Kimura Disease: A Rare Case in Vietnamese Woman

Linh Nguyet Le (✉ lenguyetlinh210@gmail.com )
University of Medicine and Pharmacy Ho Chi Minh City https://orcid.org/0009-0000-6693-834X

Linh Ngoc Tuong Tran
Duy Le Pham
https://orcid.org/0000-0001-5382-9283

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Abstract

Background: Kimura disease (KD) is a rare benign chronic inflammatory condition that predominantly affects Asian males. It is characterized by subcutaneous tissue masses in the head and neck region, enlarged lymph nodes, increased blood eosinophilia, and elevated serum total IgE levels. In this report, we describe a rare case of KD in a young Vietnamese female.

Case summary: A 31-year-old Vietnamese woman presented to the hospital with 2 masses in the bilateral cheeks and 1 mass behind the left ear that persisted for 15 years, recurrent skin itching, elevated serum total IgE levels and increased blood eosinophilia. No medical history of the individual or family was recorded. We performed an excision biopsy of the postauricular mass which revealed follicular hyperplasia with small vessel hyperplasia, diffuse infiltration of eosinophils in lymphoid follicles, and several eosinophilic microabscesses. After a comprehensive review, the final diagnosis for this patient was KD and atopic dermatitis comorbidity.

Conclusion: KD is not limited to males, as this report demonstrated. The histopathological examination plays an important role in the diagnosis of KD. This case illustrated the characteristic description of KD and highlights the need for awareness of this rare disease in Asian women.

Introduction

Kimura disease (KD) is a rare condition that was first described as an “eosinophilic hyperplastic granuloma” by Kim and Szetu in 1937 [1]. The popular name became “Kimura disease” when the condition was described by Kimura in the Japanese literature in 1948 [2]. KD is a benign disease, characterized by subcutaneous tissue masses in the head and neck region, enlarged lymph nodes, blood eosinophilia, and an increased serum level of IgE. Diagnosis is challenging; an excisional biopsy is necessary for confirmation. The disease is largely confined to Asian populations; several cases have been reported from Japan, China, and Indonesia. KD is more common in men than women (male:female ratio 3:1) [3]. Here, we describe a rare case of KD in a young Vietnamese female.

Case report

A 31-year-old Vietnamese woman presented to the Unit of Allergy and Clinical Immunology, University Medical Center of Ho Chi Minh City, Vietnam, in October 2022, complaining of masses in both cheeks and a mass behind the left ear. The masses had developed 15 years prior and had gradually increased in size. She also complained of itchy, inflammatory skin lesions of the legs and arms. The patient had sought care from 2018 and had undergone several blood and imaging tests; the results are shown in Table 1. However, no diagnosis had been made. On examination, we found non-tender diffuse masses in both buccal regions and a tender well-circumscribed mass in the left posterior auricular region (the latter 2.5 × 3.0 cm in dimensions) (Fig. 1). Eczematous lesions were apparent on both legs and both arms (Fig. 2). No other finding was remarkable.
<table>
<thead>
<tr>
<th>Subclinical results</th>
<th>Time</th>
<th>Value</th>
<th>Reference value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Complete blood count</strong></td>
<td><strong>Jan, 2018</strong></td>
<td><strong>Total leucocyte count</strong></td>
<td><strong>4000–10.000/mm³</strong></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neutrophil</td>
<td><strong>2000–7000/mm³</strong></td>
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<tr>
<td></td>
<td></td>
<td>Lymphocyte</td>
<td><strong>1000–3000/mm³</strong></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Eosinophil</td>
<td><strong>20–500/mm³</strong></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Basophil</td>
<td><strong>20–100/mm³</strong></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Red blood cell count</td>
<td><strong>4.200.000–5.400.000/mm³</strong></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Plateled count</td>
<td><strong>150.000–450.000/mm³</strong></td>
</tr>
<tr>
<td><strong>IgE test</strong></td>
<td><strong>Oct, 2022</strong></td>
<td>Total serum IgE level</td>
<td><strong>&lt; 100 IU/mL</strong></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Specific IgE (RAST)</td>
<td>Positive with <em>Dermatophagoides pteronyssinus</em> (1.0); <em>D. farinae</em> (1.2); <em>Blomia tropicalis</em> (1.5)</td>
</tr>
</tbody>
</table>
A complete blood count revealed a white blood cell count of $8.89 \times 10^9/L$ and an elevated eosinophil count ($3.7 \times 10^9/L$). The other blood cell counts were within the normal ranges (neutrophils $2.3 \times 10^9/L$, lymphocytes $2.3 \times 10^9/L$, basophils $0.039 \times 10^9/L$, red blood cells $4,170 \times 10^9/L$, platelets $262 \times 10^9/L$). The level of serum total IgE was high (15,403 IU/mL).

