CASE REPORT

Kimura's Disease: A Case Report and Review of Literature

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Abstract:

Kimura's Disease (KD) is a benign chronic inflammatory disorder attributed to an immune mediated hypersensitivity. It typically manifests by painless subcutaneous nodules in head or neck region and often accompanied by regional lymphadenopathy, salivary gland involvement along with hyper eosinophilia and elevated serum IgE. Most cases have been described predominantly in Chinese and Japanese people. Kimura's disease has been confused with Angiolymphoid Hyperplasia with Eosinophilia (ALHE), from which it probably should be distinguished as a separate entity. The course is usually benign except for the potential cosmetic disfigurement. The diagnosis may be suggested by a fine needle aspirate, but is established by a biopsy. A case of Kimura's disease in a 42 year male with recurrent left submandibular swelling is presented together with a brief review of the literature.

Keywords: Head and Neck Region, Benign, Chronic Inflammatory, Recurrent, Hypereosinophilia

Introduction:

Kimura Disease (KD) is a rare benign chronic inflammatory disease that usually involves deep subcutaneous tissue and lymph nodes of the head and neck region with frequent regional lymphadenopathy or salivary gland enlargement [1]. Other sites of involvement including the oral cavity, axilla, groin, limbs, and trunk have also been described [2]. Systemic symptoms (fever, night sweats, and weight loss) are not common. This disease is most common in middle-aged Asian men. It is endemic in Asia (China and Japan) and sporadic in the non-Asian population [2]. The peak age of onset is the third decade. Elevated serum Immunoglobulin E (IgE) levels and peripheral blood eosinophilia are also common [3]. KD was first reported by Kimm and Szeto in 1937 in China. In 1948, a Japanese doctor named Kimura *et al*. [4] published a systemic description of the disease and formally coined it as 'Kimura Disease'. Our case was classical presentation with respect to the fact that it involved all three possible areas of preference: The subcutaneous tissue, salivary gland (submandibular gland) and lymph nodes.

Case Report:

A 40 year male patient came with complaint of swelling in left submandibular region (Fig.1) since 6months which was firm in consistency, mobile, non-tender and non-adherent to the skin associated with cervical lymphadenopathy. There were no symptoms to suggest pulmonary tuberculosis such as chronic cough, night sweats, anorexia or loss of weight. Patient had history of similar swelling at same site two years ago for which excisional biopsy was done. There was no axillary or inguinal lymphadenopathy or hepatosplenomegaly. Initial laboratory investigations revealed normal indices of full blood count with slight eosinophilia with a rise in serum IgE levels (1750 IU / ml. Fine needle aspiration was inconclusive therefore Surgical excision of the lesion (Fig.2) was performed under general anesthesia**.** Histopathological study of the specimen confirmed the diagnosis of KD by demonstrating lymphoid tissue with reactive follicular hyperplasia (Fig. 3). Within the germinal

centers, deposits of abundant eosinophils and eosinophilc proteinaceous material were seen (Fig. 3). The interfollicular infiltrate was rich in eosinophils with admixed lymphocytes and eosinophilc micro-abscess (Fig. 4). The patient's recovery was uneventful and discharged well but lost to follow up.

