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Case Report

Unusual presentation of Kimura's disease: a case series

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ABSTRACT

Kimura's disease is a rare chronic inflammatory disease of unknown etiology, presenting as painless subcutaneous nodules with lymphadenopathy and peripheral eosinophilia, mainly disturbing the head and neck region. It mainly affects Asian males in their 2nd to 4th decade of life. Kimura's disease, although difficult to diagnose clinically, should be considered in the differential diagnosis of patients who have a primary lymphadenopathy with eosinophilia with or without subcutaneous nodules. It should be investigated accordingly as the disease has an indolent course and good prognosis. Herein we report two cases of Kimura's disease, of which one had unusual site of involvement.

Keywords: Kimura's, Lymphadenopathy, Eosinophilia, IgE

INTRODUCTION

Kimura's disease is a rare chronic hyper-eosinophilic disorder affecting major salivary glands and cervical lymph nodes, commonly reported in young Asian men.

Although KD is popularly termed as angiolymphoid hyperplasia, pseudopyogenic granuloma and atypical pyogenic granuloma, they are different entities. Only documented systemic involvement is nephrotic syndrome.[1]

Herein we present two such baffling cases diagnosed as Kimura's disease, contributing to better aid in their management.

CASE REPORT

Case 1

A 22 year old male presented with insidious onset of a gradually progressive asymptomatic swelling of size approximately 3x2 cm over left post-auricular area since 2 years. On examination it was an oval-shaped, non-tender, soft and non-compressible subcutaneous swelling devoid of textural changes (Figure 1). Systemic examination was unremarkable.

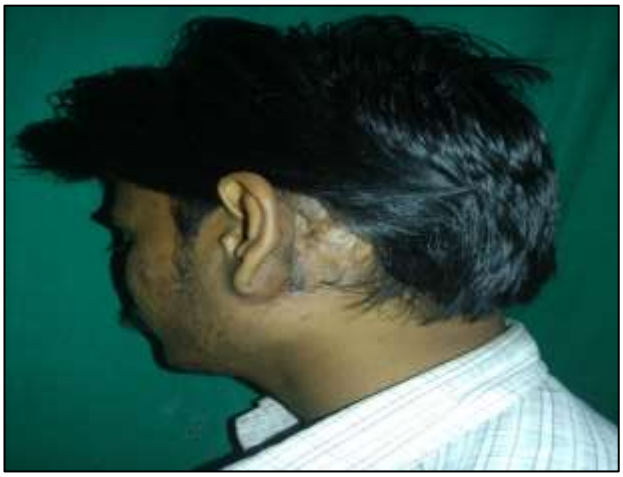

Figure 1: Case 1: 3×2 cm oval-shaped swelling over left post auricular area with normal overlying skin.

Differential diagnosis of lymphadenopathy, lipoma, dermoid cyst and Kimura's disease were considered and case was investigated accordingly.

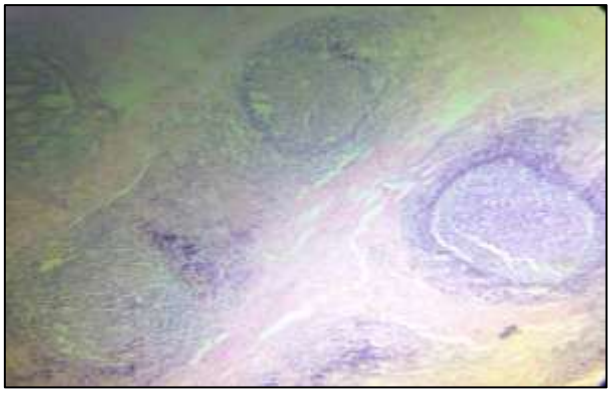

Figure 2: H and E (4X) - Lymphoid follicles with prominent germinal centres showing eosinophilic infiltration.

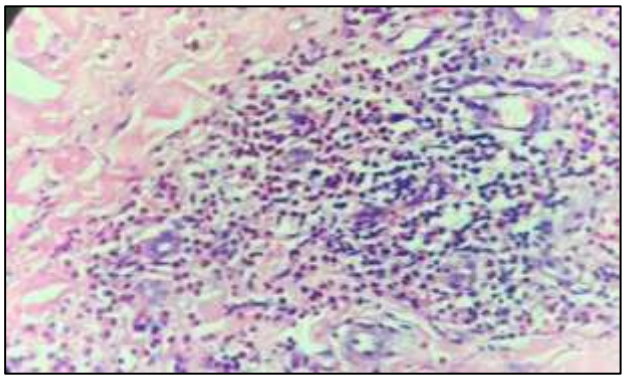

Figure 3: H and E (40X) - Lymphoid follicles with prominent germinal centres showing eosinophilic infiltration.

Hematological investigations revealed leucocytes count of 6000 cells/mm^3 with eosinophilia (6%). Absolute eosinophil count was 360 cells/mm^3. Other hematological investigations like liver function tests and renal function tests and radiological investigations were normal. Fine needle aspiration cytology yielded bloody aspirate which was inconclusive. Histopathology of the completely excised specimen showed fibro-collagenous tissue containing lymphoid follicles with prominent germinal centres showing eosinophilic infiltration and occasional hyalinized vessels suggestive of Kimura's disease (Figure 2 and 3). Final diagnosis of Kimura's disease was reached on the basis of histopathological examination and hyper-eosinophilia. There was no recurrence of lesion during 4 months of follow up.

Case 2

35 years male presented with asymptomatic raised lesion over right iliac region since 6 months. On examination, it was 5×5 cms sized, non-tender exophytic pinkish growth over right iliac region (Figure 4). Systemic examination was normal.

