Medical Reports and Case Studies
ISSN: 2572-5130

Azman, Med Rep Case Stud 2017, 2:2
DOI: 10.4172/2572-5130.1000132

Case Report Open Access

# Kimura's Disease: Uncommon Cause of Proptosis

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**Received date:** March 08, 2017, **Accepted date:** March 16, 2017, **Published date:** March 19, 2017



## Abstract

**Purpose:** To describe a case of inferior orbital mass with generalized lymphadenopathy and histopathological examination proven Kimura's disease.

**Case Study and Results:** A 17 year-old Malay man with no previous illness. Presented with two months history of progressive proptosis of the right eye (RE). Upon presentation vision was 6/7.5 RE and left eye (LE) 6/6. Ocular examination revealed RE proptosis with restricted extraocular muscle movement and optic disc swelling. Systemic examination noted multiple lymphadenopathy over cubital fossa and inguinal region. Blood investigation showed peripheral eosinophilia. CT brain and orbit revealed RE inferior orbital mass compressing the optic nerve. Subsequently RE orbital mass incisional biopsy and inguinal node excisional biopsy was done. The later showed typical features of Kimura's disease with diffuse eosinophils infiltration within the germinal centres of the lymphoid nodule. He was started on tapering dose of oral corticosteroids and responded well evidenced by reducing RE proptosis.

**Conclusion:** This case demonstrates that proptosis with lymphadenopathy does not always imply lymphoma but may have a benign pathogenesis such as Orbital Kimura's disease. Therefore it should be considered in the differential diagnosis of orbital lesions occurring in adults. Accurate diagnosis in biopsies is also crucial to avoid unnecessary radical surgery.

**Keywords:** Kimura; Angiolymphoid hyperplasia; Eosinophilic granuloma

## Introduction

Kimura's disease is a chronic inflammatory disease which presenting as a multiple painless solitary subcutaneous nodules characterized by an angiolymphoid proliferation with eosinophilia and elevated serum immunoglobulin IgE. It is a rare entity and found almost exclusively in Asian individuals especially in young males [1]. The disease was first described by Kimm and Szeto in 1937 as 'eosinophilic hyperplastic granuloma' [2]. However, the exact etiology is remain unknown.

## Case Study and Results

A 17 year-old Malay man with no previous illness. Presented to us with two months history of progressive proptosis of the right eye associated with multiple painless nodular swelling over the arm flexor and inguinal region bilaterally. No history of significant trauma prior, hyperthyroidism symptoms nor constitutional symptoms. Upon presentation vision was 6/7.5 right eye and left eye 6/6. RAPD negative. Ocular examination revealed 6mm right eye proptosis with restricted extraocular muscle movement in all direction (Figure 1) and optic disc swelling. Multiple lymphadenopathy over cubital fossa and inguinal region were noted on systemic examination.

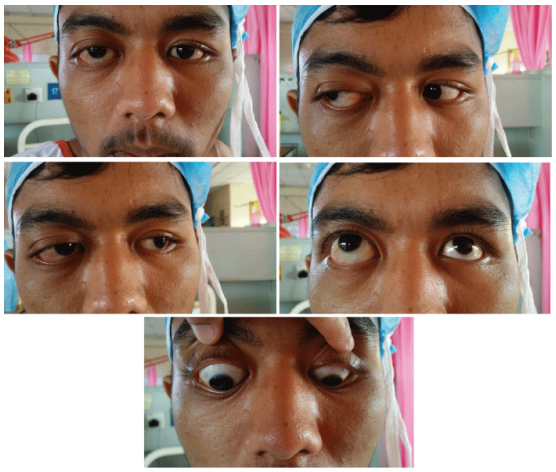

**Figure 1:** Right eye proptosis with limited extraocular muscle movement.

Full blood picture did not reveal any blood dyscrasias except for significant eosinophilia. Urine for microscopic examination performed did not show any evidence of proteinuria nor red blood cells. Thyroid function test was normal. Contrasted computed tomography of the brain and orbit revealed right eye inferior orbital mass with no clear plane of demarcation with inferior rectus muscle extending posteriorly

and compressing the optic nerve (Figure 2). Incisional biopsy of right inferior orbital mass and excisional biopsy of inguinal node was performed. The later showed typical features of Kimura's disease with diffuse eosinophils infiltration within the germinal centers of the lymphoid nodule (Figures 3-6). He was started on tapering dose of oral corticosteroids and responded well evidenced by reducing right eye proptosis.

