

A Rare Case of Kimura's Disease of the Eyebrow Region

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Abstract: Kimura's disease is a benign inflammatory disease generally involving the head and neck region. It is characterized by lymphoid follicular hyperplasia with prominent germinal centers and eosinophilic infiltrates in the inter follicular region and proliferation of post capillary venules. Peripheral blood eosinophilia and elevated serum Ig E are seen in most of the cases. A 21 year male patient came with a subcutaneous swelling beneath the left eyebrow. Differential count showed neutrophils 69%, lymphocytes 15%, eosinophils 10% and monocytes 6%. ESR was 14 mm/hour. The serum IgE levels were 492.5 IU/ml. Histo pathological examination revealed hyperplastic lymphoid follicles of varying sizes with reactive germinal centers and well formed mantle zones. We report a rare case of Kimura presenting in the subcutaneous region of the eyebrow.

Keywords: Kimura's disease, Lymphoid follicular hyperplasia, Eosinophilia, Serum Ig E, Head and neck

INTRODUCTION

Kimura's disease is a benign, slow growing inflammatory condition generally presenting as single or multiple masses involving the head and neck region. The condition is common in Asians as compared to non-Asians. The exact etiology and pathogenesis is unknown. The probable suspected etiologic factors include viral or parasitic stimulus inducing an allergic or autoimmune response [1]. The disease was first described by Kim and Szeto in 1937, but it was kimura et al who coined the term in 1948 [2]. It has been reported sporadically in other areas and ethnic groups. The disease predominantly involves the head and neck region and is frequently associated with regional lymphadenopathy. Salivary gland involvement may or may not occur. Other sites of involvement are axilla, groin, popliteal fossa, inguinal region, scalp, extremities and trunk [3, 4] Rare sites include kidneys, orbit, ears, spermatic cord and median nerve. Though the disease commonly involves the young and middle aged men, orbital involvement is generally seen in older age groups. Presence of peripheral blood eosinophilia and elevated serum Ig E levels substantiate the diagnosis [1].

CASE REPORT

A 21 year male patient was referred to Sarojini Devi Eye Hospital in November 2008. He presented with a subcutaneous swelling beneath the left eyebrow.

He complained of gradual increase in the size since last 6 months. There were no other complaints like itching, redness etc. Ocular examination, general examination revealed no other abnormality. Laboratory investigations included peripheral blood examination which revealed hemoglobin of 13 grams %, Total WBC of 9600 cells/cubic mm. Differential count showed neutrophils 69%, lymphocytes 15%, eosinophils 10% and monocytes 6%. ESR was 14 mm/hour. The serum IgE levels were 492.5 IU/ml. (Normal for our lab is upto 378 IU/ml). All the biochemical and radiological parameters were within normal limits. Liver profile was within normal limits. Renal parameters were normal. CT scan of the orbit revealed a mildly hyperdense soft tissue mass lesion in the subcutaneous plane without orbital involvement.

A biopsy was performed. Histopathological examination revealed hyperplastic lymphoid follicles of varying sizes with reactive germinal centers and well formed mantle zones (Fig. 1A, 1B). Prominent vascular proliferation was seen. Inter follicular areas showed dense eosinophilic infiltrates. Based on the histological findings, Serum IgE levels and peripheral blood eosinophilia (Fig. 2) we reported the condition as Kimura's disease. As the lesion was not well circumscribed a near total excision of the mass was done.

The patient came to the Ophthalmology Department again in August 2010 with recurrence of the swelling in the same region. Peripheral blood examination showed elevated leucocyte count. Eosinophil count was 12% and serum Ig E level were 812 IU / ml. Re excision was done. Again complete resection was not possible due to the poorly circumscribed nature of the mass. Patient again came to the hospital in March 2013 with recurrent swelling in the same region. Excision was done and intra orbital steroids were injected.

