Kimuras Disease a case report

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Abstract:
Kimuras disease is a rare chronic inflammatory disease that presents as a subcutaneous mass in the head and neck region often associated with regional cervical lymphadenopathy. We report a case of Kimuras disease in a 14yr male who presented with bilateral cervical lymphadenopathy. The diagnosis was based on characteristic histopathological findings in correlation with peripheral eosinophilia and elevated serum Immunoglobulin E levels. The histopathological differential diagnoses have also been discussed.

Keyword: Eosinophilia, Immunoglobulin E, Kimura, Lymphadenopathy

Introduction:
Kimura’s disease (KD) was initially described in 1937, in the Chinese literature, by Kimm and Szeto as an “eosinophilic hyperplastic lymphogranuloma” and it came to be known as KD after its publication by Kimura et al of similar cases in Japan under the title of “
Atypical granulation associated with hyperplastic abnormalities in the lymphoid tissue. The diagnosis of this entity has been a challenge, considerable confusion existing between this entity and Angiolymphoid hyperplasia with eosinophilia (ALHE). We report a case of Kimura’s disease in a 14yr male discussing the histopathological differential diagnosis.

**Case report:**
A 14yr boy presented to the pediatric surgical outpatient with complaints of swelling on both sides of the neck for the past 9 months. There was no history of fever or any drug intake (anticonvulsants, sufa drugs, aspirin or erythromycin). On examination, two nodules were seen, one in the right and other in the left cervical region measuring 2x1cm and 1.6x1cm respectively with well defined margins and were firm in consistency. As fine needle aspiration cytology did not yield enough material, an excision biopsy of the right cervical nodule was done and the tissue was sent for histopathological examination. We received a brown nodular piece of tissue measuring 1.5x 0.5 cm. The cut surface appeared grey white. No foci of calcification or necrosis were identified. Microscopic examination revealed a lymph node with focal sclerotic obliteration of subcapsular sinuses and follicles with prominent germinal centres (Fig.1A). There was folliculolysis, diffuse infiltration by eosinophils forming abscesses (Fig.1B). Paracortical fibrosis and prominent blood vessels in the germinal centre were present (fig.2A). An occasional Warthin Finkeldey giant cell was noted (fig.2B). No parasites or granulomas were identified in the multiple level sections studied. Follicles with CD20+ germinal centres and bcl-2 + mantle zones were demonstrated by immunohistochemistry. Thus the morphological features supported by immunohistochemistry favoured the diagnosis of Kimura’s disease.

**Discussion:**
Kimura’s disease is a rare chronic inflammatory disorder of unknown etiology. It presents as a deep, subcutaneous mass in the head and neck region and is frequently associated with regional lymphadenopathy or salivary gland involvement. The majority of patients with Kimura’s disease have been reported in Asian population in China, Taiwan, and Japan. Sporadic cases of Kimura’s disease have been described in other ethnic groups including Caucasians, Blacks, Hispanics and Arabs. Most of the patients are young males in the second and third decades of life, although this entity has been described over a wide age group (8.5yrs-64yrs) and the common sites include head and neck (76%), particularly infraauricular, retroauricular, orbit, eyelid, palate, pharynx. Our patient was a 14 yr boy who presented with right and left cervical lymphadenopathy.

The clinical course of Kimura’s disease is generally benign and self limited. Kimura’s disease may be complicated by renal involvement. Proteinuria may occur in 12 % to 16 % of cases. Nephrotic syndrome is the most common presentation, a wide spectrum of histologic lesions such as minimal change disease, mesangioproliferative glomerulonephritis, focal segmental glomerulosclerosis, membranous nephropathy, IgM nephropathy and IgA nephropathy have been described. Urine analysis and biochemical investigations ruled out any renal involvement in the present case.
Kimura’s disease is usually accompanied by peripheral blood eosinophilia which may reach 10% to 70% and increased serum IgE concentrations (800 to 35,000 IU/ml). Isolated cases with normal peripheral blood eosinophils and serum IgE levels have been reported. Raised eosinophils of 33% and serum IgE levels of 25,000 IU/ml were seen in the present case also.

While manifesting as enlarged lymphnodes, the constant histopathological features include preserved nodal architecture, florid germinal center hyperplasia, eosinophilic infiltration and postcapillary venule proliferation. Other features include capsular fibrosis, subcapsular sinus obliteration, polykaryocytes, vascularization of the germinal center and eosinophilic abscess. All these features were appreciated in our case. Over the years, there has been considerable confusion between Kimura’s disease and ALHE with early reports using both terms synonymously. Despite a few common histopathological features, ALHE and Kimura’s disease can be differentiated from each other. Large irregular blood vessels lined by endothelial cells with cytoplasmic vacuoles are characteristic of ALHE. Small blood vessels lined by non-protruberant endothelial cells devoid of cytoplasmic vacuoles and relative abundance of eosinophils, lymphoid follicles are diagnostic of Kimura’s disease. Eosinophils are small in number or practically absent in ALHE but they are always abundant in KD. When manifesting as a subcutaneous nodule, KD is deeply located extending into skeletal muscle. Extension into muscle is not seen in ALHE. The other entities to be considered in the differential diagnosis are parasitic lymphadenitis (filarial), langerhans cell histiocytosis, drug reactions and Hodgkins lymphoma in view of the presence of abundant eosinophils. Absence of granulomatous reaction and a parasite, absence of langerhans histiocytes, drug history (anticonvulsants, sufa drugs, aspirin or erythromycin) and Reed- Sternberg cells respectively ruled out these possibilities.

The etiopathogenesis of KD is unclear, although it might be a self limited or autoimmune response triggered by an unknown stimulus. It has been speculated that a viral or parasitic trigger may alter CD4 cell immunoregulation or induce an IgE mediated type-I hypersensitivity resulting in the release of eosinophilotrophic cytokines causing eosinophilia and elevated serum IgE levels.

The optimal treatment for KD is not well established. The range of treatment options include conservative treatment, steroid therapy, radiotherapy, cryotherapy, laser fulguration and surgical excision. Nevertheless, the treatment outcome after excision is variable and recurrence is common. In cases treated with surgical excision alone, the recurrence can be as high as 25%.

References:


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