

Kimura's Disease - A Rare Clinical Presentation of a Subcutaneous Breast Lesion in an Asian Female

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ABSTRACT

We report a case of a 54 year old Asian woman referred to our hospital for a subcutaneous right breast swelling for the last one year. No other systemic symptoms were present and a mild hypereosinophilia was found in routine blood tests. To rule out any neoplastic lesion and reach a specific diagnosis, Excision biopsy of the lesion under local anaesthesia was performed. Histopathological examination showed lymphoid infiltrates with prominent reactive germinal centres along with infiltration by eosinophils and vascular proliferation, a diagnosis of Kimura's disease of breast was made. The patient was put on symptomatic treatment and clinical follow up revealed complete regression of the swelling and disease. This case has been reported due to its extreme rarity and highlights the need for including Kimura's disease in the differential diagnosis of subcutaneous breast swellings.

Keywords: Kimura's disease, Subcutaneous swelling, Inflammatory disorder

INTRODUCTION

Kimura's disease is a chronic inflammatory disorder of unknown aetiology. It is seen in an endemic form in the orient [1] but also in other parts of the world including the United States and Europe [2]. It typically presents as painless, sometimes disfiguring subcutaneous nodules predominantly in the head and neck [3]. It is often accompanied by regional lymphadenopathy, raised eosinophil count and markedly elevated serum Immunoglobulin E (IgE) levels [1,4]. On histopathology the lesions are characterised by reactive lymphoid follicles with eosinophilic infiltrations, sometimes forming eosinophilic abscesses [1,5,6]. Initially Kimura's disease can be confused with inflammatory lesion or lymphoma, but a definite diagnosis can be established after biopsy. Surgical excision the prognosis is good and malignant transformation has not been reported till date. Early diagnosis of the disease could spare the patient unnecessary and potentially harmful diagnostic procedures. The Kimura's disease of the breast should be kept in differential diagnosis of breast swellings in rarest of rare cases.

CASE REPORT

A 54 year old Asian female presented to our Surgical OPD with a painless, pruritic, subcutaneous swelling on the Right breast (upper outer quadrant). She reported that it was initially small and had progressively increased in size over 1 year. She has no other associated symptoms. Her medical,

surgical and family histories were non-contributory. She was not taking any medications and had no drug allergies. There was no history of travel to any other country and she denied tobacco, alcohol or drug use. On physical examination she had a single discrete subcutaneous swelling on the upper outer quadrant of the right breast measuring 2 × 2 cm. The swelling was fixed and non-tender with smooth surface. There was no other swelling on the left breast. Laboratory values were significant for a white blood cell (WBC) count of 8.9/10³ µl (reference range: 4.5-10.5/10³ µl) with an eosinophil count of 880 cells/mcl (reference range: 15-550 cells/mcl). Her Hb level was 12.6 g/dl (reference range: 11.5-15 g/dl). Serum IgE level was 1599 ku/L (reference range: ≤ 114 ku/L). Other parameters were within normal limits.

After surgical excision, histopathology showed skin lined tissue with underlying dermis showing abundant lymphoid follicles with germinal centres. There was a dense infiltration by mature eosinophils along with prominent

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vascular proliferation (**Figures 1-3**). Focal eosinophilic abscesses were also seen and a diagnosis of Kimura's disease was established. The patient had an uneventful postoperative course and is doing well after 4 months follow up.

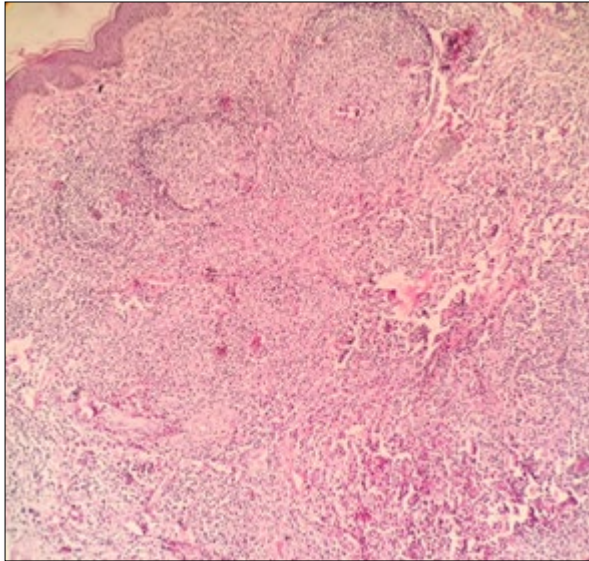


Figure 1. Epidermis lined biopsy showing lymphoid follicles (H&E 4x).