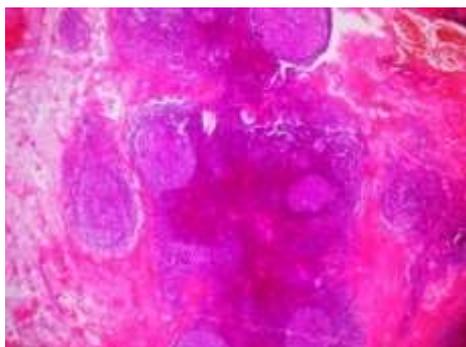


Fig. 1 A: Shows reactive lymphoid follicles with prominent germinal centers and adjacent areas of fibrosis



Fig. 1B: Shows reactive lymphoid follicles with prominent germinal centers and adjacent areas of fibrosis

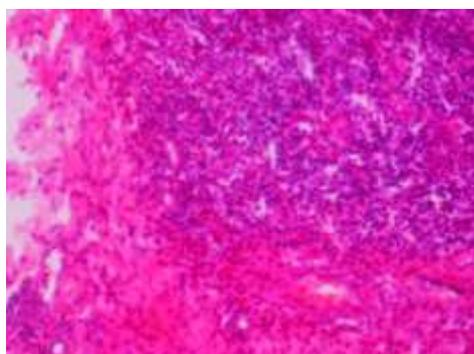


Fig. 2: Higher magnification of a follicle showing capillary proliferation and dense eosinophilic infiltrate along with lymphocytes, plasma cells with surrounding fibrosis

DISCUSSION

Kimura's disease is an uncommon chronic inflammatory disorder of unknown etiology presenting typically as subcutaneous nodular masses especially involving the head and neck region.

Our patient is a 21 year old male. In our patient only a solitary swelling beneath the left eyebrow was noted, no other swellings or lymphnode enlargement was seen. Males are more commonly affected than females, the male to female ratio according to various studies ranged from 7:1 to 3.5:1 [2]. Solitary or multiple lesions can occur. Unilateral or bilateral involvement can occur [5]. The typical presentation is a painless subcutaneous swelling in the head and neck region [6]. Orbital and periorbital involvement is rare but a few cases have been reported in literature [7]. In this case all the biochemical and radiological parameters were within normal limits. Liver profile and renal parameters were normal. Renal involvement is frequently seen in patients of Kimura's disease. It may present as nephrotic syndrome or in lesser than 1% of patients as proteinuria [1, 3]. Other renal manifestations include Minimal Change Disease, Membranous Nephropathy and Mesangio proliferative Nephropathy. Many allergic conditions may also occur in association with Kimura's disease like asthma, urticaria, eczema, pruritis, rhinitis etc. [4, 5].

The activated CD 4 cells of the Th2 phenotype release cytokines IL-2, IL-5 and IL-13 which result in high serum IgE and prominent eosinophilia [8]. According to the other studies it was proposed that the disease is purely of an atopic nature because tests for all the probable parasites were negative and no evidence of insect bite was identified.

But the fact that the disease is more prevalent in Asian population and very rare in the west suggests that racial and/or cultural factors also play a significant role in the etiology of Kimura disease [2].

Diagnosis of Kimura disease is made by excisional biopsy with the typical cellular, vascular and fibrous components. Histopathologically the disease is characterized by reactive lymphoid follicles with prominent germinal centres, inflammatory cell infiltration composed of dense eosinophilic infiltrates in a background of abundant lymphocytes and plasma cells, eosinophilic microabscesses with central necrosis and mast cells [5, 8]. Capillary proliferation and fibrosis are prominent features. The capillaries are thin walled high endothelial venules.

Laboratory tests often reveal peripheral blood eosinophilia and elevated serum IgE levels. CT and MRI scan help to delineate the extent of the disease. In this case CT scan of the orbit revealed a mildly

hyperdense soft tissue mass lesion in the subcutaneous plane without orbital involvement. On CT scan with contrast enhancement the lesion appears as homogenous slightly hyper attenuated mass. MRI will differentiate its precise nature from other soft tissue tumors. This lesion appears as heterogenous-hyperintense sometimes slightly hypointense on T1 and T2 weighted images. According to one study IHC of the lesion demonstrated prominent perithelial cells by alpha smooth muscle actin antibody in the majority of blood vessels [4, 6, 8].

Kimura is commonly compared with ALHE (angiolymploid hyperplasia with peripheral eosinophilia). Vascular hyperplasia is more prominent in ALHE and the endothelial cells are epithelioid and/or vacuolated [2, 8].

The treatment options include surgical excision, systemic and intralesional steroids, radiotherapy and cytotoxic agents. Surgery as the only treatment yielded limited success with multiple recurrences.

CONCLUSION

Kimura's disease though uncommon should be considered in Oriental population presenting as superficial masses in head and neck region. Though the disease is harmless and has no potential for malignant transformation, early diagnosis helps to avoid possible loss of vision and spares the patient from unnecessary investigations.

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