Kimura's disease-A rare entity.

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Abstract:

Kimura disease is a rare chronic inflammatory disorder of unknown etiology primarily seen in young Asian males. It is a benign disease with an indolent course, gradually increasing in size over months or years. It usually presents as a mass lesion in the subcutaneous tissue of the head and neck region or major salivary glands area often associated with regional lymphadenopathy. In some cases lymph node enlargement is the only manifestation of the disease. It has no potential for malignant transformation. Spontaneous involution is rare. The main concern in this disease is its capacity to grow and cause disfigurement. We reported a case of a 16year old male who presented with bilateral cervical swelling and unilateral submandibular swelling. Specimen of one of the cervical swellings was received in histopathology department. Microscopy revealed there was reactive follicular hyperplasia with infiltration of giant cells of warthin-finkeldey type, eosinophils and vascular proliferation. Diagnosis of Kimura's Disease was given on histopathology.

Keywords: - Lymphadenopathy, Subcutaneous tissue, Warthin-Finkeldey type of Giant cells.

Introduction:

Kimura's Disease or Eosinophilic hyperplastic lymphogranuloma is a chronic inflammatory disorder of unknown etiology mostly seen in young Asian males.[1]It takes the forms of large subcutaneous masses that are often concentrated around ears or located adjacent to major salivary glands along with frequent lymph node involvement and peripheral eosinophilia. The disease was thought to be identical to Epithelioid hemangioma or Angiolymphoid hyperplasia with Eosinophilia (ALHE) but recent literature indicates that they are both completely different conditions with only few superficial similarities.

This case is reported for its rare possible occurrence.

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Case Report:

A 16 year old male patient presented in ENT department of our hospital with painless left sided neck swelling since 1 year which was gradually increasing in size. Then patient developed bilateral multiple swelling in cervical region with cervical pain. On clinical examination, swellings were mobile, nontender with well defined borders. Complete Blood Count of patient revealed eosinophilia. Excisional biopsy from left sided posterior cervical swelling was taken and was

sent for histopathological examination.

Gross Examination

Grossly 1.5x1x0.5cm sized greyish white; soft to firm tissue mass was received.

Microscopic Examination

Histopathological examination revealed reactive follicular hyperplasia showing germinal centers in follicles. [Image 1&2]. In between, there were infiltrations of multinucleated giant cells (warthin-finkeldey type) [Image 3]. There was presence of vascular proliferation and many eosinophils in parafollicular areas. [Image 1&2]

Image-1 Lymphocytic Infiltrate, Sinus Histiocytes, Eosinophils and Reactive Follicle, Vascular proliferation [H&E 10X]

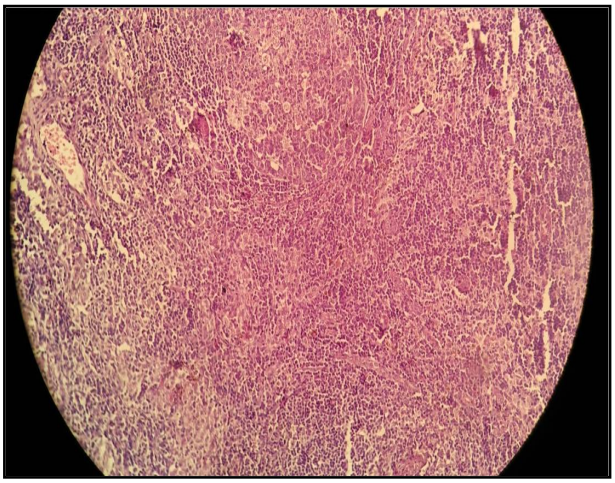


Image 2 Vascular proliferation, Reactive follicle [H&E 10X]

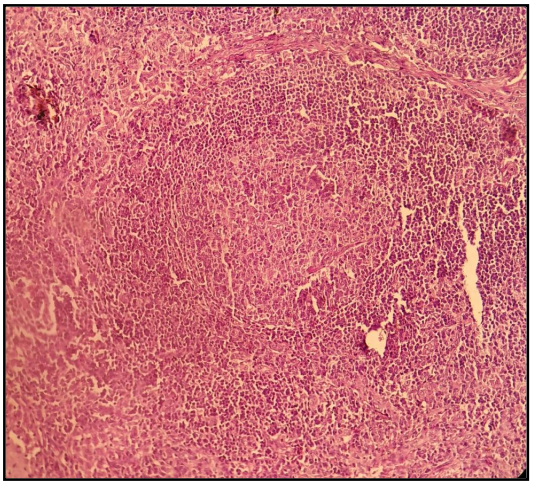
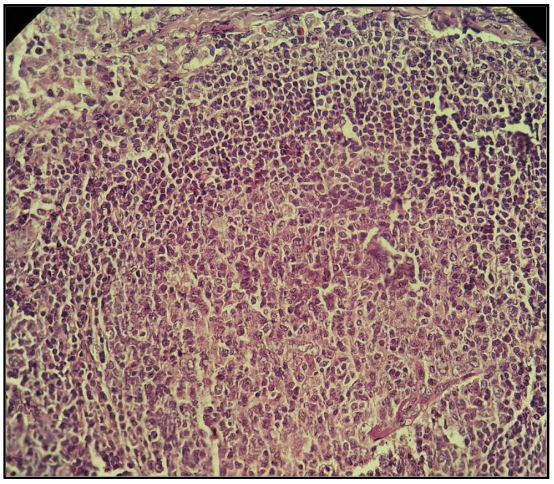


