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Kimura's disease: a rare diagnosis of a forehead swelling

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Abstract: *Kimura's disease is a rare chronic benign disorder, presenting as an asymptomatic tumour like subcutaneous swelling, usually located in the head and neck region and is more prevalent among young adult males of Asian origin. It exhibits the clinical triad of a painless head or neck mass, eosinophilia, and an elevated IgE level. Here, we present a case report of this rare entity.*
Keywords: *Kimura's Disease, Eosinophilia, Parotid Gland, Lymphadenopathy, Angiolymphoid hyperplasia*

I. Introduction

Kimura's disease is a rare chronic benign disorder, presenting as an asymptomatic tumour like subcutaneous swelling, usually located in the head and neck region. The disease was first described in the Chinese literature while the histological description was first published by Kimura et al in 1948[1]. Although a rare entity in the west, it is more prevalent among young adult males of Asian origin[2]. The etiopathogenesis of the disease remains unknown, but, it is now considered to be an aberrant immunological reaction to some unknown antigenic stimulus[2]. It is often accompanied by lymphadenopathy, salivary gland swelling, associated with tissue and blood eosinophilia, and raised serum IgE. Renal involvement may be seen in almost half of the cases.[2] Since it presents as an asymptomatic swelling, it is often ignored by the patient, and often the diagnosis might be missed in the Indian scenario, where the disease is not so common.

II. Case Report

A 50 year old male patient presented to us with a painless asymptomatic subcutaneous swelling over the forehead region of one year duration that was gradually increasing in size. He also had a low backache, on and off, since 3-4 months with a history of weight loss, though not significant. The forehead lesion was approximately 1.5× 1.5 cm in size, round, firm in consistency, non-tender, mobile with no changes of the overlying skin. The general physical and systemic examination of the patient revealed no abnormality except for cervical lymphadenopathy. The lymph nodes were 3-4 in number, 2-3cm in size, bilateral, non-tender, soft to firm in consistency, discrete, mobile. The differential diagnoses considered were lipoma, dermoid cyst, sebaceous cyst, Kimura's disease and secondary metastatic deposit from an undetected malignancy.

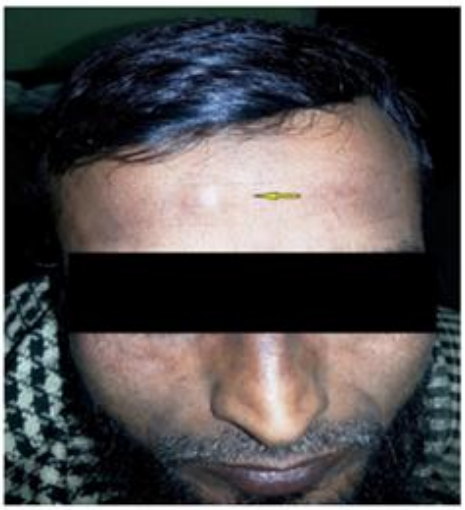

Fig 1: Kimura's disease: The patient had forehead swelling.

The routine blood examination revealed eosinophilia, with normal total leucocyte count and hemoglobin. Peripheral blood smear was normal. The liver function tests, renal function tests, X-ray chest and lumbosacral spine, USG whole abdomen were within normal limits. USG neck and parotids detected the cervical lymphadenopathy and lumpiness of the parotid glands. Serum IgE level was found to be elevated (922 IU/ml). FNAC was done from the forehead swelling which showed groups of large cells having pleomorphic nuclei with prominent nucleoli which was confusing, as these were suggestive of malignancy. Therefore, excision biopsy was done from the forehead swelling, and it revealed an inflammatory mass of tissue composed of lymphoid follicles with prominent germinal centres with areas of eoisinophilic infiltration, and surrounded by fibrocollagenous tissue. Vascular proliferation was also noted comprising of thin walled blood vessels. These histopathological features were consistent with Kimura's disease. Thus conglomerating the clinical, laboratory and histopathological features, that is, clinical nature of the swelling with lymphadenopathy, parotid lumpiness on ultrasonography, elevated serum IgE and from the histopathological findings, the conclusive diagnosis of Kimura's disease was arrived at.

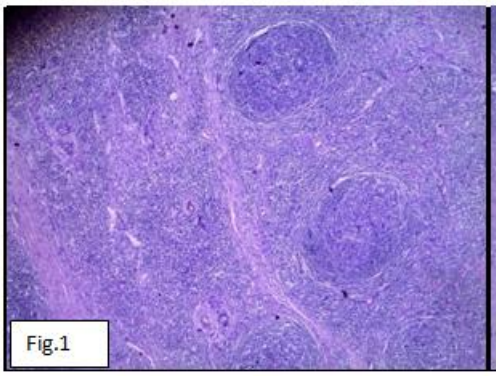

Fig1: Lymphoid follicles with prominent germinal centre in a background of fibrocollagenous tissue.

