Kimura's Disease As Diagnostic Dilemma

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ABSTRACT

Kimura's disease is a chronic inflammatory disease of unknown aetiology. The classical triad includes painless subcutaneous mass in head and neck, eosinophilia and elevated serum immunoglobulin levels. Kimura's disease is rare, besides can produce a diagnostic confusion as seen in our case deviating from its triad of presentation described in literature. A 14 year male child came with painless swelling over right post auricular region since 6 months. He had surgical excision for similar swelling at same site one year back. Complete surgical excision was done and sent for histopathology, which was reported, Kimura's disease. After 3 years of follow up there was no recurrence. Our case showed Kimura's disease can present with normal eosinophil count and serum immunoglobulin levels and histopathology alone is confirmatory. We also suggest surgery is the treatment of choice for Kimura's disease either in primary or recurrence.

Keywords: Kimura's, Lymphadenopathy, Painless swelling

INTRODUCTION

Kimura's disease or eosinophilic hyperplastic lymphogranuloma is a chronic inflammatory disease of unknown aetiology[1]. This benign condition is characterized by a triad of painless subcutaneous masses commonly in head and neck, eosinophilia and markedly elevated serum Immunoglobulin levels[2,3]. The main objective is to report a rare case like Kimura's disease producing a diagnostic confusion by deviating from its classical triad of presentation as described in literature. Our case of young male presented as recurrent painless subcutaneous mass in right post auricular region without any eosinophilia and raised serum immunoglobulin levels which are part of classical triad of Kimura's disease. We managed this case with complete surgical excision despite its recurrence following previous surgery. The patient was followed for 3 years without any sequale of disease suggesting a complete surgical excision may be curative despite its recurrence.

CASE REPORT

A 14 year old male presented to out patient department with painless, soft tissue swelling over the right post auricular region extending up to angle of mandible since 6 months which was gradually increased in size(fig 1). History suggested, he had similar swelling at same site one year back which was surgically removed. On examination the swelling was 10 x 7 cm in size, non tender, globular in shape with well defined margins and firm in consistency.

Ultrosonography showed a well defined hypoechoic collection of size 8 x 10 mm in right post auricular region with surrounding inflammation with multiple enlarged lymphnodes in bilateral post auricular, parotid and level 1 regions. Fine needle aspiration cytology given a differential diagnosis of Kimura's disease or eosinophilic granuloma. Complete blood picture was with in normal limits with no peripheral eosinophilia. Serum immunoglobulin levels are also normal.

For definitive diagnosis, complete surgical excision of the lesion was done via a right post auricular skin incision and dissecting subcutaneously over the swelling all around. The swelling is excised completely and sent for histopathology which was reported as Kimura's disease(fig.2). The patient was followed every 6 months for 3 years without recurrence.

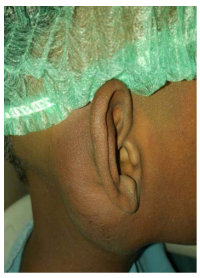

Fig.1: Swelling at presentation

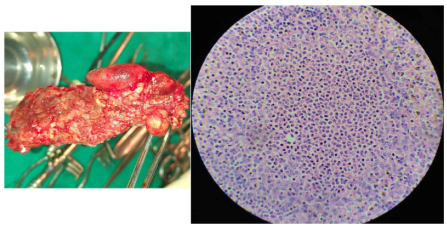

Fig.2: Gross and microscopic pictures of kimura's

DISCUSSION

Kimura's disease is a chronic disease typically manifested by painless subcutaneous nodules commonly in head and neck. It was first described in China by Kim and Szeto

and later Kimura and colleagues in 1948 in Japan made a more detailed description[1].

Exact prevalence of Kimura's is not known. Most cases of this are reported in East and South East Asia with very few cases in Europe and United states. A male predominance is seen with a male : female ratio of 3.5 – 9 : 1[2]. Most cases are reported in 2nd to 3 rd decade of life.

The cause and pathogenesis of Kimura's is unclear, although allergic responses, trauma and autoimmune reponse have been implicated as trigger factors. It has been speculated that a viral or parasite trigger may alter T cell immunoregulation or induce an IgE mediated type 1 hypersentivity resulting in the release of eosinophiliotrophic cytokines. Immunohisto chemical studies performed on skin, lymphnodes and peripheral blood have shown marked proliferation of HLA-DR CD4 cells, which inturn may precipitate the high serum IgG and marked eosinophilia[3].

The lesions are usually in deep subcutaneous tissues, more often associated with regional lymphadenopathy and salivary gland involvement[4,5]. Other sites of involvement include oral cavity, axilla, limbs, groin and trunk6. Renal involvement, usually extra membranous glomerulonephritis is found in 60 % patient and protenuria in 12 to 16 % of cases is not seen in our case[7,8].

The differential diagnosis for Kimura's includes tuberculosis, nodal metastasis, lymphoma, eosinophilic granuloma, granulomatous diseases, salivary gland pathologies, lymphadenitis, drug reaction and angiolymphoid hyperplasia with eosinophilia(ALHE).

The most controversial of these is ALHE as at time ALHE and Kimura's have been considered to be the same disease. Recent literature however describes these as two distinct entities[9,10,11]. As per literature laboratory investigations typically reveal eosinophilic and elevated serum immunoglobulin levels. The degree of eosinophilia may correlate with the size of lesions and recurrences may be more likely in patient with eosinophil count more than 50% and serum immunoglobulin more than 10,000 mIU/ml[12].

However in our case the laboratory results showed there is no increase in eosinophil count or elevated serum immunoglobulin levels producing a diagnostic dilemma.

Histopathology alone is confirmatory and so needs a surgical excision of lesion. Histologically Kimura's disease presents as preserved lymph node architecture with reactive and prominent germinal centers. Dense eosinophilic infiltration of the interfollicular zones, lysis of the follicles, and occasionally microabscesses are seen. Granuloma formations contain infiltration of eosinophils, lymphocytes, plasma cells, and histiocytes. Tissue fibrosis, sclerosis, and vascular proliferation are also present.Vessels remain thin-walled with cubical endothelial cells present. Rarely the features include progressive destruction of germinal centers, presence of polykaryocytes (which are not pathognomic for that disease). Immunofluorescence tests show germinal centers containing heavy IgE deposits and variable amounts of IgG, IgM,and fibrinogen[13].

Systemic steroid, radiotherapy and surgical modalities have been tried with variable success. Recently, Messina-Doucet et. al have proposed a treatment protocol for this disease. They suggested that surgical excision is the treatment of choice followed by regular urinary protein estimation to rule out any renal involvement. Initial local recurrence can be managed by surgery alone but if recurrences are frequent or renal involvement appears systemic steroid should be started[14].

Similarly in our case, there was a localised recurrence which was treated by surgical excision. So we made a complete surgical excision and after 3 years of follow up there was no recurrence suggesting a complete surgical excision can be curative. So we suggest surgery as first choice for primary or recurrence.

CONCLUSION

Kimura's disease despite a rare disorder should be considered as differential diagnosis when presenting as subcutaneous mass associated with multiple lymphadenopathy. Although in our case, eosinophilia and elevated serum immunoglobulin may be absent, should not form grounds of exclusion as they form classical triad of Kimura's disease.

Our case had showed histopatholgy alone can differentiate from other similar lesions especially ALHE which also can present with normal eosinophil count and serum immunoglobulin levels and based on laboratory investigations should not exclude the Kimura's disease. We also recommend surgery is the treatment of choice for Kimura's disease either in primary or recurrence.

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