Kimura's disease: an unusual case of neck mass

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Abstract Kimura's disease usually affects young men of Asian descent and is characterized by swelling in the region of head and neck. A case of Kimura's disease is reported who presented with neck mass. Fine needle aspiration cytology was doubtful of lymphoma. Histopathological examination of excised specimen was reported as Kimura's disease.

Keywords Kimura's Disease · Immunoglobulin E · Eosinophilia · Angiolymphoid hyperplasia

Introduction

Kimura's is a rare idiopathic condition which is seen mostly in young men of Asian descent. The disease is characterized by swelling in the region of head and neck with involvement of subcutaneous tissue, major salivary glands and / or lymph nodes. Patients almost always have eosinophilia and elevated serum Immunoglobulin E levels. The diagnosis is established by biopsy.

Case report

A 32-years-old healthy young adult reported to ENT OPD with the symptom of a slowly increasing swelling in front of his neck for past 10 months. The patient did not have any systemic symptom such as fever, night sweat, weight loss, loss of appetite, cough or weakness. He was actively performing all works involved in his job as an aircraft fitter.

Clinical examination revealed a man of average built and nutrition with normal vital signs. Local examination showed a 3 × 3 cm size firm, mobile, nontender swelling in submental region. The swelling had smooth surface, well defined margins with a normal skin over it. No other lesion was seen in the neck. No abnormality was found in oral cavity, oropharynx, larynx and hypopharynx.

The patient had mild eosinophilia (8%) in his peripheral blood. Routine and microscopical examination of urine was normal. His HIV status was negative. FNAC was suggestive of Hodgkin's disease and an excision biopsy was recommended by the Pathologist. The lesion was excised in toto under local anaesthesia.

Histopathological examination revealed lymphoid follicles with germinal center at the periphery. In between the follicles the stroma showed vascular proliferation, lymphoplasmacytic infiltrates and numerous eosinophils. At places the stroma showed hyalinization and hyalinized vessels. No Reed Sternberg cells were seen. No epithelioid endothelial

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proliferation was noted. The findings were consistent with Kimura's disease (Fig. 1).

The patient was further evaluated for Immunoglobulin profile. IgE was found raised considerably at 425.00 IU/ml (reference range being less than 180.00 IU/ml). He was given oral steroid for two weeks. Three years follow up has shown no recurrence.

Discussion

Kimura's disease was first described as "eosinophilic hyperplastic granuloma" by Kim and Szeto in China in 1937 [1]. Kimura himself reported on the disease in Japan in 1948 [2]. This rare clinical condition mainly affects young Asian men. Male: female ratio is 3:1.

The most common presentation is a gradually increasing swelling in the region of head and neck over a period of months or years. The lesion primarily involves the subcutaneous soft tissues and sometimes the salivary glands and lymph nodes; though cases involving oral cavity, conjunctiva, eyelid, tympanic membrane, skeletal muscle, prostate, kidney, peripheral nerve and epiglottis have been reported.

Peripheral blood eosinophilia and an elevated serum immunoglobulin E (IgE) levels are consistent laboratory findings in Kimura's disease. About 12% of patients also have associated renal disease characterized by proteinuria [3].

The definitive diagnosis of Kimura's disease is made by histologic examination of the excised lesion. The key microscopic findings are marked lymphoid hyperplasia with pronounced eosinophilic infiltration. Foci of necrosis are seen with vascularization of the paracortex and deposition of hyaline material within follicles. Polykaryotic giant cells are common feature. Deposition of IgE in reticular fashion

between the follicles and on the surface of the non-granulated mast cells can be seen on immunostaining [4].

When the diagnosis is attempted by fine needle aspiration cytology the polymorphous infiltrate with eosinophilia and the presence of giant cells raise the suspicion of Hodgkin's disease but the absence of Reed Sternberg cells helps to rule it out [5].

Another important differential diagnosis is Angiolymphoid Hyperplasia with Eosinophilia (ALHE). The common features shared by these conditions include male predominance, predilection for head and neck region, tendency to recur and vascular nature of the lesion with lymphoid and eosinophilic infiltrates. However the ALHE lesions are multiple dermal popular or nodular eruptions in older patients less frequently accompanied by eosinophilia in peripheral blood. The main histopathological difference is the presence of "histiocytoid" and "epithelioid" blood vessels in ALHE, a feature that is lacking in Kimura's disease [6].

The etiology of Kimura's disease is still unknown. The consistent laboratory findings of peripheral blood eosinophilia, raised serum IgE levels and the histologic picture depicting local tissue eosinophilic and lymphoplasmacytic infiltration suggest an allergic process though no specific allergen have been identified. The parasitic cause has also been looked into but it has not been established. The current concept is that the disturbance resulting in excess production of eosinophil and IgE in Kimura's disease is a product of interaction between Type 1 and Type 2 T helper cells (Th 1 and Th 2) which results in excessive elaboration of eosinophilotropic cytokines such as Interleukin 4 [7].

The treatment of choice for localized disease is surgical excision [1]. There have been reports of recurrence in 15% to 40% of cases even after apparently adequate surgical excisions and excision biopsies which may be because of poor delineation of the lesion.

Kimura's disease also responds well to oral steroids [8], but relapses occur after the treatment is stopped. Other modes of treatment include intralesional injection of steroids and radiation therapy. Kimura's disease has no potential for malignancy.

Fig. 1 Micro-photograph of the tumour showing vascular proliferation with lymphocytic and eosinophilic infiltrate. H& E. Magnification $\times 40$

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