



## KIMURA'S DISEASE: A RARE CAUSE OF NECK SWELLING

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### ABSTRACT

KIMURA'S DISEASE is a rare condition and manifests as solitary or multiple subcutaneous nodules in the head and neck region, with eosinophilia and adenopathy. It is a benign chronic inflammatory disorder of unknown etiology. Histologically the lesions are characterized by hyperplastic lymphoid tissue, an inflammatory infiltrate rich in eosinophils and a proliferation of postcapillary venules. Hypereosinophilia in the blood and elevated levels of circulating IgE are found. The disease progresses slowly and is endemic to Asians. Treatment consists of surgical resection and administration of parenteral steroids in relapse cases. The prognosis is good and no malignant transformation has been ever observed.

### Introduction:

Kimura's disease was first described in China in 1937, but it was not referred to as Kimura's disease until its description in the Japanese literature in 1948.<sup>1,2</sup> It is a benign chronic inflammatory disorder of unknown etiology. It mainly involves the lymph nodes and subcutaneous tissue of head and neck region. It is often associated with regional lymphadenopathy. Rare sites include the kidney, orbit, ear, spermatic cord and nerves. The classical triad of Kimura's disease include single or multiple painless, slowly enlarging soft tissue mass, associated lymphadenopathy and peripheral eosinophilia, raised ESR & serum IgE. The disease usually presents as a single or multiple painless nodule.

The disease has a male predominance with peak age of onset in the third decade. Laboratory findings include peripheral eosinophilia and elevated serum IgE levels. Various systemic manifestations are reported, which include nephrotic syndrome, bronchial asthma, and gastrointestinal involvement. Renal involvement with proteinuria and nephrotic syndrome is the only systemic manifestation which has significant impact on progression of disease and its further prognosis. Renal impairment is probably due to immune complex-mediated damage or to Th2-dominant immune response disorders. Histological examination of the lesion shows lymphocyte infiltration, numerous eosinophils, germinal center formation and proliferative blood vessels.<sup>3</sup> Surgical excision is considered as the first line of treatment. Oral steroids are advised in cases of relapse. We are reporting this rare entity of Kimura's disease associated with right side neck mass in a patient managed successfully by surgical excision followed by oral steroids.

#### CASE REPORT

A 25 year old male patient presented to the department of Otorhinolaryngology in our institution with history of swelling on the Right side of the neck for 3 years. There was no history of fever, night sweats, weight loss. There was no past/family history relevant to tuberculosis. He had no history of taking medication and had no drug allergies. Other medical history was unremarkable. There was history of excision of the same mass 2 years back but it recurs after 4 months. No documents were available for the same. Clinical examination revealed swelling on the Right side of the neck in the supraclavicular region (figure 1). The swelling around 4x7 cm extends from midline to the right lateral side of the neck.

Three dots were present over the swelling indicating that some traditional treatment was carried out. The swelling was soft, non tender and the overlying skin was free from inner structures. There was no axillary, inguinal or any other palpable lymph nodes. Nasopharyngoscopy was normal. Chest X-ray was also normal.

Routine blood investigations were done. Blood picture shows peripheral eosinophilia and increased serum total IgE concentrations. All others laboratory tests were normal. To know about the vascularity of the mass Contrast Enhanced CT neck was done and it showed non enhancing mass. FNAC was done and it revealed moderate cellularity with mixed population of lymphoid cells and eosinophils suggesting lymphadenitis. Biopsy of the swelling was advised for confirmation. So excisional biopsy was planned.

The mass was removed under general anesthesia and sent for histopathological examination. The mass was measured 6.0 × 4.0 cm (figure 2). Histopathological examination of the excised lesions showed hyperplastic lymphoid tissue with proliferating germinal centers, moderate-to massive infiltration by eosinophils in their interfollicular and perivascular zones and a proliferation of postcapillary venules. There was no atypia/malignant change. These histologic features were consistent with Kimura's disease. Post-operatively patient was given oral steroids in tapering dose for 6weeks. The patient is symptom free till date.