Ultrasound revealed oval, heterogeneous, diffuse hypoechoic masses in both buccal regions, of dimensions $8 \times 15$ and $13 \times 20$ mm in the left and right cheeks, respectively. In addition, poorly echogenic oval structures were observed: hilar lymph nodes (grade +) and inflammatory nodules $\leq 10 \times 24$ mm in size at the angle of the jaw and along both sides of the sternocleidomastoid muscle, and behind the left ear (the latter mass was $\leq 9 \times 23$ mm in dimensions) (Fig. 3).

We performed fine-needle aspiration (FNA) of the left postauricular mass; this revealed a polymorphous cell population of lymphocytes and eosinophils. The FNA data did not aid diagnosis. An excision biopsy
of the postauricular mass revealed follicular hyperplasia, small vessel hyperplasia, diffuse infiltration of eosinophils into lymphoid follicles, and several eosinophilic microabscesses (Fig. 4). We suspected kidney injury, but renal examinations revealed no renal involvement. A diagnosis of KD with comorbid atopic dermatitis was confirmed.

**Discussion**

KD is a rare, benign, chronic inflammatory disorder of (principally) young Asian males; the male:female ratio is 3:1 [3–5]. However, cases have been reported worldwide at ages of 1 to 66 years [6]. KD is characterized by painless, firm, diffuse, single, or multiple subcutaneous masses 1 to 7 cm in diameter, principally in the head and/or neck (76%), particularly around the parotid gland and in the submandibular region. Although extremely rare, lesions have also been reported in the axillary and inguinal regions, the trunk, abdomen, chest wall, peripheral extremities, epiglottis, long bones, breasts, genitals, orbits, and ocular appendages [7, 8]. The overlying skin is usually not significantly affected. Sometimes, skin itching, pigmentation, thickening, local erosion, or even ulceration may be evident. Associated regional lymphadenopathy and salivary gland enlargement are common [9]. Systemic associations include nephrotic syndrome, eczema, asthma, sinusitis, tuberculosis, and Loeffler syndrome [10–12]. Typically, the kidneys and skin are affected; the incidence of renal pathology ranges from 10–60% [7, 13, 14].

In terms of laboratory findings, an elevated blood eosinophil count and a high level of serum total IgE are the most prominent features of KD. These parameters are crucial in terms of diagnosis, treatment, and prognosis. Imaging findings such as those from ultrasound (US), computed tomography (CT), and magnetic resonance (MR) are nonspecific but reveal the lesional morphologies and the anatomical distributions [8].

A definitive KD diagnosis requires histopathological examination. The prominent histopathological characteristics include follicular hyperplasia with active germinal centers and small vessel hyperplasia. The diffuse interfollicular infiltrates are rich in eosinophils, lymphocytes, plasma cells, and mast cells. Sometimes, several eosinophilic microabscesses and fibrosis are observed [9, 15]. Histologically, lymphocytic eosinophilic hyperplasia (ALHE) is similar to KD and commonly affects women in the third to fourth decade of life. However, it is characterized by vascular proliferation with many large epithelioid or histiocytoid endothelial cells; eosinophilic infiltration is rare. ALHE lesions are smaller than those of KD, more numerous, more superficial, more erythematous, and more likely to bleed when irritated. ALHE is rarely associated with systemic disease, the lymph nodes, or the salivary glands. Table 2 summarizes the unique features of KD and ALHE [16]. Based on the histopathological and clinical findings, a diagnosis of ALHE could be excluded in our present case. Other KD differential diagnoses include Hodgkin and non-Hodgkin lymphoma, allergic granulomatosis, Kikuchi disease, and Mikulicz disease [17].
Table 2
Comparison of Angiolymphoid Hyperplasia with Eosinophilia and Kimura Disease