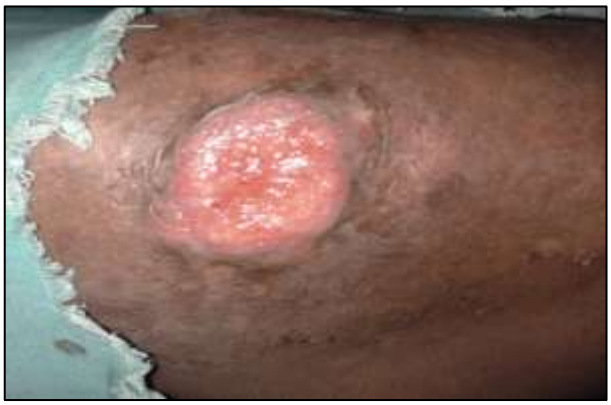

Figure 4: Case 2: 5×5 cms exophytic pinkish mass over right iliac region.

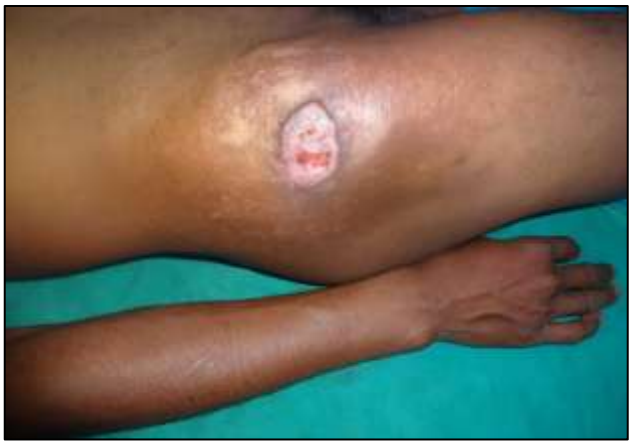

Figure 5: Case 2: 5×5 cms ulcer with granulation tissue over right iliac region.

Pheohyphomycosis, chromoblastomycosis, verrucous carcinoma, amelanotic melanoma, angio-lymphoid hyperplasia with eosinophilia were kept as differentials.

Hematological investigations revealed leucocytes count of 6800 cells/mm^3 with eosinophilia (7%). Absolute eosinophil count was 476 cells/mm^3. Other hematological investigations and radiological investigations were normal. Incisional biopsy was performed for histopathology examination along with special stains for fungal elements. It revealed dense inflammatory infiltrate composed of lymphocytes, plasma cells and many eosinophils in sub-epithelium. At places lymphoid follicles were seen with prominent germinal centre and eosinophils. Vascular proliferation was suggestive of Kimura's disease. Periodic acid-Schiff and Sterheimer-Malbin stains were non- contributory.

Patient was treated with intralesional triamcinolone acetonide, 2 injections 3 weeks apart. Patient showed regression of lesion (Figure 5).

DISCUSSION

Kimura's disease (KD) is an unusual benign condition of uncertain etiology. Described for the first time in China by Kimm and Szeto in 1937, Kimura and colleagues in 1948 in Japan have made a more detailed description. Since then, although 200 cases have been published to date, mostly from the far East.[2] It has been rarely reported from India. Only single case report of Kimura disease of the eyelid was reported from Indian subcontinent.[3]

KD is usually seen in young Asian adults between 20 to 40 years with male predominance (3:1). It involves subcutaneous tissues (preauricular, submandibular), the major salivary glands, and cervical lymph nodes with other rare sites like eyelids, orbit, oral cavity, groin, postauricular area, trunk, and limbs.[4] Both our cases presented with unusual site of involvement leading to diagnostic difficulties.

Various theories for the origin of KD, including impairment or interference with immune regulation, atopic reaction to a persistent antigenic stimulus by arthropod bites, virus and neoplasm have been suggested.[1] In our cases no such etiology could be elicited.

The disease is manifested by an abnormal proliferation of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and eosinophilic inflammatory infiltrate suggest that KD might be a kind of hypersensitivity reaction. Several lines of evidence indicate that lymphocyte T-helper 2 (Th2) might also play a role.[5] Eosinophils counts and serum IgE were elevated in our cases.

KD may affect the kidneys in up to 60% of patients, presenting as all types of glomerulonephritis or as nephritic syndrome (12%) fortunately both our cases lacked any systemic involvement.[4] Diagnosis through FNAC is misleading and can easily be mistaken for a malignant disorder. In presence of unusual clinical features, diagnosis is based on mainly histopathology. As in our cases clinical features were misleading, diagnostic clue was histopathological examination.

T-cell lymphoma, Kaposi Sarcoma, Hodgkin's disease, and angio- lymphoid hyperplasia with eosinophilia (ALHE) are potential differential diagnoses. In contrast to Kimura's disease, in ALHE, numerous thick and thin walled vessels lined with characteristic edematous endothelial cells associated with variable lymphocytic and eosinophilic infiltrate are seen.

In the active phase, the lesion has many cytologic features of malignancy and may be misdiagnosed as angiosarcoma, but it is essentially benign and usually surgical excision results in cure.[6]

In KD conservative approach may be sufficient with the use of other modalities of treatment. Treatment options include surgical excision, radiotherapy, immune-suppressive agents and corticosteroids (systemic and intralesional). Spontaneous remissions may be observed. However, the excision of the mass remains the treatment of choice especially in young, although local recurrences are frequent.

Systemic corticosteroids may be indicated in case of relapse or in patients with renal impairment. Patients should be followed up to monitor renal involvement.

CONCLUSION

Kimura's disease, although difficult to diagnose, with an indolent course and good prognosis should be considered in the differential diagnosis in patients presenting with subcutaneous nodules with hyper-eosinophilia with or without primary lymphadenopathy and should be investigated accordingly.

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