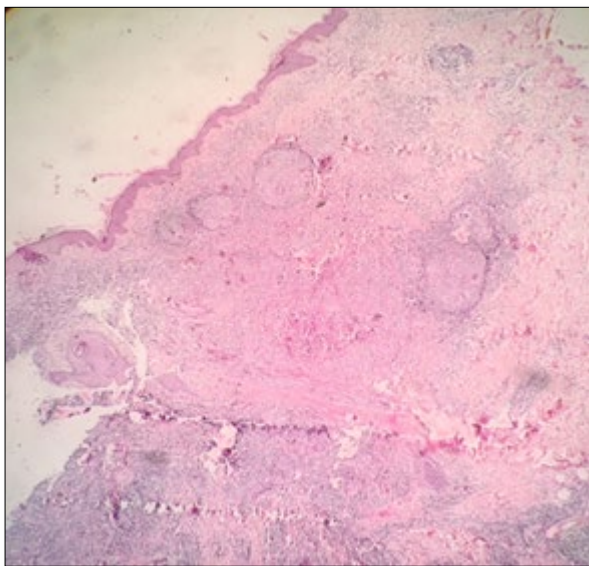


Figure 2. Aggregates of eosinophils and proliferating vascular channels (10x).

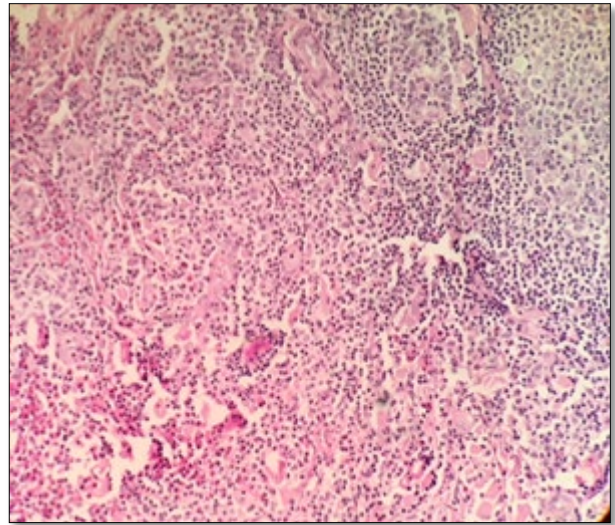


Figure 3. Aggregates of eosinophils and proliferating vascular channels (H&E 20x).

DISCUSSION

Kimura's disease is a rare entity in the west but endemic in Asia. It is a benign chronic inflammatory disease with unknown aetiology, although a possible relation to Epstein Barr Virus (EBV) has been suggested [7]. Another case has been reported in an HIV positive patient after primary infection with HHV-8, suggesting a possible relation to Kimura's disease [8].

The disease has a male predominance and the clinical presentation is variable. It typically presents as solitary or multiple subcutaneous nodules slowly increasing in size. The lesions are variably painful and pruritic; however the overlying skin is normal. The most common site of involvement is the cervical region [9]. It may occur at other unusual sites including limbs, groin and scalp and as retro auricular masses [10]. Occurrence of this condition in a female as a subcutaneous breast lesion is a rare clinical presentation. There is an important association between Kimura's disease and renal diseases particularly nephritic syndrome and the frequency can go up to 60%; well exceeding that of the general population [11-13]. Laboratory findings include peripheral eosinophilia and elevated serum IgE levels.

Definitive diagnosis of Kimura's disease is based on histological findings. The constant features are prominent lymphoid follicles with florid germinal centres, eosinophilic infiltration and eosinophilic microabscess formation. The interfollicular zone shows vascular proliferation [14]. Immunochemical findings are IgE reticular network in the germinal centre and IgE coated non degranulated mast cells [6]. A surprising finding has been the presence of a clonal rearrangement of the T-cell receptor delta gene in a single case of the disease [15].

Despite early statements to the contrary, current evidence suggests that Kimura's disease and disease known to dermatologists as Angio Lymphoid Hyperplasia with Eosinophilia (ALHE) are different entities, specifically the former disorder lacks the epithelioid (histiocytoid) endothelial cells that are the morphologic hallmark of the latter. Both may present with fibrous tissue proliferation with a plasma cell infiltrate, however epithelial and non-epithelial adnexal structures may be spared [16,17].

The treatment of Kimura's disease is variable, with surgical excision being the obvious choice. However, the lesion has a tendency to recur, other therapeutic options such as radiation, systemic corticosteroids and cytotoxic agents have tried with inconsistent results. The management of Kimura's disease differs with the condition of patient, with surgical, medical or radiotherapy being the line of treatment. Surgery may be contemplated in younger patients with localised primary lesions or in localised recurrences. In patients with recurrent disease with renal involvement, intra lesional steroids and oral steroids, cyclophosphamide and anti-platelet drugs have been used with reasonable success rates; however, radiotherapy may be indicated in unresectable masses and recurrent masses unresponsive to medical therapy.

CONCLUSION

Kimura's disease is a medically benign disorder, which is typically chronic. Although it has no known malignant potential, prompt and accurate diagnosis can be made sparing the patient any unnecessary and potentially harmful diagnostic procedure. The purpose of this article is to emphasize the importance of knowledge regarding Kimura's disease among clinicians and pathologists, since it can mimic various inflammatory and neoplastic conditions. A rare clinical presentation of Kimura's disease should be kept in mind while evaluating lesions in the breast region.

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