Image 3 Giant cells of Warthin-Finkeldey type [H&E 40X]



Fine Needle Aspiration Cytology (FNAC) of the one of the swellings of the patient done before showed findings of pleomorphic adenoma of salivary gland. Advice for Serum IgE levels was given for correlation and that came out higher. (657 IU/ml)

Correlating all of these clinicopathological findings, diagnosis of Kimura's Disease

was made. On follow up of the patient as patient was prescribed steroids and anti inflammatory drugs, he responded well with decrease in size of all of the swellings progressively.

Discussion:

Kimura disease was first reported by Chinese authors Kimm & Szeto in 1937 with the term eosinophilic hyperplastic lymphogranuloma.[2] The disorder received its current name in 1948 when Kimura et al noted the vascular component & referred to it as an unusual granulation tissue combined with hyperplastic changes in lymphoid tissue.[1] Most cases of Kimura disease are found in Asians & it is endemic in the far East.[3] Young males are commonly affected, the median age being 28-32 years. It usually presents as a mass lesion in the subcutaneous tissue of head & neck region or the major salivary glands, often associated with regional lymphadenopathy. Sometimes lymph node enlargement is the only manifestation of the disease. The patient may present with painless slowly enlarging soft tissue mass, blood & tissue eosinophilia and markedly increased serum Immunoglobulin E. Periauricular, parotid & submandibular regions are commonly affected. Renal involvement can cause nephrotic syndrome. Pathophysiology of Kimura disease is unknown, although an allergic reaction, trauma & an autoimmune process have all been implicated as the possible causes. Histopathological study is recommended to confirm the diagnosis. Lymphoid follicles with prominent germinal centres characterise Kimura disease. Marked eosinophilic infiltration sometimes forming eosinophilic microabscesses along with capillary proliferation forming canalized vessels with flat endothelium.[3,5] Differentiating Kimura disease from ALHE requires a strict analysis of clinical & histological features because the diseases are similar & were once thought to be same disorder. Both the diseases usually present with soft tissue masses in head & neck region, but in ALHE the lesions are mostly dermal or subcutaneous & not found in lymph nodes, which is a common location for Kimura disease. Clinically Kimura disease is believed to be a disease of the Asians & ALHE one of the western world. ALHE occurs in older, predominantly female population. Kimura disease is primarily a disease of young males. Regional lymphadenopathy, peripheral blood eosinophilia & raised serum IgE levels which are characteristic of Kimura disease are rarely seen in ALHE. The histology of ALHE is typified by an exuberant proliferation of capillary vessels some of which may not be canalized. These are lined by epithelioid or histiocytoid endothelial cells which are not seen in Kimura disease.[5]Tendency towards renal involvement & nephrotic syndrome is restricted to Kimura disease & not observed in ALHE.[4]Because of presence of eosinophils along with polymorphous population of reactive Lymph Node, Hodgkin Lymphoma should always be kept in differential diagnosis of this disease. But there were no atypical cells and Reed Sternberg cells, so Hodgkin Lymphoma can be excluded easily. Spontaneous resolution, though rare, has been reported .Most patients have a prolonged course with slow enlargement of the mass. Surgical excision is the treatment of choice to preserve cosmesis & function. However recurrence rate can be as high as 25%.Other treatment modalities include conservative treatment with the aim of spontaneous involution, intralesional or oral steroids, radiotherapy, cryotherapy, laser & cytotoxic agents. However the overall prognosis of Kimura disease is reported to be good.

Conclusion:

Kimura disease is a locally disfiguring disease with indolent course, whose true importance lies in its gradually increasing in size over months or years. A high index of suspicion is needed to diagnose this condition. This case report draws attention on such a rare chronic inflammatory disease which mimics neoplastic conditions, ALHE and Hodgkin Lymphoma. This disease should be considered in differential diagnosis of patients presented with head & neck mass and lymphadenopathy and investigated accordingly as this disease has good prognosis. Knowledge of signature features of Kimura's disease put the physicians in a better position to evaluate its clinical outcome and better treatment.

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