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Case report

KIMURA'S DISEASE: A RARE CASE REPORT

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ABSTRACT

Introduction: Kimura's disease is of unknown etiology. **Case report**: This report describes an interesting case of Kimura's disease in an 18 years old male manifesting as a unilateral left sided parotid swelling with multiple cervical lymphadenopathy. There was eosinophilia and increased levels of circulating IgE. Patient was treated with surgical excision and corticotherapy, cetrizine was prescribed. Histologically there were numerous follicles with prominent germinal centres. Interfollicular area was infiltrated with eosinophils and plasma cells. **Conclusion:** Any swelling in the head and neck region associated with eosinophilia should make one to suspect the Kimura's disease.

Keywords: eosinophilia, lymphadenopathy, angiolymphoid hyperplasia

INTRODUCTION

The history of Kimura's disease dates back to 1937 when HT Kimm and C Szeto first described about it.[1] Kimura et al in the year 1948 was the pioneer to describe the microscopic features of the disease. Hence, the disease is recognised by his name.[1] This disease is seen in young Asian males and the cause of this rare entity is not known. Only 200 cases are reported since its histological description and sporadic in the rest of the world.[2] Clinically the disease presents as swelling in the head and neck region characterised by eosinophilia in blood and tissue with marked elevated serum immunoglobulin E (IgE) levels.[3] This report describes a rare case of Kimura's disease in a 18 years old male who presented with parotid swelling

CASE REPORT

An 18 years old male presented to the ENT outpatient department with the swelling on the left side of the face below the left ear. It was sudden in onset and progressed gradually to attain the size of 12X10cms. There was no past history of prolonged fever, cough, loss of weight and loss of appetite. He was non-alcoholic and non-smoker. The family history was unremarkable. On palpation the swelling was diffuse, non-tender, firm, multiple over left parotid region The swelling was measuring 12x10cms, mobile, skin over the swelling was pinchable and hyperpigmented. Other groups of the lymph nodes were not enlarged. There was no hepatosplenomegaly. Bilateral ear and facial nerve examination were within normal limits. The vital signs and laboratory investigations were normal except there was eosinophilia and elevated IgE levels as shown in table-1. Neck ultrasonography revealed enlarged parotid gland with altered echotexture, multiple enlarged pre and post auricular lymphnodes. Fine needle aspiration biopsy revealed features of reactive lymphadenitis. After a course of antibiotics the swelling was excised and sent for the histopathological examination. He was treated with

levofloxacin, acclofenac, cetrizine and B- complex tablets.

The specimen was received in the histopathology section. Macroscopic examination revealed a well circumscribed mass measuring 12X8X5cms. External surface was unremarkable and cut section showed grey white homogenous areas as shown in figure - 1. The specimen was processed routinely in the histopathological section and the sections were stained with haematoxylin and eosin stain (H/E). Microscopic examination revealed the architecture of the lymph nodes with numerous follicles showing hyperplastic feature with germinal centers. Many eosinophils and plasma cells were present in the interfollicular area. Also seen are perifollicular fibrosis, prominent blood vessels with endothelial proliferation and mild hyaline change as shown in figure 2. The diagnosis of Kimura's disease was made.

Table 1: Shows vital signs and laboratory findings

Vital Signs: Pulse Rate: 86/minute, Blood Pressure: 130/80 mm of Hg, Respiratory Rate: 28/minute Temperature: 38.5^{o}c
Laboratory Investigations: Hb%: 11.8g/dl (normal:12.5-14.5g/dl) RBC Count: 3.8millions/cmm (normal:4.5-5.5millions/cmm) Haematocrit: 34% (normal: 35% to 45%) WBC Count: 9800 cells/cmm (normal: 4000 to 11,000/cmm) Differential Count: Neutrophils: 65%, Lymphocytes: 23%, Eosinophils: 10%, Monocytes: 02% Basophils: 00% Bleeding Time-3 minutes 30 seconds (Normal:2-6minutes) Clotting Time-4 minutes 30 seconds (Normal: 2-8minutes) Absolute eosinophil count: 600 cells/cmm (Normal:40-440cells/cmm) Serum IgE:4.4mg/dl (Normal: 0.01 – 0.04 mg/dl) Stool tests: negative for parasites Random Blood Sugar: 90mg/dl (normal: 80 to 110mg/dl) Blood Urea: 19mg/dl (normal: 5 to 25mg/dl) Serum Creatinine: 0.8mg/dl (normal: 0.8 to 1.2mg/dl)
Serological Investigations: HIV-1 & 2:Negative , HBsAg : Negative
Routine Urine Examination: Albumin: Nil, Sugar: Nil, Urine micro: 1-2 pus cells/HPF

Table 2: Shows the difference between kimura's disease and angiofollicular hyperplasia with eosinophilia[1]

