INTRODUCTION

Kimura disease is an extremely rare disease. It is characterised by chronic inflammatory disorder involving the subcutaneous tissue at the head and neck region, major salivary gland or lymph nodes with presence of peripheral blood eosinophilia and an elevated IgE concentration. It is of unknown aetiology and most reported cases involve men of Chinese and Japanese origins. It can also occur occasionally in Caucasians but rarely in Africans. Males are more affected than females. The age at presentation described in literature is between the second and third decade of life [1, 2].

CASE PRESENTATION

A 34-year-old Malay lady presented with gradually enlarging right nasal swelling of one-year duration. The swelling was painless and not associated with nasal blockage or nasal discharge. She had no eye symptoms and no associated constitutional symptoms. Physical examination showed a firm swelling over the right part of her nasal dorsum, which was non-tender, not warm and immobile. It measured 2 cm x 2 cm, oval in shape with broad base and well-defined margins. The overlying skin was normal (Figure 1). Nasal endoscopy showed no extension of the mass into the right nasal cavity. Surgical excision was performed where histopathological examination (HPE) reported findings that were consistent with Kimura disease. Kimura disease, a rare benign disease, can affect the head and neck structures. Manifestation at the nasal region is an extremely rare occurrence. HPE is needed to confirm the diagnosis.

ABSTRACT

Kimura disease is a rare chronic inflammatory disorder affecting the subcutaneous tissue. It is of unknown etiology mainly occurring in the head and neck region. A 34-year-old Malay lady presented with a gradually enlarging, painless right nasal bridge mass of one-year duration. Clinical examination revealed a firm, immobile swelling measuring 2 cm x 2 cm. Nasal endoscopy showed no extension of the mass into the right nasal cavity. Surgical excision was performed where histopathological examination (HPE) reported findings that were consistent with Kimura disease. Kimura disease, a rare benign disease, can affect the head and neck structures. Manifestation at the nasal region is an extremely rare occurrence. HPE is needed to confirm the diagnosis.

KEYWORDS: Kimura disease; nose; histopathological examination
**Figure 1** Right nasal dorsum mass with dryness of overlying skin

**Figure 2** Vascular proliferation with lymphoid cells and dense eosinophilic Infiltrates

**Figure 3** Post-operative scar in the nasolabial groove
DISCUSSION

Kimura and Sieto first described Kimura disease in seven patients with benign lymph nodes enlargement [3]. Since there was coexistent eosinophilic infiltration, it was termed as eosinophilic hyperplastic lymphogranuloma [3]. Kimura et al [4] later in 1948 noted a vascular involvement and described it as an unusual granulation with hyperplastic changes of the lymphatic tissue and since then this condition has been widely known as Kimura disease.

Kimura disease often occurs at the head and neck region involving the subcutaneous tissue, major salivary glands and lymph nodes. Other parts of body such as oral cavity, orbit, axilla, groin and spermatic cord, skeletal muscle and prostate can also be affected [5]. Kimura disease is generally a localised process with a benign clinical course.

Middle aged Asian men are most commonly affected. China and Japan are endemic countries but sporadic cases have been described elsewhere [6]. Peak age of onset is in the third decade of life with a marked male preponderance with male to female ratio of 3.5 to 7 [1]. Aetiology of the disease is unknown. Increased production of eosinophilotropic cytokines like IL4 is postulated to be the cause apart from allergic reaction and infection [7, 8].

Kimura disease usually presents as a solitary or multiple deep, slow growing subcutaneous nodule located in the head and neck region accompanied by satellite lymphadenopathy or salivary gland hypertrophy mainly of the parotid and submaxillary glands. Lymph node enlargement can also be the only manifestation of the disease. Peripheral blood eosinophilia and an elevated serum IgE level are consistent laboratory findings in Kimura disease [9].

In diagnosing Kimura disease, the clinical aspect of the disease together with HPE evidence of the disease is mandatory since FNAC can be misleading. There are chronic inflammatory infiltrates of lymphocytes that will form lymphoid follicles, which are often accompanied by vascular proliferation and fibrosis [9, 10]. Immunofluorescence studies show germinal centre containing heavy IgE deposits and variable amount of IgG, IgM and fibrinogen.

Radiological studies for Kimura disease are variable and non-specific. CT scan and MRI are not diagnostic. However, the imaging from these may help to locate the extent of the disease [11]. Contrasted CT scan showing the vascular nature of lesion is not a consistent finding while MRI with contrast enhancement (Gd DTPA), may demonstrate high T1 and T2 weighted signal intensities in the involved tissue.

There are several differential diagnosis of Kimura disease when it is confined to the nose, which include angiolymphoid hyperplasia with eosinophilia (ALHE), tuberculosis, Kaposi sarcoma, eosinophilia granuloma, epitheliod hemangioma and angiofollicular hyperplasia [12]. Differential diagnosis between Kimura disease and ALHE has been a challenge. However, in contrast to ALHE, in Kimura disease the germinal centres are destroyed due to heavy infiltration of eosinophil and there is absence of vacuolated endothelial cells, which are not seen in ALHE.

Various modalities have been suggested to treat Kimura disease. However, no consensus has been reached yet. Conservative treatment includes oral steroids but the lesions usually get enlarged again when steroid treatment is stopped. Therefore, continuation of constant low dose steroid therapy is often considered for a successful outcome [13]. Treatment of choice for localized disease is surgical excision [14]. However there have been reports of recurrence in 15 to 40% of cases even after apparently adequate surgical excision. Thus, surgery and subsequent steroid treatment are proposed as an alternative treatment [15]. Other treatment options include intralesional steroid injection and radiation therapy. Radiation therapy with doses of 20-30 Gy is useful to control lesion that either relapses after surgery or is unresponsive to steroids [15].

CONCLUSION

Kimura disease is a rare benign chronic inflammatory disorder of unknown aetiology, which can occur in the head and neck region. However, involvement of this disease at the nose is extremely rare. It often follows a prolonged, indolent course with no evidence of malignant transformation. Hence the prognosis of Kimura disease is good.
Conflict of Interest
Authors declare none.

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Author’s Contribution
Drafting the manuscript: ZM, ARI
Data collection: ZM, ARI
Data analysis: ZM, ARI, IM
Final approval: ZM, IM

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