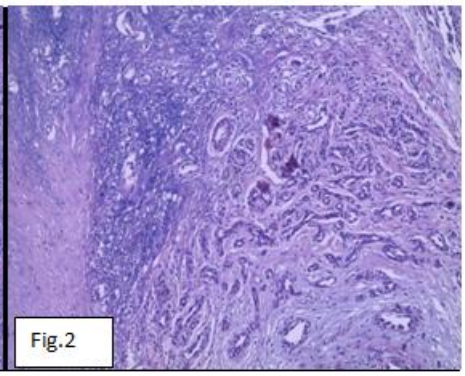

Fig 2: Proliferation of thin walled blood vessels

III. Discussion

Kimura's disease is a rare entity of unknown origin. Although the etiology of the disease remains unclear, it is thought to be an aberrant immune reaction to some unknown antigenic stimulus.2

Most of the patients are male, oriental young adults, and the prevalence in patients of other ethnicities is considered low. The disease is characterized by the development of soft subcutaneous masses commonly over the head and neck region, often accompanied by blood eosinophilia and marked increase in serum levels of immunoglobulin E (IgE). It can often have a relapsing course and recurrences are seen to occur despite treatment. [2,3]

Kimura's disease should be suspected when a patient— particularly an Asian male—exhibits the clinical triad of a painless head or neck mass, eosinophilia, and an elevated serum IgE level.4 There can also be salivary gland swellings and renal involvement presenting as nephrotic syndrome.3 The soft tissue masses in Kimura's disease often mimick an inflammatory or neoplastic process clinically. Our patient had a single asymptomatic swelling which on FNAC showed plump endothelial cells on pap smear that are characteristic of Kimura's disease, which were mistaken for malignant cells.

Salient histopathological findings of Kimura's disease are characterized by lymphoid infiltration with reactive follicles, dense fibrotic stroma and a mixed inflammatory cell infiltrate with numerous eosinophils which may form eosinophilic abscesses. All of these can develop in subcutaneous tissue, salivary glands, and lymph nodes.6 Vascular channels are also seen. The histopathological findings of lymphoid follicles with tissue eosinophils in a background of fibrocollagenous tissue along with vascular proliferating channels seen in our patient were consistent with Kimura's disease. Renal involvement can also occur in Kimura's disease, often presenting as nephritic syndrome due to various types of gomerulonephritis[4], which was however absent in our case.

A close histopathological differential is ALHE(angiolymphoidhyperplasia with eosinophilia). In ALHE, the lymphoid infiltration is more diffuse and only occasionally lymphoid follicles are found. Eosinophilic infiltration is more variable, ranging from sparse to abundant, and eosinophilic abscesses are rare. In ALHE, thick-walled blood vessels are lined with hypertrophic low cuboidal to dome-shaped "epitheloid" or "histiocytoid" endothelial cells containing vacuolated eosinophilic cytoplasm with vesicular nuclei. These endothelial cells protrude into the vascular lumen, occasionally resulting in a "cobblestone" appearance, and they may occlude the lumen or form solid cords. In contrast, the blood vessels in Kimura's disease show either

flattened or low cuboidal endothelial cells which do not exhibit the characteristics of the "epitheloid" or "histiocytoid" cells.[4]

However, a controversy exists whether Kimura's disease and ALHE are two separate entities, or different spectrum of the same disease. Chong et al in 2006 reported a case with coexisting features of both Kimura's disease and ALHE, in a 40 year old Chinese female.4 The lesions of Kimura's disease are in the deep subcutis whereas in ALHE it is mostly small dermal palpulonodules. Lymphadenopathy is also rare in ALHE. While Kimura's is common in the young Oriental males, ALHE usually is seen to occur in middle aged females. [4,6]

Till date no consensus has been reached regarding the treatment of Kimura's disease but various modalities have been tried, like surgical excision, steroid therapy, radiotherapy, cyclosporine, cryotherapy, pulsed dye laser and orl retinoids. [4,5] Messina-Doucet et al have suggested surgical excision as the treatment of choice for definitive diagnosis and initial management.[7] However recurrence can occur in the treated lesions. Localized initial recurrence can be managed by surgical excision. If recurrence becomes frequent, oral corticosteroids should be initiated. If all else fails, radiotherapy can be considered, but side-effects are the limiting factor for use in benign lesions.[4]

Thus, to conclude, although Kimura disease is a rare disorder, it should be considered in the differential diagnosis of any subcutaneous swelling demonstrating an eosinophilic infiltrate and prominent follicular hyperplasia. With proper histologic criteria, we think a correct diagnosis can be achieved, especially when combined with pertinent clinical information and laboratory studies.

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