

Kimura Disease : A Rare Case Report of 2 Cases Diagnosed on FNAC with Review of Literature

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Abstract

Kimura disease is a chronic inflammatory disease, which frequently affects middle-aged Asian men, although children are seldom affected by it. This may rarely be encountered in children, and the knowledge of this fact is essential to rule out the remote possibility of Kimura's disease in children with a slow-growing painless mass in the head and neck region. Therefore, the characteristics of the Kimura disease of childhood have not been well illustrated. Here we are reporting 2 cases of Kimura disease in 17 years & 22 years old female. (Head and neck are the most frequently involved sites of subcutaneous masses). On laboratory investigations, in both the patients are having peripheral blood eosinophilia and elevated immunoglobulin E level were seen. Clinicians need to be aware of this disease when dealing with patients with lymphadenopathy. A standard and effective treatment protocol would improve the outcome of the Kimura disease. Here we report a case of Kimura disease in rare age group.

Keywords

Kimura's disease, Eosinophilia

Introduction

Kimura disease (KD) is a chronic inflammatory disorder of unknown etiology, that most commonly present with painless, unilateral cervical lymphadenopathy. First described by Kimm and Szeto [1] in 1937 as 'eosinophilic hyperplastic lymphogranuloma', it gained prominence as Kimura's disease following a report by Kimura & coworkers in 1948, which elaborated on an 'unusual granulation combined with hyperplastic changes in lymphoid tissue'[2]. Kimura's disease primarily involves the head and neck region, presenting as deep, subcutaneous masses in the preauricular region, forehead or scalp[3]. According to previous medical literatures, kimura disease has a high recurrence rate so early and definite diagnosis of disease is vital for effective treatment plan 4 . As a triad of painless subcutaneous masses in the head and neck region, it is characterised by blood and tissue eosinophilia with clearly elevated serum immunoglobulin E (IgE) levels. The optimal treatment for KD remains controversial.

Case Report 1

A 17 years old female presented in the ENT OPD with chief complaints of right side cervical painful swelling since one year (Figure 1a). The patient did not complain of any increase in the size of swelling, reduction in salivary flow or pus discharge. There was no history of weight loss, low grade fever or night sweating. There was no history for dental treatment that reveals any incidence of tooth-related pain or space infection. Medical, surgical and family histories were non-contributory. She had normal development. Patient was investigated routinely & all the biochemical investigations were found to be normal except eosinophilia & increased serum IgE levels.

Case report 2

A 22 years old female presented in the medicine OPD with chief complaints of left sided posterior cervical swelling for 6 months (Figure 1b). Head and neck examination revealed a 4x3 cm, firm, mobile, non tender mass over the left posterior cervical swelling. Other systemic examination was unremarkable. There was no symptoms to suggest pulmonary tuberculosis such as chronic cough, night sweats, anorexia or weight loss. There was no axillary or inguinal lymphadenopathy or hepatosplenomegaly. Initial lab investigations revealed normal indices of full blood count with slight eosinophilia with rise in serum IgE levels.

FNAC was performed in both the cases which showed moderate cellularity consisting of population of lymphocytes, immunoblasts & large number of eosinophils (Figure 2a & 2b). A provisional diagnosis of Kimura's disease was made. Biopsy was advised for confirmation of diagnosis. Subsequently excision biopsy was done in both the cases which confirmed

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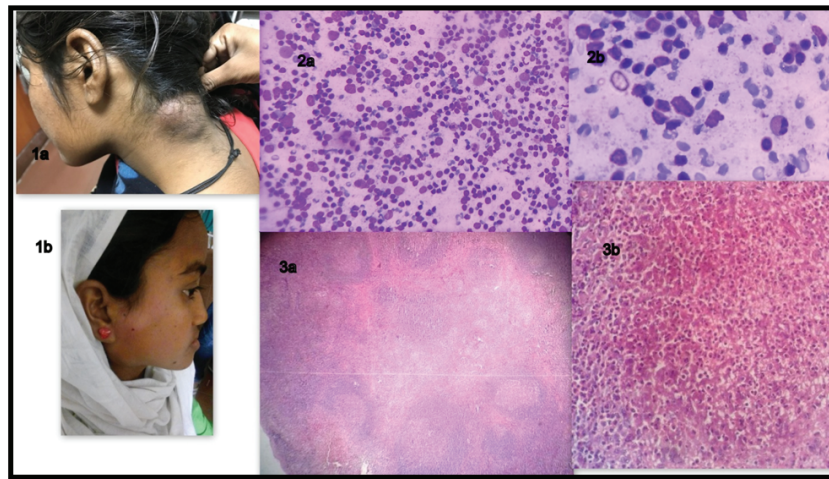


Figure 1a & b: Clinical picture of both the patient with right sided cervical swelling and left posterior cervical swelling.