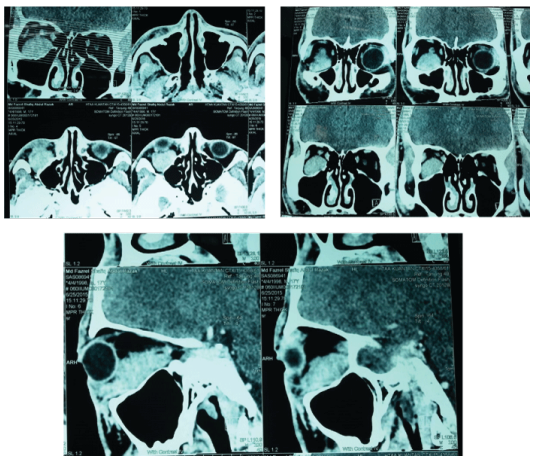

**Figure 2:** CECT scan demonstrate right eye inferior orbital mass.

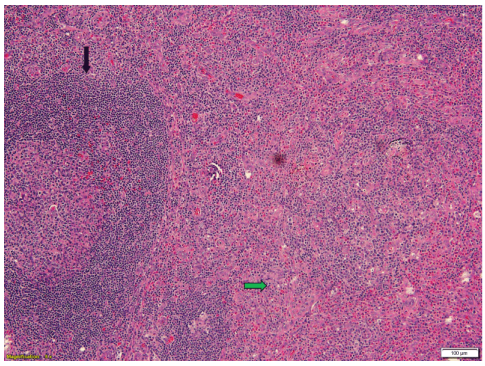

**Figure 3:** The lymph node displaying reactive lymphoid follicles with prominent germinal centers (black arrow). There are numerous eosinophils and plasma cells at the paracortex (green arrow). (Magnification x100).

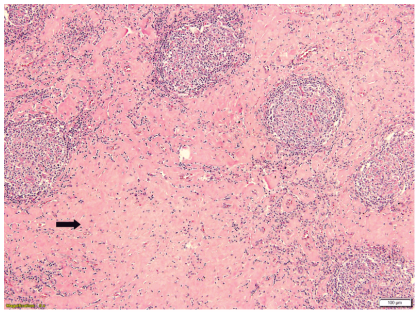

**Figure 4:** There are marked paracortical sclerosis in between the follicles (arrow). (Magnification x40).

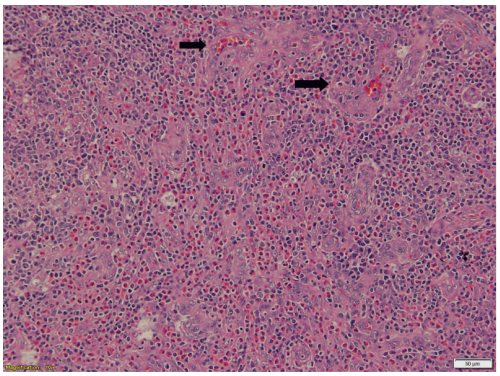

**Figure 5:** There are extensive infiltration of mature eosinophils forming eosinophils microabscesses. The arrows are pointing towards the vascular proliferation at the paracortex. (Magnification x200).

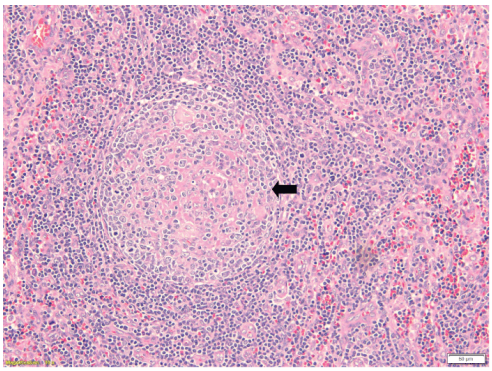

**Figure 6:** The germinal centres are showing interstitial fibrosis and deposition of proteinaceous material (arrow) in between the germinal centre cells. They are rimmed by mature lymphocytes. (Magnification x200).

## Discussion

In 1976 Takenaka et al. reported the first case of orbital Kimura's disease [3]. Kimura's disease has a predilection for the head and neck region but orbital involvement has been reported in only 14 cases [4-7]. Regional lymph node involvement occurs in up to 75% of the cases. Systemic associations includes asthma and nephrotic syndrome [8]. Recurrence is common, estimated between 15 to 40% of the cases but fatalities have not been reported [9]. Most of the patient presented as proptosis and on imaging orbital masses were demonstrated. Nearly all patient with Kimura disease have peripheral blood eosinophilia and elevated serum IgE. Histological findings from biopsy may show lymphoid nodules with discrete germinal centers and marked eosinophilic infiltrate or abscesses. Most of the patient responded to excision of the mass and systemic corticosteroid.

Orbital Kimura's disease may be easily mistaken for a malignant disorder such as orbital lymphoma and Kaposi's sarcoma. Therefore other than clinical findings and imaging, biopsy and histopathological examination play a vital role in managing the patient. In this patient, we manage to get the accurate diagnosis as his histopathological examination showed typical features of Kimura's disease.

## Conclusion

This case demonstrates that proptosis with lymphadenopathy does not always imply lymphoma but may have a benign pathogenesis such as Orbital Kimura's disease. Therefore it should be considered in the differential diagnosis of orbital lesions occurring in adults especially in our population. Accurate diagnosis in biopsies is also crucial to avoid unnecessary radical surgery.

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