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Case Report

## Kimura's disease of the parapharyngeal space: A clinical case study

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### Abstract

Kimura's disease (KD) is a rare idiopathic condition of unknown etiology. The disease is characterized by swelling and lesions in the head and neck region, with involvement of the subcutaneous soft tissue, major salivary glands and lymph nodes. Patients almost always have eosinophilia and elevated serum immunoglobulin E levels. The diagnosis is established by biopsy. KD is usually self-limiting. We report a case of Kimura's disease in a female and review the literature. The present report highlights the need for increased awareness by all otolaryngologist of this clinical impressive entity.

**Keywords:** Kimura's disease; eosinophilia; Parapharyngeal mass; immunoglobulin E

### Introduction

Kimura disease (KD) is a chronic inflammatory disorder of unknown etiology, most commonly manifesting as painless unilateral cervical lymphadenopathy or subcutaneous masses in the head or neck region<sup>1-3</sup>. The first report of KD was from China in 1937, in which Kimm and Szeto described 7 cases of a condition they termed "eosinophilic hyperplastic lymphogranuloma." The disorder received its current name in 1948, when Kimura et al. noted the vascular component and referred to it as an "unusual granulation combined with hyperplastic changes in lymphoid tissue."

Controversy exists in the literature regarding whether KD and angiolymphoid hyperplasia with eosinophilia (ALHE) are the same entity<sup>4-6</sup>. Some authors believe that KD represents a chronic deeper form of ALHE; however, most recent papers distinguish the two on the basis of clinical and histopathologic characteristics, which are detailed below. Kimura's disease is a chronic inflammatory condition which presents with a characteristic triad of signs and

symptoms, namely a painless, slowly enlarging soft tissue mass (or masses), associated lymphadenopathy and peripheral eosinophilia<sup>7-9</sup>. Eighty-five per cent of cases occur in men<sup>10-11</sup>.

We report a case of Kimura's disease in a female and review the literature. The present report highlights the need for increased awareness by all otolaryngologist of this clinically impressive entity.

### Case Report

A 32-year-old woman presented with a complaint of a slowly enlarging mass in the right parapharyngeal space. History revealed that he had this swelling for six months and was treated with antibiotics following which it used to subside but again appeared within 15 days of stopping antibiotic. She reported no epistaxis, nasal obstruction, dysphagia, hemoptysis, or other symptoms. There was no history of pain, fever, night sweats, or weight loss. She was a nonsmoker, and the rest of her medical history was unremarkable. On physical examination, he seemed well. A nontender, nonfluctuant

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firm mass measuring 6 x 6 x 4 cm was palpable in the left anterior triangle of the neck, extending from the level of the right nasopharynx to the aryepiglottic fold inferiorly. No warmth or redness was noted on the overlying skin. Neither axillary or inguinal lymphadenopathy nor hepatosplenomegaly was noted. The rest of the physical examination was normal. Nasal endoscopy showed that the mass protruded into the right nasopharynx and oropharynx. The overlying mucosa was intact and appeared to be normal. Laboratory data included a hemoglobin of 13.1g/dL, platelet count of  $387 \times 10^9/L$ , and white cell count of  $10.8 \times 10^9/L$ : differential showed 26% eosinophils ( $2.81 \times 10^9/L$ ), 46% segmented neutrophils, 22% lymphocytes, and 6% monocytes. The erythrocyte sedimentation rate was 11 mm/hour. Results of serum electrolytes, liver function tests, albumin, blood urea nitrogen, and creatinine were normal. A chest radiograph and skeletal survey were unremarkable.

Computed tomography (CT) detected a mass in the parapharyngeal space with increased contrast uptake (Fig 2). Findings on all other investigations--namely, urea and electrolyte measurements, liver function tests, Epstein-Barr virus serology, and chest radiographs were normal.

Fine needle aspiration cytology revealed inflammatory cells and differential count revealed marked eosinophilia. As the patient was not having any local infection clinical diagnosis was not reached. Hence excision biopsy was performed which revealed a dense inflammatory infiltrate and fibrosis (Figures 1 and 3). The infiltrate was characterized by lymphoid tissues with germinal centers and numerous eosinophils with microabscess formation. A prominent proliferation of small venule-sized vessels was noted. There was no evidence of malignancy and no organisms were noted.