Fig 1 showing right sided neck mass



Fig 2 showing excised mass

#### Discussion:

Kimura's disease was first described in Chinese literature in 1937. But in 1948, the terminology 'Kimura's disease' was first time mentioned in the Japanese literature. It is also called as eosinophilic lymphogranuloma. It is a benign chronic inflammatory disorder with unknown etiology and pathogenesis. Allergic reactions, infections and autoimmune reactions with an aberrant immune response have been suggested as possible etiological factors.<sup>3</sup>

It is most commonly seen in Asian continent (especially in China and Japan) and sporadic in the non-Asian population. There is marked male predominance with male to female ratio of 3.5 to 7 and the peak age of onset is during the third decade.<sup>4</sup> Kimura's disease usually presents as a slow growing solitary or multiple deep subcutaneous nodules, accompanied by satellite adenopathies and/or salivary gland hypertrophy (mainly parotid and submaxillary glands). As the masses enlarge very slowly, patient is usually asymptomatic. The nodules can be pruritic and painful but the overlying skin is normal. Some rare sites involved are oral cavity, groin, limbs, trunk, eyelids, orbit and lacrimal glands.

Laboratory findings include peripheral blood eosinophilia and elevated serum IgE levels.<sup>5</sup> Proteinuria must be systematically evaluated. For diagnosis of Kimura's disease, fine needle aspiration cytology (FNAC) is a safe procedure, but has only limited value. Diagnosis is usually based on the excision biopsy. Histopathological examination of involved lymph node reveals a characteristic triad of florid reactive follicular hyperplasia, increased vascularity and marked eosinophilia of the paracortical region. The germinal centres are often well vascularized and contains polykaryocytes, proteinaceous deposits and eosinophils. Nodal and extra nodal soft tissue lymphoid follicles show folliculolysis with infiltration by eosinophils.

The immunochemistry staining with IgE shows a characteristic reticular staining pattern on the processes of follicular dendritic cells of the germinal centres.<sup>6</sup>

The differential diagnosis include angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin's lymphoma, Langerhan's cell histiocytosis, allergic granulomatosis, drug reaction, parasitic lymphadenitis. Both Kimuras and ALHE diseases usually present with soft tissue masses in the head and neck region, but in angiolymphoid hyperplasia with eosinophilia, the lesions are mostly dermal or subcutaneous and not often in lymph nodes. Angiolymphoid hyperplasia with eosinophilia is more typically seen in middle-aged women and Kimura's disease in younger men. Histological changes suggest that ALHE is neoplastic in origin, whereas Kimura's disease is an immune-mediated disorder.<sup>7</sup>

The optimal treatment for Kimura's disease is controversial till today. If the mass is asymptomatic and nondisfiguring then wait and watch policy has to be applied. In symptomatic or disfiguring mass treatment is mainly surgical resection, but relapses are frequent. Corticosteroids are used in relapse cases but disease usually recur on treatment withdrawal. Certain tumors may become refractory to treatment. Radiation treatment is usually used for the local control of lesions not responsive to steroids, and total doses of 20 to 30 Gy have proved effective. Other treatments include the intralesional administration of steroids, cytotoxic agents, and electrodesiccation. The prognosis of Kimura's disease is good and no malignant transformation has been reported.<sup>8</sup>

## CONCLUSION

Kimura's disease is a benign chronic inflammatory disorder of unknown etiology. It is a rare entity usually seen in Asians and has male predominance. It presents with painless soft tissue nodules and lymphadenopathy in the head and neck region. Laboratory examinations reveal peripheral eosinophilia and elevated serum IgE levels. Diagnosis is confirmed histologically. Surgical resection is the first line of treatment but relapses are frequent. Steroids are given in relapse cases. It has a benign indolent course.

## References:

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