Figure 2a & b: Photomicrograph shows polymorphous population of lymphocytes in various stages along with numerous eosinophils. (MGG 100X)

Figure 3a & b: Photomicrograph shows Eosinophils which were prominent in interfollicular areas in scattered lymphocytes and plasma cells. Eosinophilic folliculolysis was seen. (H&E 100X, 400X)

the diagnosis of Kimura's disease. The excised specimen measures 3.0x2.0x1.6 cm & in size and on cut section was pale gray. The microscopic examination revealed several reactive lymphoid follicles. Numerous thin-walled vessels were present, often grouped in small foci, their endothelial lining was flattened with no epithelioid or vacuolated cells. Eosinophils were prominent in interfollicular areas in scattered lymphocytes and plasma cells. Eosinophilic folliculolysis was seen (Figure 3a & 3b). The final histopathological diagnosis of Kimura's disease was given in both the cases. CD1A marker was negative on immunohistochemistry (positive result is an indicator of langerhan cell histiocytosis). Both the patient did not get any complain after surgery during follow up.

Discussion

Kimura's disease or eosinophilic hyperplastic lymphogranuloma is a chronic inflammatory disease. It is difficult to differentiate from angiolymphoid hyperplasia with eosinophilia (ALHE). The Kimura's disease is a primary inflammatory process with secondary vascular proliferation whereas ALHE is primary arteriovascular malformations with secondary inflammation. It is mainly seen on the head and neck region. The lesion is benign but might be confused with malignant lesions. Kimura's Disease is often seen in the second and third decades of life. Patients with Kimura's disease have been shown to have high levels of circulating eosinophilic cationic protein and major basic protein, with heavy concentrations of IgE in their tissues. Allergic or parasitic aetiologies for Kimura's disease have been actively sought, but not identified. The clinical course of Kimura's disease is benign.

The differential diagnosis for Kimura's disease includes such entities as eosinophilic granuloma, Mikulicz's disease, acute non-lymphocytic leukaemia, Hodgkin's disease, follicular lymphoma, angioimmunoblastic lymphadenopathy, and angiolymphoid hyperplasia with eosinophilia. Except for angiolymphoid hyperplasia with eosinophilia, the clinical and histological features of these diseases easily distinguish them from Kimura's disease. The absence of Reed-Sternberg cells helps to exclude Hodgkin's disease. Unfortunately, T cell lymphomas can present with polymorphonuclear lymphocytes and eosinophilia, making the distinction difficult. Although atypical, histiocytosis-X can present with subcutaneous masses, the diagnosis is made by finding the characteristic abnormal histiocytes and detecting CD1A marker. Angiolymphoid hyperplasia with eosinophilia is more typically seen in middle-aged women and Kimura's disease in younger men [5]. Immunoperoxidase studies show IgE reticular networks in germinal centers.

There is no consensus on the management of Kimura's disease. Surgery is performed as a therapeutic/diagnostic procedure.

Conservative treatment includes oral steroids, cyclosporine & leflunomide which are reported to be responsible for decreasing size of the enlarged lymph nodes, but there is no evidence of reduction of the affected salivary gland size. The prognosis of Kimura's disease is good, no malignant transformation is observed [5-7]. Widespread disseminated intravascular thrombosis is also reported in literature, affecting mesenteric and renal veins (thrombotic storm)[6]. So follow up of these cases is must. The clinical course of Kimura disease is often progressive, and the main problem concerning treatment is disease recurrence.

Conclusion

Although it is very rare, nevertheless it shows the universal distribution of this disease. The goal of its diagnosis is to treat it & to prevent morbidity & complications. The diagnosis of Kimura disease should not be a problem when combined with the results of a pathological examination, together with markedly high levels of eosinophils in peripheral blood and serum IgE. Correlation with these clinico pathological findings is essential as differentiation on imaging alone may be difficult. Follow up is needed in every case.

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