

KIMURA DISEASE IN OLDER ADULTS: TWO CASE REPORTS AND PATIENT'S PERSPECTIVE

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ABSTRACT

Background: Kimura's disease (KD) is a rare, chronic, benign inflammatory disorder that mostly affects men of Asian descent during their 2nd–4th decades of life and is less common in older adults. Typically, the disease presents with painless, slow-growing subcutaneous swellings, increased peripheral eosinophilia, and elevated serum IgE levels. While KD sometimes mimics neoplastic conditions, the diagnosis is generally confirmed by excisional histopathological findings. **Case Reports:** We present two cases (68-year-old and 72-year-old males, respectively). Elevated serum immunoglobulin E (IgE) and peripheral blood eosinophilia are significant clinical clues for diagnosing KD in both cases. Surgical excision and selected steroid therapy are the modes of treatment. No recurrences were reported during the 12- and 18-month follow-ups, respectively. **Conclusion:** The old-age onset of the disease underscores its importance, particularly as many older adults have compromised health status therefore Kimura disease in older adults requires special attention and prompt treatment. A note on the patients' perspectives is provided.

KEYWORDS

Aged, Kimura Disease, IgE, Eosinophilia, Biologics, Misdiagnosis

INTRODUCTION

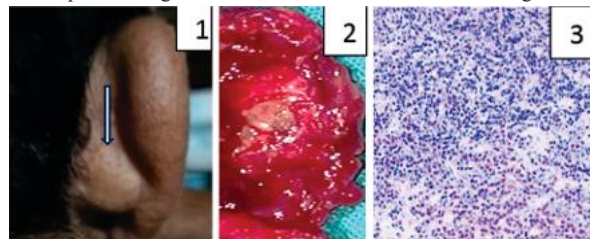
Kimura disease (KD) is a rare, benign, painless, chronic inflammatory disorder of unknown aetiology, which triggers subcutaneous swellings and lymphadenopathy¹. No malignant transformation of KD has yet been reported but the disease is infamous for its high recurrence rate². Because Kimura disease predominantly presents as nodular swelling in the head and neck, its symptoms often share or mimic those of other diseases, frequently leading to misdiagnosis³. Uncommonly Kimura disease has been detected in other parts of the body including visceral organs^{4,5}. This disease is most prevalent in the young Asian population while infrequently cases have been documented among Caucasians, Africans, and Americans^{4,6,7}. An elevated serum immunoglobulin E (IgE) and peripheral blood eosinophilia are the key clinical clues for diagnosing KD, which is then confirmed by histopathology⁸. Florid germinal centre hyperplasia with Warthin-Finkeldey polykaryocytes, extensive eosinophilic infiltrates and hyperplastic postcapillary venules are hallmark histopathological features². Renal involvement is the most significant systemic complication in Kimura disease, where proteinuria is a common indicator¹. Ultrasonography (USG)⁹, magnetic resonance imaging (MRI)^{5,10}, computed tomography (CT)¹¹, immunohistochemistry (IHC)⁶, fine needle aspiration cytology (FNAC)¹², hematological studies, and excisional biopsy are some of the diagnostic tools to study the nature of the lesion for diagnosis of KD. The primary choice of treatment is surgical excision for localized KD and excisional biopsy. Conservative treatment like oral corticosteroids, immunosuppressants or low-dose radiation are recommended to the small lesions or to prevent recurrence after surgical excision^{13,14}. However, target therapy (biologics) is the emerging effective treatment, while chemotherapy is a last-resort decision for refractory cases^{15,16}. Although Kimura disease is most commonly reported in the second to fourth decade of life¹⁷, we present two cases of older-age onset in patients of Indian descent. Because the study of KD in patients over 50 years age is inadequate, it is relevant to add some more observations to the existing knowledge.

Case Report 1

A 68-year-old male patient presented to the ENT outpatient department (OPD) of a tertiary hospital with complaints of right post auricular mass for the past 9 months (Fig1). He had no family history of any malignancy. There was no history of fever, night sweats, anorexia, or weight loss; however, patient has comorbid insomnia, dementia and hypertension. On clinical examination the swelling (2x1.5 cm) found nontender and painless. No satellite nodes were felt nearby. Ultrasonography showed a hypoechoic subcutaneous mass of 1.8 X 1.5 cm size. Fine needle aspiration cytology (FNAC) of the lesion is characterised by polymorphous reactive lymphoid cells, numerous eosinophils and a few Warthin-Finkeldey-like giant cells, suggestive of Kimura's disease.

Complete blood count showed haemoglobin of 13.5 g/dl, TLC 9700 /cumm with an absolute eosinophil count 27%. Liver function and renal function test found in normal range. Serum IgE was markedly

raised to 2642.3 IU/mL. No abnormal level of protein (proteinuria) detected in urine sample. As a front-line treatment, surgical excision with adequate margin was performed to debulk the lesion (Fig.2) and the mass was given for excisional biopsy. The histopathological results revealed the lymphoid tissue with follicles and eosinophil infiltration, confirming Kimura disease (Fig3). Post-surgery steroid therapy for Kimura disease was initiated with Wysolone 30 mg [oral prednisolone] for two weeks then tapered to 20mg for one month and then 5 mg for three months. No recurrence was noticed till 1 year of follow up. Eosinophilia and IgE level also came down to their normal range.

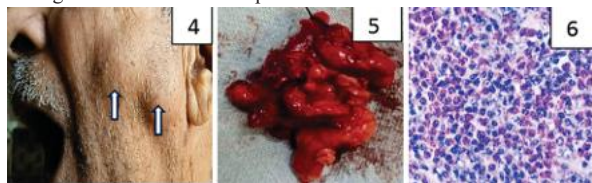


Figs. Showing Right Post Auricular Subcutaneous Kimura Lesion (1), Surgical Excision Mass (2) and Lymphoid Tissue with Follicle and Eosinophil Infiltration in Histopathology of Excisional Biopsy (3).

Case Report 2

A 72-year-old male patient presented to the OPD of oral and maxillofacial surgery in a tertiary hospital with a chief complaint of two soft masses and swelling on the left upper facial region. As other comorbid the patient had osteoarthritis, diabetes, hypertension. The patient had also the history of carious molar teeth extraction two months prior to presentation. On clinical examination the swelling size was 2.0 x 1.8 cm and 2.0 x 1.5 approx. (Fig.4). Palpable lymph nodes were not felt in the surrounding. The masses were nontender, painless and slow growing since last 1 year. Extraoral examination showed distinct lesions that were not attached to the ipsilateral maxilla or mandible. Stretching of the surrounding oral mucosa and overlying skin revealed no evidence of inflammation. Ultrasonography described the normal parotid salivary ducts and moderately enlarged lymph nodes. FNAC findings specifically lymphoid aggregates and eosinophilic infiltrate, indicative of Kimura disease. Proteinuria, haematuria, and casts were not there in the urine sample. Absolute eosinophil count was 39% and IgE was markedly raised to 4163.8 IU/mL. Surgical excision was done to debulk the masses (Fig.2). Incisional histopathology revealed follicular hyperplasia, reactive germinal centres, and eosinophilic infiltrates with microabscesses, confirming Kimura disease (Fig.3). Postoperatively, the patient was prescribed oral Omnacortil (prednisolone) at 1 mg/kg/day for four weeks, then the dose tapered to half for the following four weeks. Additionally, Xolair (omalizumab) 300 mg was injected subcutaneously at monthly intervals for three months. All parameters

of KD normalized within six months, and no recurrence was reported during the 18-month follow-up.



Figs. Showing Left Upper Facial Swellings (4), Excision Mass (5), Lymphoid Tissue with Follicle and Eosinophil Infiltration in Excisional Histopathology of Biopsy (6).

DISCUSSION

Kimura disease is an aberrant immune response of the body to an unknown aetiology. The pathogenesis of KD often involves IgE-mediated hypersensitivity, T-helper 2 (Th2) cell activation, and elevated interleukins (IL-4, IL-5, IL-13), eosinophils, and IgE production¹⁸. Specific KD genes have not yet been identified; however, the onset of the disease, mostly in young individuals of Asian ethnicity strongly indicates a gene-environment interaction¹⁸. Despite this established age-demographic trend, we report here two elderly cases.

Commonly affected sites of KD include head and neck region involving preauricular, postauricular and facial sites. Few cases of preauricular, postauricular, and neck site presentation reported in elderly patients^{13,15,19}. Localised swelling with mandibular KD mass²⁰, hard palate²¹ and a soft palate²² mass are¹³ also reported in older age patients.

Treatment of KD generally includes surgical excision, oral or parenteral steroids, administration, immunosuppressive agents, local low-dose radiotherapy. These treatments are recommended in combination or in isolation.

Simple excision of mandibular and facial KD masses in older adults was reported with no recurrence²³⁻²⁵. Exclusive wide resection with negative margin in two older patients did not show any recurrence KD over 5 years follow up⁵. Similarly, to prevent recurrence, a wide resection with negative margins was performed on a 64-year-old patient presenting with a clinically established mass on an extremity⁶.

In some cases, high-dose corticosteroid therapy was given to downsize the facial mass before surgical excision^{26,27}.

For two old males presenting with recurrent KD of the neck over two years, a combination of surgical excision and moderate-dose radiotherapy with standard fractionation were reported as promising treatment option^{14,21}.

Post excision short term oral administration of Prednisolone (1 mg/kg/day) claimed no recurrence of a facial KD mass²⁸. Whereas an elderly patient (59) with KD mass on neck has been successfully treated with surgical excision followed by immunosuppressant and a corticosteroid drug¹³. Comparable treatment was also given to a 70-year-old patient with painless KD mass in inguinal lymph node²⁹. A recurrence case of KD was reported in a 60 years old patient due to self-discontinuation of steroid against medical advice²¹.

A patient (62) with refractory KD mass on eyelid did not respond to any of above therapy but managed with CVP (Cyclophosphamide, Vincristine, Prednisone) chemotherapy¹⁶.

Of late, monoclonal antibodies (biologics) such as dupilumab, omalizumab (an anti-IgE antibody), and mepolizumab (an anti-IL-5 antibody) have been used as targeted therapy for long-standing and refractory KD (Fukami 2023)¹¹. While a 57-year-old patient with 7 years history of pre and post auricular KD was treated with corticosteroids (prednisone) followed by anti-IgE therapy of omalizumab. This treatment neither reduce the elevated IgE nor the peripheral eosinophil, but responded to dupilumab in manipulated doses for 4 months. Thus, a multi monoclonal antibody therapy could control the long-standing KD¹⁵. To a patient (55) with 14 years history of post auricular KD lesion and refractory prurigo as comorbidity, oral cyclosporine monotherapy and cyclosporine- prednisolone combination therapy was ineffective. However, biologic treatment (dupilumab) could effectively address both the issues.¹¹

Although no malignant transformation of Kimura Disease has been recorded, a case of coexisting lymphoma and KD was recently reported in a 72-year-old patient³⁰ where treatment is challenge.

In the present case studies, suitable treatment options are explored for non-recurrence of KD as older adults have compromised health status. A patient perspective, including take-home messages regarding KD management in old age is provided in Box 1.

CONCLUSION

Kimura disease in older adults requires special attention and prompt management, as these patients often have multiple comorbidities. Timely diagnosis and treatment can help to avoid complications of KD. Moreover, the self-care guidance always supports early recovery and prevention of KD recurrence in elderly patients.

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