Kimura's disease	**Angiofollicular hyperplasia with eosinophilia**
Young man (2^{nd}-3^{rd} decade)	Middle aged woman (3^{rd}-5^{th} decade)
Oriental	Westerner
Voluminous subcutaneous mass, adenopathy, salivary gland involvement	Subcutaneous or dermal cervical papules or nodules; overlying skin is erythematous or pigmented; rare adenopathies
Follicular hyperplasia, eosinophil infiltration of interfollicular and perivascular zones with abcess formation and lysis, postcapillary venule proliferation	Same aspect. Histiocytoid vessels with particular endothelial cells (different sized nuclei and protrusion into the vessel lumen)
Elevated total IgE, hypereosinophilia, benign pathology	No increase in IgE, hypereosinophilia. Benign form of a group of vascular proliferation diseases ranging from hemangioendothelioma to epitheloid angiosarcoma

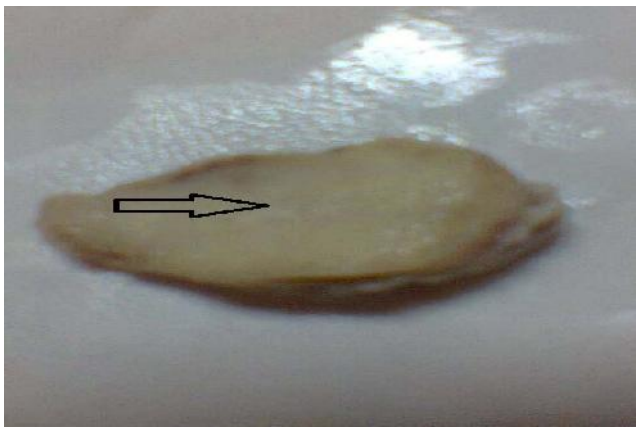

Fig 1: Cut section of the specimen showing homogenous grey white areas.

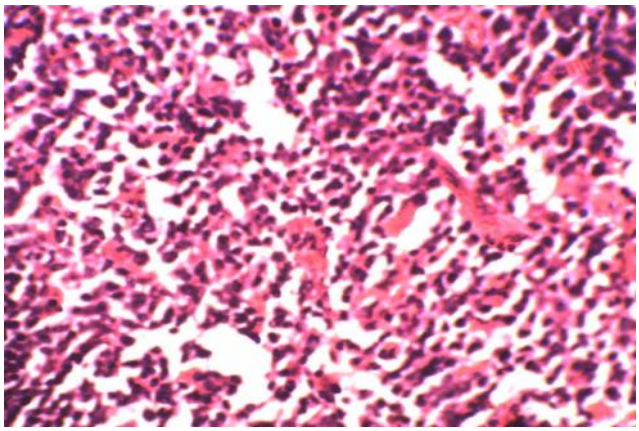

Fig 2: Showing the microscopic picture of Kimura's disease 40X(H/E).

DISCUSSION

Kimura's disease is a chronic inflammatory condition where in the etiology may not be established. Hence, there was a misconception between this disease and angiolymphoid hyperplasia with eosinophilia (ALHE). But later it was proved that they were two separate disorders. Autoimmune/allergic response was seen in Kimura's disease as against benign vascular hyperplasia in ALHE.[4]

There is no age preponderance to this disease. However 2nd and 3rd decade have been reported in many literature, which also collaborates with the present case.[5] Won Jun Choi et al reviewed 54 patients and found that the mean age of the patients was 33.1 years with head and neck region being more commonly involved[5]. Other areas to be involved are kidneys, orbit, ears, spermatic cord and median nerve.[5] Clinical differential diagnosis should include chronic inflammatory disorder of the salivary glands, granulomatous gland disease (cat-scratch disease, sarcoidosis) and autoimmune disease (Sjogren's syndrome), benign and malignant lesions of the gland and cysts should be differentiated. However the following conditions like lymphoma, metastasis, ALHE, Langerhans cell histiocytosis, florid follicular hyperplasia, Castleman's disease, drug reaction, dermatopathic lymphadenopathy, parasitic lymphadenitis and allergic granulomatosis of Chung and Strauss are to be considered in differential diagnosis.[3]

Laboratory analysis showed peripheral blood eosinophilia and increased serum total IgE concentration. There was no proteinuria. Regional lymphadenopathy, peripheral blood eosinophilia and increased IgE levels are commonly seen in Kimura's disease, but not in ALHE. Renal involvement is found in half the patients of Kimura's disease. Yuen et al [6] reported that renal involvement is about 60%. Table-2 reveals the differences and similarities between Kimura's disease and ALHE.[1]

Surgical excision of the lesion is the first line therapy. Corticosteroids therapy is partly unsuccessful. Radiation and cytotoxic therapy has to be considered for the refractory lesions which do not respond to surgery. Cetrizine (histamine [H-1] receptor blockers) induced a complete remission in a corticosteroid dependent patient within 2 months of treatment.

CONCLUSION

This rare case report underlines the importance of taking into account Kimura's disease when there is eosinophilia in the peripheral blood and swelling in the head and neck region.

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