

Kimura's Disease: A Rare Cause of Postauricular Swelling

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ABSTRACT

Introduction

Kimura's Disease is a chronic inflammatory disorder of lymph node which is very rare in Indian population.

Case Report

A 15 year old boy with multiple postauricular swelling for 18 months presenting in OPD and diagnosed having eosinophilia. Then excision biopsy was taken, which indicates Kimura's Disease. Patient was treated with high dose of corticosteroid.

Conclusion

Kimura's disease, though rare should be kept in mind for treating a patient with lymphadenopathy with eosinophilia or high IgE level, because it can spare the patient unnecessary invasive procedure.

Keywords

Kimura Disease; Eosinophilic Lymphadenitis; Lymphadenopathy

Kimura's disease is a rare, benign, chronic inflammatory disorder of unknown aetiology that involves lymph nodes and subcutaneous tissue of head and neck region. The condition is clinically characterized by triad of painless subcutaneous masses, blood and tissue eosinophilia and markedly increased serum IgE concentrations. The disorder is primarily seen in East Asian male patients in the third decade of life, though rare in Indian. Although it may mimic a neoplastic condition, early correct diagnosis of Kimura's disease may spare the patient from unnecessary invasive diagnostic procedure. We present such a case in a 15 year old boy who presented with bilateral postauricular swelling and focus on treatment modalities in our discussion.

Case Report

A 15 year old boy was referred to our outpatient department for workup for multiple bilateral postauricular subcutaneous swelling for 18 months. He was seen by his primary care physician who treated him with different types of oral antibiotics for many weeks. There was no history of fever, sore throat, cough, ear discharge or weight loss. Physical examinations showed an apparently well boy with multiple distinct palpable

masses at the both postauricular areas which were non-tender, firm, non-fluctuant and freely mobile (Fig. 1). The largest discrete mass was 2.5 cm in diameter. The rest of the general physical and systemic examinations were normal.

Peripheral blood smear showed 11% eosinophils with absolute eosinophil count of 950/cumm. Chest radiograph was within normal limits. Tuberculosis skin test and rapid Infectious mononucleosis assay were negative.

Because of concern that the mass might be some lymphoproliferative disorder or neoplasm, an excision biopsy was taken. Histopathological examination of the mass showed eosinophilic infiltration and vascular proliferation with prominent endothelial cells within the extra nodal soft tissue associated with lymphoid hyperplasia and germinal centre formation. Serum IgE levels were estimated which showed - 5300 KIU/ml (Normal - 150 KIU/ml) further supported the diagnosis of Kimura's Disease.

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Fig.1. Postauricular swelling of a 15 year old boy

After 6 months follow up, the mass began to recur and then treatment with oral prednisolone was started at a dose of 40mg/day for the initial period of 6 week and subsequently tapered every week by 10 mg/day over next 4 weeks. After 4 weeks of treatment eosinophilia was decreased and became well within normal limits in 6 weeks. IgE levels fell to 144 KIU/ml gradually. There was no local recurrence at 9 months follow up.

Discussion

Kimura's Disease was first described by Kim and Szeto¹ in Chinese literature in 1937, but was later characterized by Kimura et al in 1948 in a manuscript titled "On the unusual granulation combined with hyperplastic changes of lymphatic tissue."² It is now recognized as a benign cause of painless localized lymphadenopathy of Asians, especially boys of Chinese and Japanese origin.³ The clinical triad of subcutaneous nodule in head and neck region, prominent peripheral eosinophilia and highly elevated IgE, particularly when seen in an Asian male [in 2nd and 3rd decades of life, although can become apparent at any age] is highly suggestive of Kimura's Disease.

The clinical course of Kimura's Disease is benign and self-limited. The subcutaneous masses or adenopathy are usually found in head and neck region with occasional

pruritus of overlying skin. It sometimes affects axillary, periauricular, inguinal or epitrochlear node.⁴ Most patients have a prolonged course with slow enlargement of masses. Occasional spontaneous resolution is known. These lesions do not have any malignant potential. Kimura's Disease may be complicated by renal involvement. In case of renal involvement, nephrotic syndrome is the commonest presentation, proteinuria may occur in 12 - 16 % of cases.⁵ But in our case, there was normal renal function and there was no evidence of proteinuria.

The pathophysiology of Kimura's Disease is not well understood at this time, but may relate to a disturbance in the normal rate of product of an interaction between types 1 and 2 helper cells. Such a derangement could result in excessive elaboration of eosinophilotropic cytokines.⁶ These patients have been shown to have high levels of circulating eosinophilic cationic protein and major basic protein with heavy concentration of IgE in their tissues.⁷ Allergic or parasitic etiologies for Kimura's Disease have been actively sought, but not proven.

The pathology of Kimura's Disease is characterized by prominent germinal centres in involved lymph nodes containing cellular, vascular and fibrous components. The cellular component consists of dense eosinophilic infiltrate in background of abundant lymphocytes and plasma cells, eosinophilic micro-abscesses with central necrosis.^{8,9} Other characteristics may include sclerosis, proteinaceous material and / or vascularity in the germinal centres and polykaryocytes of Warthin-Finkeldey type.¹⁰

The lesions can be further investigated with ultrasonography, CT scan and MRI. But their radiographic features are not pathognomonic. The diagnosis is made only by histopathological examination of excisional biopsy specimen of lesion.

The differential diagnosis of Kimura's Disease includes other entities such as Tuberculous adenitis, Mikulicz's Disease, Eosinophilic Granuloma, ALHE (Angiolymphoid Hyperplasia with Eosinophilia), Cylindroma, Pyogenic granuloma. It should also mimic some serious disorder like Hodgkin's Disease, Acute non-lymphocytic leukemia, follicular lymphoma

and Kaposi's sarcoma. Except for Angiolymphoid Hyperplasia with Eosinophilia, the clinical and histological features of these diseases easily distinguish them from Kimura's Disease.

There is no consensus on management of Kimura's Disease. Various treatment modalities have been tried with variable success. Three major therapeutic options exist for Kimura's Disease. Excision of the mass remains the treatment of choice if the entire lesion can be removed, but local recurrence is common. Localized initial regrowth can often be managed with repeated surgical excision. Local irradiation has occasionally been used to treat recurrent, persistent or refractory cases to surgical and medical therapy, recalcitrant and large lesions, young patient or when surgery is not feasible.¹¹ Finally, the pharmacotherapy of Kimura's Disease has mainly involved the use of systemic and intralesional corticosteroids for an adequate period have been shown to reduce the size of the lesion but the lesion tends to recur when the drugs are discontinued.^{11,12} Cyclosporine, oral pentoxifylline, all trans retinoic acid with prednisolone, imatinib, leflunomide, azathioprine and vincristine have been tried in the management for Kimura's Disease with variable responses.¹³

The choice of treatment modalities should be individualized. Recurrence is common with all the modalities of treatment.

Our patient had excision of postauricular mass but after 6 months, recurrence of mass happened. Then the patient was treated with 1mg/kg/day of oral prednisolone for 6 weeks which allowed gradual regression of mass. The dose is then tapered over next 4 weeks.

Conclusion

Kimura's Disease may be suspected when eosinophilia or high IgE level are seen in a patient of slowly progressive head and neck swelling or lymphadenopathy.

If not properly diagnosed, the neck mass or postauricular mass may be initially mistaken for a malignancy leading to unnecessary and potentially invasive investigation. However due to a well-obtained clinical history and histological awareness a proper diagnosis has been established in our case.

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