Kimura’s Disease Presenting as Bilateral Postauricular Swelling - A Case Report

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Abstract: Background: Kimura’s disease is a slowly progressive inflammatory disease of unknown aetiology. Kimm and Szeto reported the first case in 1937 in China and termed it as “eosinophilic hyperplastic lymphogranuloma”. The condition is characterized by a triad of painless cervical lymphadenopathy or painless subcutaneous masses in head and neck region, tissue & blood eosinophilia, and markedly elevated serum immunoglobulin E levels. Cases: Middle aged man, presented with bilateral postauricular swelling, with no significant physical findings. Investigations revealed eosinophilia and raised IgE levels in both the cases. USG- Bilateral multiple cervical lymphadenopathy. Microscopy: It was reported as lymphoid hyperplasia on FNAC. The histopathology confirmed the condition to be Kimura’s disease. Conclusion: Although rare the Kimura’s disease makes the differential diagnosis of head and neck masses. This article highlights that: the clinicians should be aware of this rather rare and impressive clinical entity and may spare the patient from unnecessary invasive diagnostic procedures.

Keywords: Eosinophilia, Immunoglobulin, Kimura’s disease, Lymphadenopathy

1. Introduction

Kimura’s disease is a slowly progressive inflammatory disease of unknown aetiology [1]. Kimm and Szeto reported the first case in 1937 in China and termed it as “eosinophilic hyperplastic lymphogranuloma” [2]. It is named after Kimura et al. who referred to it as unusual granulation combined with hyperplastic changes in lymphoid tissue [3]. The condition is characterized by a triad of painless cervical lymphadenopathy or painless subcutaneous masses in head and neck region, tissue & blood eosinophilia, and markedly elevated serum immunoglobulin E levels.

Kimura’s disease is more common in young Asian males of Chinese and Japanese origin [4]. However, in Indian literature there is only limited information available about this condition. Here we report two cases of this rare and impressive entity presenting with cervical lymphadenopathy, which were clinically suspected to be lymphoma or tuberculosis. This report thus highlights the importance of knowing this clinical entity.

2. Case Report

Case one, is a 53 years male, who presented to the ENT outpatient department with complaints of swelling in bilateral postauricular region since four months. On local examination, there was a 4x3 cm swelling on right postauricular region and a 2x2 cm swelling in the left postauricular region. Both the swelling were firm, non mobile and non tender. The overlying skin was normal and there was no increase in local temperature. Rest of the general physical and systemic examination was unremarkable.

The complete blood count and peripheral smear showed eosinophilia with eosinophil count of 22%. The ultrasonography of the site showed multiple lymphadenopathy. His serum IgE levels were raised with a value of 935 IU/ml. All the other investigations showed normal results. The patient underwent fine needle aspiration from the larger right sided postauricular swelling. It was reported as lymphoid hyperplasia.

An excisional biopsy from the right sided postauricular swelling was performed and sent for histopathological examination. The histopathology confirmed the condition to be Kimura’s disease.

Case two is, a 40 years male with bilateral postauricular swelling since two months with right side more than left. Both the swelling were 4x3 cm and 3x2 cm respectively, firm, non tender with normal overlying skin and temperature. His general physical and systemic examination was normal. His local site ultrasonography showed multiple lymphadenopathy.

His complete blood count and peripheral blood showed 26% eosinophilia. Serum IgE levels were raised to 7055 IU/ml. Cytology from right postauricular swelling was suggestive of reactive lymphadenopathy. Histopathological examination showed the features of Kimura’s disease.

On histopathological examination, both the cases showed lymph node with reactive follicles with presence of dense eosinophilic infiltration in interfollicular area and some of the germinal centres along with formation of eosinophilic micro abscesses at places (Figure-1& 2). Endothelial proliferation with sclerosis of vessel walls and focal vascularisation of germinal centres were also seen.

Histopathological Examination; Photo micrographs.
Kimura’s disease was first described by Kimm and Szeto in 1937 in China and termed it as “eosinophilic hyperplastic lymphogranuloma” [2]. It is named after Kimura et al. who described it in 1948. It is a benign slowly progressive condition involving subcutaneous issues, major salivary gland, lymph nodes mainly in head and neck area. However, other sites like oral cavity, groin, trunk and limbs may also be involved. It is endemic in China and Japan in young males. It usually presents as a non-tender subcutaneous swelling in head and neck region, mainly in preauricular and submandibular area, associated with lymphadenopathy, marked peripheral eosinophilia and an elevated IgE level [5]. It may be mistaken for a malignant disorder.

It is a chronic inflammatory condition with uncertain etiology. Some proposed etiologies include neoplasia, parasitic infection, atopic reaction or immune response to various pathogens [5]. However, no causative agent has been isolated from these lesions [6]. No malignant changes have been reported [7] FNAC is usually inconclusive and diagnosis can be established by histopathological examination.

The histopathological features of Kimura’s disease include follicular hyperplasia with reactive germinal centres, with IgE deposition and eosinophilic infiltration of germinal centres and interfollicular zones. Lysis of follicles with microabscess formation may also be seen [8]. Some degree of vascular proliferation of germinal centers, increased postcapillary venules in the paracortex, and sclerosis are also observed. Immunoperoxidase studies show IgE reticular network in germinal centers [9]. This was not done in our cases due to lack of facility.

Kimura’s disease has got conditions like eosinophilic granuloma, Mikulicz’s disease, Acute lymphocytic leukemia, Hodgkin’s disease, Angioimmunoblastic lymphadenopathy and Angiolympoid hyperplasia with eosinophilia in the differential diagnosis [10]. Most of these can be differentiated on clinical and histological basis. Among these, Angiolympoid hyperplasia with eosinophilia (ALHE) was a histological challenge earlier and was thought to be a different variation of same disease. Now it is widely accepted that these two are separate disease entities [11]. ALHE presents as multiple or solitary subcutaneous nodules in head and neck region but affects middle aged Caucasian females as compared to Kimura’s disease which is seen in young Asian males [12]. On histopathology, unlike Kimura’s disease, ALHE shows aggregates and lobules of vascular endothelium lined by plump endothelial cells. Other features like regional lymphadenopathy, peripheral blood eosinophilia and elevated IgE levels are uncommon in ALHE.

The management of Kimura’s disease include observation, medical therapies, surgical excision and radiotherapy. Overall, it has got a slow clinical course with good prognosis, but rarely there may be local recurrence. Some of the cases may show renal complications manifesting as nephrotic syndrome [13].

4. Conclusion

Although rare the Kimura’s disease makes the differential diagnosis of head and neck masses. It can be diagnosed easily by its clinical, specific histopathological features, associated peripheral eosinophilia and elevated IgE levels. This article highlights that; the clinicians should be aware of this rather rare and impressive clinical entity and may spare the patient from unnecessary invasive diagnostic procedures.

References


