Abstract:
A 24 year old female patient presented with multiple, painless swellings in the neck and axilla for 6 months. Routine investigations were inconclusive except elevated eosinophilic count. Clinically it appeared to be lymphoma. Fine needle aspiration cytology was reported to be a reactive hyperplasia while excision biopsy of the cervical lymph node revealed the diagnosis as Kimura’s disease. This disorder should be suspected when painless cervical adenopathy, hypereosinophilia and hyper-IgE is present, particularly in male Asian patients.

Keyword: Kimura’s disease, lymphadenopathy, eosinophilia and hyper-immunoglobulinaemia.

Introduction
Kimura’s Disease (KD) is an allergic inflammatory disorder of unknown cause characterized by swelling in the head and neck region with the involvement of subcutaneous tissue, major salivary glands and lymph nodes. It is a benign condition but may be mistaken for malignant disease. Kimura’s disease is often accompanied by lymphadenopathy, eosinophilia and elevated IgE concentration. Renal involvement is its only systemic manifestation. Various treatment modalities have been tried including oral and intralesional steroid, surgical excision, cryotherapy, laser therapy and radiotherapy. However, the disease is usually persistent and difficult to eradicate.

Case History
A 24 yr old female, presented with complaints of multiple cervical swelling on both sides of the neck since 6 months which was slowly increasing in size. She had no history of any constitutional symptoms. Patient gave history of FNAC from the cervical swelling 15 days back, as advised by the treating physician at a private clinic, which was reported as reactive hyperplasia. On physical examination she was fit and healthy woman. The significant clinical finding was bilateral cervical lymphadenopathy in the level II, III, IV and V on both sides of the neck. The swellings were firm, non tender and immobile. The largest node present on the left side of the neck measuring 7x6 cm in size. The overlying skin was normal with no discharging sinus. Bilateral axillary lymph nodes were also palpable.
Investigations
1 Routine blood investigations and chest x-ray were normal except for eosinophilic count (21%)
2 CT NECK: Showed bilateral level II, III, IV, V and VI lymphadenopathy
3 CT CHEST: Showed cystic necrosis in the sternoclavicular compartment with invasion to the anterior mediastinal lymph nodes with multiple axillary lymph nodes
4 Excision biopsy of left level II lymph node revealed features suggestive of Kimura’s disease (figure 2)
5 IHC was done to rule out lymphoma due to multiple nodal involvement which proved to be negative

Histopathology findings:
Biopsy from level II lymph node showed multiple foci of necrosis, microabscess and karyohytic debris. Also seen aggregates of histiocytes admixed with numerous eosinophils. The stroma showed vascular proliferation and focal areas of lymph stasis. No epitheloid granulomas were seen. Picture suggestive of Kimura’s disease.
Figure 2: section showing eosinophilic tissue infiltration consistent with Kimura’s disease.

With this pathological diagnosis, literature was reviewed and specific investigations were done.
1) Immunoglobulin levels were done and the levels of which were as follows:
   a) IgG: 17 g/l (normal range: 5 to 12 g/l)
   b) IgE: 920 microgram (normal range: 30 to 280 microgram)
2) Candida specific antibody level was not significant.
3) Renal function test was normal and confirmed that patient had no renal involvement.

Based on hypereosinophilia, histology and negative serological testing for candida, a clinical diagnosis of Kimura’s disease was made and, Patient was put on oral steroids as surgery was not feasible due to massive nodal involvement in bilateral neck, both axilla and anterior mediastinum.

Figure 1: female patient with cervical lymphadenopathy.
Later, she was referred to radiotherapy department where we have decided not to irradiate her but advised regular follow up. She remains well on follow up without constitutional symptoms or renal involvement.

**Role of radiotherapy**

Radiotherapy may be indicated for patients with recurrent tumor or relapse after steroid withdrawal. Hareyama et al achieved local control in 90% of cases using radiotherapy at dosages of 26-30 Gy and they strongly recommend that no surgical procedure other than a biopsy should be carried out. Limited field radiation should be considered involving the lesion and adjacent lymph nodes, with an optimum dosage of 26-30 Gy irrespective of tumor size (Hareyama et al. 1998). However, considering the benign nature of the disease, radiotherapy should be considered only in cases of recurrent or disfiguring lesions.

**Discussion**

Kimura's disease is a benign condition where the aetiology is not known. It was first described in 1937 by Kimm and Szeto in China; who reported seven cases of benign lymph node enlargement with eosinophilic infiltrate, which they termed as eosinophilic hyperplastic lymphogranuloma (Kimm & Szeto 1937). Kimura et al. (1948) from Japan reported a similar finding and described it as an "unusual granulation and hyperplastic changes of lymphatic tissue", and since then this condition has become widely known as Kimura's disease (Kimura et al. 1948). Most cases of KD reported in males especially in the Oriental population. KD disease can occur at any age but most of the cases have reported in the second and third decades of life, with 80-87% of the affected patients being males (Sawada & Nomura 1984; Peters et al. 1986; Kuo et al. 1988). The pathophysiology of Kimura's disease is not understood at this time, but may relate to a disturbance in the normal rate of production of eosinophils and IgE. It is believed to be a product of an interaction between types 1 and 2 (Th1 and Th2) T helper cells. Such a derangement could result in excessive elaboration of eosinophilotrophic cytokines such as interleukin 4. Patients with Kimura's disease have been shown to have high levels of circulating eosinophilic protein, with heavy concentrations of IgE in their tissues. Allergic, bacterial, viral (e.g. Epstein-Barr virus), fungal (e.g. candida albicans) and parasitic aetiologies have also been linked to KD but not identified (Takenaka et al. 1976; Tabata et al. 1992; Wierenga et al. 1993).

The differential diagnosis for KD are many and they include angiolymphoid hyperplasia with eosinophilia (ALHE), lymphocytoma, Hodgkin's disease, lymphoma, Kaposi's sarcoma, tuberculosis, eosinophilic granuloma, epithelial haemangioma, angiofollicular hyperplasia, low grade angiosarcoma, hamartoma, Mikulicz disease and nodal metastasis. KD can be easily distinguishable from the clinical and histological features of these diseases, except for ALHE (Kung et al. 1984; Urabe et al. 1987). The clinical course of Kimura's disease is benign. The subcutaneous masses are usually found in the head and neck region, sometimes affecting the parotid or minor salivary glands, axillary and inguinal nodes. If not treated, these masses tend to slowly enlarge and may eventually become disfiguring. There is no consensus on the management aspect however a conservative surgical approach may be sufficient with use of other modality of treatment reserved.
References:


