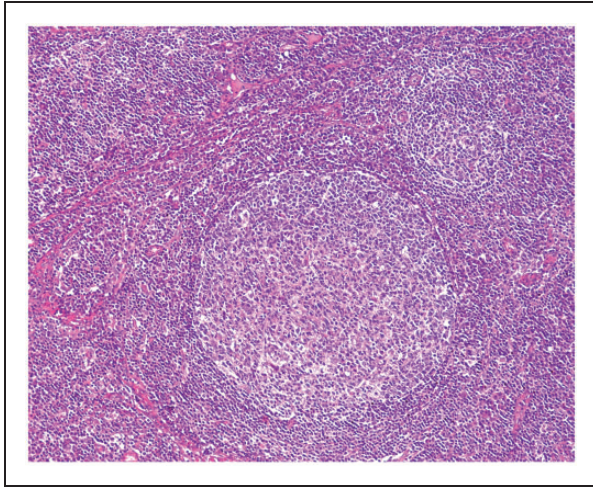


Figure 1. (Haematoxylin & Eosin $\times 100$) Inguinal lymph node biopsy. Note the multiple follicles of variable sizes with prominent germinal centres and diffuse infiltration by eosinophils, lymphocytes and plasma cell.

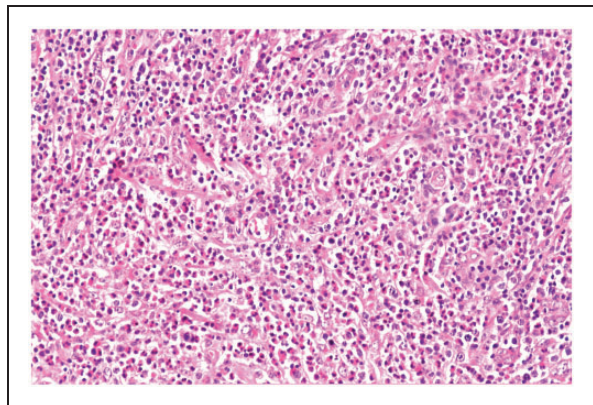


Figure 2. (Haematoxylin & Eosin $\times 200$) Inguinal lymph node biopsy. Note several proliferating and hyalinised vessels. Interfollicular region showing focal collection of eosinophils.

mobile left side axillary lymphadenopathy of 2.5×2 cm size. Fine-needle aspiration cytology (FNAC) examination revealed lymphoid hyperplasia and significant number of eosinophils. Considering the recurrence of KD, treatment with oral Prednisolone (2 mg/kg/day) for 4 weeks was commenced. The lymph node swelling subsided, and steroid therapy was tapered off over the following 4 weeks. No new lesions have appeared on regular follow-up.

Discussion

KD was first described in China in 1937 by Kimm and Szeto.³ KD typically manifests as non-tender subcutaneous masses in the head and neck region,

with cervical lymphadenopathy.⁴ Viswanatha reported 18 patients of KD; all of them presented with painless swelling in head and neck region.⁵ Chen *et al.* reported only three cases of KD with inguinal lymphadenopathy out of 21 cases.⁶ The index case demonstrated inguinal and axillary lymphadenopathy without any subcutaneous lesion, which is rare.

Cutaneous eosinophilic vasculitis a rare condition of skin is reported in a few patients of KD.⁷ Renal function should be monitored as KD is associated with renal abnormalities, specially nephrotic syndrome, in about 20% cases.⁸ Its aetiology is thought to be an abnormal immune response to an unidentified antigenic stimulus.

The histopathologic features identified with KD are florid germinal centres, eosinophilic infiltration, eosinophilic abscesses, sclerosis in the germinal centres and paracortex, and vascularisation of the germinal centres.^{4,9} Typical histopathological findings of the excised lymph node were found (Figures 1 and 2).

FNAC is found to be helpful in some cases, but definitive diagnosis can be obtained by histological examination of the excised lesion.¹⁰ Excisional biopsy is preferred for the initial diagnosis as it helps in the exclusion of malignant lymphoma, angiolymphoid hyperplasia with eosinophilia and other reactive lymphadenopathies.

The differential diagnosis includes: (1) Hodgkin's lymphoma, which may have lymph nodes infiltrated by eosinophils, but Reed-Sternberg cells should be visible. Eosinophilia is found in around 15% patients of Hodgkin's disease;¹¹ (2) myeloid leukaemia, which may have an increase in eosinophils, but has metamyelocytes present; (3) angiolymphoid hyperplasia (characterised by florid vascular proliferation with atypical endothelial cells surrounded by a lymphocytic and eosinophilic infiltrate), which is associated typically with subcutaneous lesions and is usually seen in middle-aged women rather than young men.¹² Histopathologically KD and angiolymphoid hyperplasia are difficult to differentiate. However raised IgE levels and lymphadenopathy are typically associated with KD; (4) Sarcoidosis, presents with hilar lymphadenopathy, pulmonary infiltration, and skin and ocular lesions. Peripheral lymphadenopathy (inguinal and cervical) is also reported. The diagnosis can be established on the basis of clinical and radiologic findings, supported by histologic evidence of non-caseating epithelioid-cell granulomas.¹³

Other causes of eosinophilia are typically due to parasitic infestation such as trichinella or schistosoma, or hypersensitivity reactions to food, drugs or asthmatic stimuli, and these should be looked for.

Surgical excision and systemic steroid therapy is the treatment of choice for KD. Recurrence is frequent after the cessation of treatment. Local radiotherapy

can be considered for refractory lesions. Prognosis of KD is found to be good with no malignant transformation.² Although KD is a rare disorder, it should be considered in the differential diagnosis of any eosinophilic-related lymphadenopathy.

Declaration of conflicting interests

None declared.

Funding

This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

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