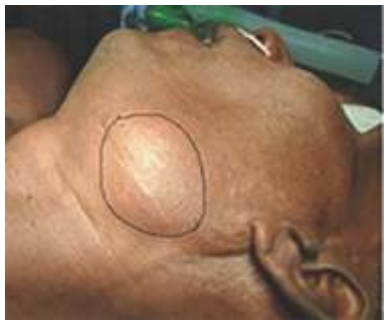

Fig. 1: Swelling in Left Submandibular Region

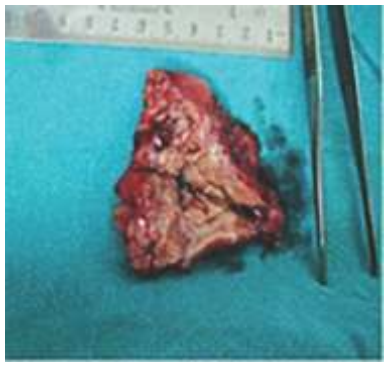

Fig. 2: Specimen excised in Toto

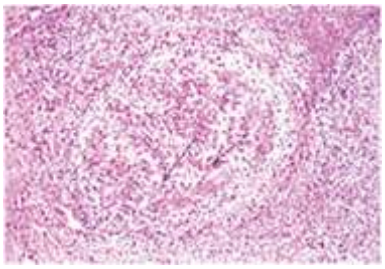

Fig. 3: Lymphoid Follicle Showing Innumerable Eosinophils with Deposits of Eosinophilic Proteinaceous Material (Black Arrows)

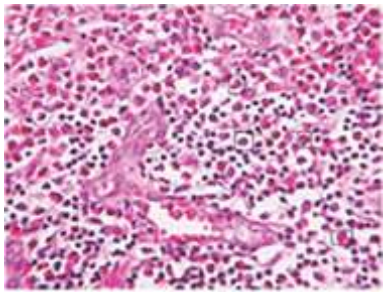

Fig. 4: Intense Eosinophilic Infiltration with Formation of Eosinophilic Micro-abscesses (Rectangles)

Discussion:

The etiology and the pathogenesis of KD are unknown. The disease is classified as a benign reactive process. Allergic reactions, infections, and autoimmune reactions with an aberrant immune reaction have been suggested [5]. The findings of increased eosinophils, mast cells, and levels of interleukin-5 and IgE suggest an abnormal T-cell stimulation to a hypersensitivity-type reaction [5]. The histological picture with the formation of lymphoid follicles and intense aggregates of eosinophils (which sometimes form micro-abscesses), vascular proliferation and fibrosis is highly suggestive of KD [6]. KD is endemic in the far east but only a few cases have been reported in non-Orientals. For many years it was thought that KD in Orientals corresponded with the later stages of ALHE in Caucasians [7]. This may explain why the disease has not previously been reported as a separate entity in the English otolaryngological literature despite a predilection for the head and neck region. A path breaking study by Rosai *et al*. (1979) [8] eventually clarified this misconception, and thus KD and ALHE were established as two distinct entities. Diagnosis of KD is always a clinical dilemma with no specific diagnostic guideline. Clinically, among other differential diagnosis of KD would include Kikuchi disease,

Mikulicz's disease and most importantly Hodgkin and non-Hodgkin lymphoma.

Fine Needle Aspiration Cytology (FNAC) is useful as an initial investigation of KD, with the main cytologic features of high number of eosinophils in a background of lymphoid cells. Nevertheless, histopathology examination of excised lesion is required for a definitive diagnosis as cytology may sometimes be difficult to interpret.

There is no agreement or uniform treatment protocol of KD [9]. Therefore, its management involves a multidisciplinary approach. Treatment alternatives for KD consist of surgical resection, cytotoxic and radiation therapy, as well as regional or systemic steroids therapy [9]. In asymptomatic cases, conservative observation is often adequate as lesions occasionally undergo spontaneous resolution. The role of surgery is mainly for diagnostic and cosmetic purposes. Systemic corticosteroids may be used with or without other treatment modalities. Treatment with intralesional corticosteroid (triamcinolone acetonide) had been reported to have good outcome [9]. Relapses are common and recurrence rate is as high as 40% despite early treatment of the patients [10]. Though there is no consensus for the treatment of recurrent disease, the overall outcome is good as there is no association with malignancy.

Conclusion:

Even though rare, KD should be considered in the differential diagnosis of a recurrent head and neck subcutaneous mass. FNAC is a supportive tool, but histopathology is indispensable for diagnosis. The aim of management primarily is preserving function and cosmetics of the tissues affected. Although, prognosis can be good, a complete cure is sometimes unattainable.

References

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