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Medical Sciences **Case Report ……!!!**

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KIMURA'S DISEASE – A CASE REPORT

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ABSTRACT

Kimuras disease is a rare disease with wide variety of local and systemic presentation. The disease is characterized by swelling and lesions in the head and neck region, with involvement of the subcutaneous soft tissue, major salivary glands, and lymph nodes and rarely kidneys. Here we present a case of 32 year old female patient who was diagnosed with kimuras disease at our institution. Post operatively there is no sign of recurrence since last one year.

INTRODUCTION

Kimuras disease is a benign rare chronic inflammatory disorder characterized by subdermal lesions in the head or neck or painless unilateral inflammation of the cervical lymph nodes[1].It is generally limited to skin, lymph nodes,and salivary glands, but patients with Kimura disease and nephrotic syndrome and focal segmental glomerulosclerosis have been reported[7]. Though the disease is benign, its course is chronic and recurrent. Variety of treatment including surgical excision, steroid therapy, immunosuppressants, antihistamines, radiotherapy has been described with variable results.

CASE REPORT

A 32 Year old female presented to Out patient Department of E.N.T, Dr Baba Saheb Ambedkar Hospital, New Delhi with complaints of right nasal dorsum mass since 2 years which was progressive in nature since 2 months, and significant disfigurement of the face. F.N.A.C was done and it revealed florid reactive lymphoid population comprising of small and large lymphocytes with immunoblasts with fare number of eosinophils, there were also present capillary fragments and swollen endothelial cells (Fig.-1) Routine Blood investigations revealed eosinophilia (9%), renal function tests and other routine investigations were within normal limit. Excision biopsy was done. The mass was very vascular and was sent for histopathology examination. Histology of the specimen revealed un-encapsulated lesion with the predominance of reactive lymphoid follicles. There were collections of eosinophils with small abscesses at the periphery of the lymphoid follicles. It also shows small sized blood vessels with swollen endothelial cells at the periphery of the lymphoid follicles (Fig-2). Based on these histo-pathological findings diagnosis of kimuras disease was made. At present patient had six weeks of steroid therapy and she is on regular follow up at monthly interval for one year without any recurrence.

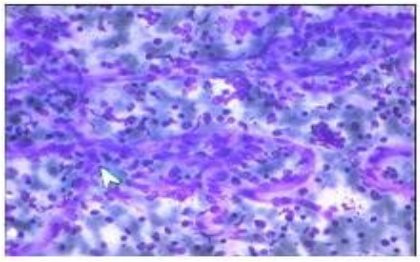

Fig-1 F.N.A.C revealed lymphocytes, eosinophils and capillary fragments

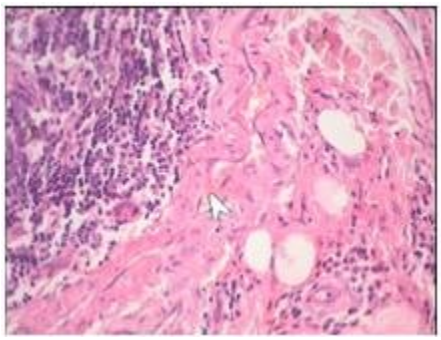

Fig-2 Reactive lymphoid follicles with eosinophilic abscess and swollen endothelial cells

Fig -3 Post op photograph

DISCUSSION

Kim and Szeto in china were the first to describe the disease as an "eosinophilic hyperplastic granuloma". Kimura himself reported on the disease in 1948.[1] This rare disease is endemic in different parts of Asia. It mainly affects young asian men (median age: 26years; male:female ratio>3.1).[1] Although it has been reported in other areas and ethnic groups.[4] It mainly presents as a swelling in head and neck region. It primarily involves the subcutaneous soft tissues of the postauricular region and sometimes the salivary glands and lymph nodes.[1,2] Very rarely it has been reported to involve other sites, such as the oral cavity, conjuctiva, eyelid, tympanic membrane, skeletal muscle, prostate, peripheral nerves, kidneys and epiglottis.[4,5,6] The disease onset is gradual which increases over a period of months to years. Peripheral blood eosinophilia is almost always present. Around 12% of the patients have renal disease characterized by proteinuria.[6,7] Clinically, the differentials include inflammatory and neoplastic lesions, chronic granulomatous diseases, infections (e.g., toxoplasmosis) and lymphomas both hodgkins and non-hodgkins. Radiological investigations in the form of CT and MRI can determine the

extent of the disease as well the lymph node involvement.[8] Fine needle aspiration cytology is helpful only in few cases.[9] The definitive diagnosis can be made by histopathological examination of the lesion. Microscopically, the key findings are marked lymphocytic hyperplasia, paracortical vascular proliferation with significant eosinophilic infiltration. Immunochemistry can show deposition of IgE in a reticular fashion between the follicles and on the surface of non-degranulated mast cells. Specific sequences of Epstein virus in the the skin lesions of a confirmed case of Kimura's disease have been seen & no parasites have been identified inspite of raised eosinophil count and focal necrosis which are highly suggestive [7]. The absence of Reed-Sternberg cells helps distuinguish Kimura's from Hodgkin's disease [2]. The aetiology of the disease is uncertain and presence of high eosinophil count and elevated serum levels of IgE suggests an allergic or hypersensitivity process. Surgery has been considered the mainstay of therapy, however, recurrence is common. The Kimuras disease responds well to oral steroids, but relapses often occur after treatment is stopped. Cyclosporine has been reported to induce remission in patients with Kimuras disease. However, recurrences of the lesions have been observed once the therapy is stopped. Steroids were then stopped and patient was put on 10mg/ d of cetrizine to prevent skin lesions.[10]

CONCLUSION

Kimuras disease though rarely seen should be kept as a differential diagnosis in a patient presenting with mass in head and neck region. Recurrence is high even after complete removal and patient should be on regular follow up and managed with intralesional and systemic steroids if recurrence occurs.

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