<table>
<thead>
<tr>
<th></th>
<th>ALHE</th>
<th>KD</th>
</tr>
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<tbody>
<tr>
<td>Gender</td>
<td>Typically middle-aged females</td>
<td>Predominantly young adult Asian males</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Pruritus, pain, pulsation</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>Lesion type and location</td>
<td>Small and superficial, with overlying erythema; head and neck region</td>
<td>Large, mainly subcutaneous; overlying skin normal; head and neck region; may involve regional lymph nodes and salivary glands</td>
</tr>
<tr>
<td>Lymphoid follicles</td>
<td>Uncommon</td>
<td>Prominent lymphoid follicles with germinal centers</td>
</tr>
<tr>
<td>Vascular proliferation</td>
<td>Prominent vascular proliferation with large epithelioid/ histiocytoid endothelial cells; evidence of underlying vascular malformation may be evident</td>
<td>Some stromal vascularity with unremarkable endothelial cells</td>
</tr>
<tr>
<td>Fibrosis</td>
<td>Absent or limited</td>
<td>Prominent</td>
</tr>
<tr>
<td>Serum IgE level</td>
<td>Normal</td>
<td>Increased</td>
</tr>
<tr>
<td>Nephropathy</td>
<td>Absent</td>
<td>Present in up to 20% of patients</td>
</tr>
</tbody>
</table>

Fitzpatrick's Dermatology 9th Edition [16]

The etiology of KD is unclear. The condition may be an allergic reaction caused by infection with *Candida albicans*, parasites, or viruses, or arthropod bites, or may be due to dysregulation of T cell responses in patients with endocrine disorders and/or autoimmune diseases. It could also stem from a Th2 immune response triggering deposition of eosinophils in diseased tissue [18][19].

There is currently no standard KD treatment [9]. The primary treatments include surgical excision, systemic steroids, cytotoxic drugs, radiation therapy, and chemotherapy [18]. Intralesional injection of corticosteroids has afforded good results. The disease recurrence rate attains 40% [17]. KD is benign and self-limited; malignant transformation has not been recorded. However, several complications of KD have been reported, including cerebral artery, jugular vein, pulmonary, mesenteric, and multiple arterial embolisms in the extremities [8, 18].

The diagnosis of KD in our patient was based on the typical epidemiological features (age and race), the medical history, clinical features, blood tests, and histological findings. This case was accompanied by eczematous skin lesions but no renal dysfunction was detected.

**Conclusion**
KD is a rare, benign, chronic inflammatory condition characterized by single or multiple subcutaneous tissue masses in the head and neck region, enlarged lymph nodes, increased blood eosinophilia, and elevated serum levels of IgE. Diagnosis is primarily based on histopathological examination. The principal management is surgical excision. Although the prognosis is good, the recurrence rate is high.

**Declarations**

Author contribution All authors contributed to the study conception and design. Case preparation was performed by LNT. Tran and DL. Pham. The first draft of the manuscript was written by LN. Le and all authors commented on previous versions of the manuscript. LN. Le was involved in the correction of the manuscript after review process. DL. Pham supervised all the process. All authors read and approved the final manuscript.

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**Data availability** The original clinical datasets generated during the case are available from the corresponding author on reasonable request.

**Ethics approval** The Local Ethics Committee approval is not required because patient signed informed consent and no study was performed.

**Consent to participate** Written informed consent was obtained from the patient.

**Consent for publication** The authors affirm that the patient provided informed consent for publication of the data and the images in Fig. 1A, B, 2, 3A, B, C, D, 4A, B and 5.

**Competing interests** The authors declare no competing interests.

**References**


**Figures**
Figure 1

Masses in both cheeks and behind the left ear
Figure 2

Eczematous lesions (erythema, vesicles) on the right lower leg
Figure 3

Ultrasonography: Ultrasound revealed (A) a mass in the left cheek; (B) a mass in the right cheek; (C) swollen lymph nodes along both sides of the sternocleidomastoid muscle; and (D) a left postauricular mass.

Figure 4

Histopathological examination of the left postauricular mass: A, Follicular hyperplasia with small vessel hyperplasia and diffuse infiltration of eosinophils into lymphoid follicles (H&E staining × 100); B, Eosinophilic microabscesses (H&E staining × 400)

Supplementary Files
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