Fig1. Computed tomography (CT) detected a mass in the parapharyngeal space (lateral view)

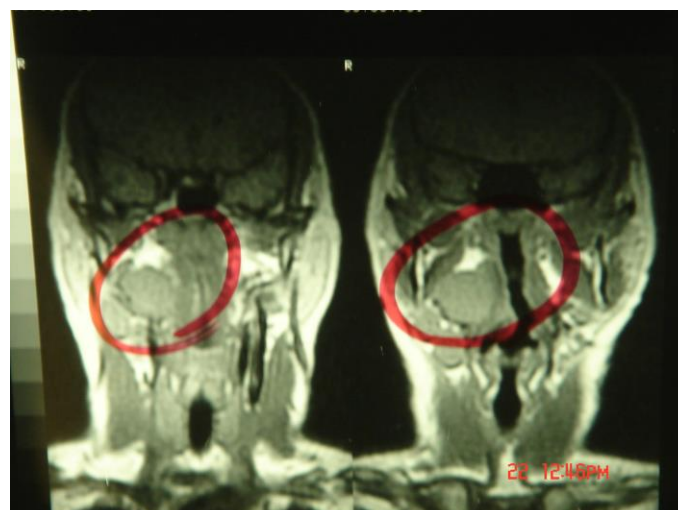
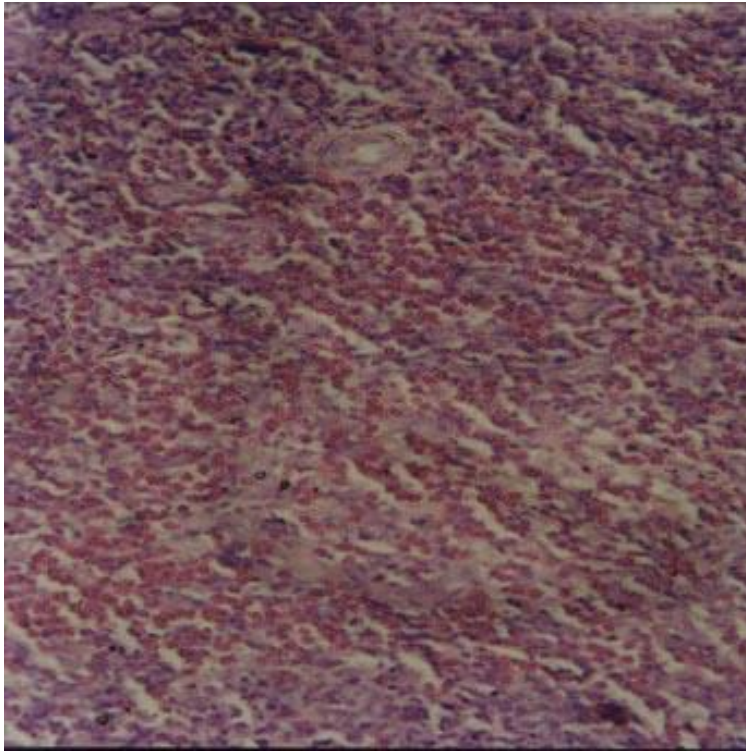


Fig2. Computed tomography (CT) detected a mass in the parapharyngeal space (coronal view)



**Fig3. Photomicrograph of the mass shows dense infiltrate of lymphocytes with germinal center formation. There are foci of eosinophilic infiltrate forming eosinophilic microabscesses.**

## Discussion

Kimura's disease is an important category of reactive lymphadenopathy in the Oriental population. The enlarged nodes are mostly located in the head and neck region. Salient pathological changes include florid germinal centers, Warthin-Finkeldey type polykaryocytes, and vascularization of germinal centers, increased postcapillary venules in the paracortex, eosinophilic infiltration, and sclerosis. The pathophysiology of KD remains unknown, although an allergic reaction, trauma, and an autoimmune process have all been implicated as the possible cause. The disease is manifested by an abnormal proliferation of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate suggest that KD may be a hypersensitivity reaction. Some evidence indicates that TH2 lymphocytes may also play a role, but further investigation is needed.

KD is generally limited to the skin, lymph nodes, and salivary glands, but patients with KD and nephrotic syndrome have been reported. The basis of this association is unclear.

The pathology of Kimura's disease is quite different from that of angiolymphoid hyperplasia with eosinophilia (epithelioid hemangioma). Immunoperoxidase studies

show IgE reticular networks in germinal centers. Nondegranulated surface IgE-positive mast cells are present in the paracortex. The authors propose that Kimura's disease represents an aberrant immune reaction to an as yet unknown stimulus. Although the individual histological features are nonspecific, the constellation of features is highly characteristic of Kimura's disease. Since lymphadenopathy can herald involvement of other tissues and the prognosis is excellent, accurate diagnosis of this disease in lymph node biopsies may spare the patients unnecessary radical surgery.

The treatment of Kimura's disease is not well established. Surgery, corticosteroid therapy, and radiotherapy have all been used. Surgery has a crucial role in that it can establish the diagnosis while removing large compressive or cosmetically unacceptable lesions. Most authors believe that the recurrence rate is fairly high with surgical excision alone<sup>12</sup>. Corticosteroid therapies usually produce good results initially, but recurrence is common after discontinuation of therapy<sup>13</sup>. Radiation has been used mainly to treat recalcitrant or large lesions<sup>14</sup>. Two other unestablished treatment modalities are cryotherapy and laser fulguration; the results of both are variable and very unpredictable. While recurrences are common, no case of metastasis or malignant degeneration has ever been reported<sup>6</sup>.

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