Case Report

Asia Pacific Allergy

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Kimura disease: A rare case in Vietnamese woman

Linh Nguyet Le[1], Linh Ngoc Tuong Tran[2], and Duy Le Pham[3,4,*]

ABSTRACT

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. 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 that 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. In 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.

Keywords: Angiolymphoid hyperplasia with eosinophilia; eosinophilia; IgE; Kimura; Kimura disease; subcutaneous masses

1. 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.

2. 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 nontender 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.

A complete blood count revealed a white blood cell count of 8.89×10^9/L and an elevated eosinophil count (3.7×10^9/L). The other blood cell counts were within the normal ranges (neutrophils, 2.3×10^9/L; lymphocytes, 2.3×10^9/L; basophils, 0.039×10^9/L; red blood cells, $4{,}170 \times 10^3$/L; platelets, 262×10^3/L). The level of serum total IgE was high (15,403 IU/mL).

Ultrasound (US) revealed oval, heterogeneous, diffuse hypoechoic masses in both buccal regions, of dimensions 8 × 15 and 13 × 20 mm in the left and right cheeks, respectively. In addition, poorly echogenic oval structures were observed: hilar lymph nodes (grade +) and inflammatory nodules ≤10 × 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 ≤9 × 23 mm in dimensions) (Fig. 3).

We performed fine-needle aspiration (FNA) of the left postauricular mass; this revealed a polymorphous cell population of

[1]Department of Dermatology, University of Medicine and Pharmacy at Ho Chi Minh City, Ho Chi Minh City, Vietnam, [2]Department of Otorhinolaryngology, University Medical Center Ho Chi Minh City, Ho Chi Minh City, Vietnam, [3]Department of Physiology, Pathophysiology & Immunology, University of Medicine and Pharmacy at Ho Chi Minh City, Ho Chi Minh City, Vietnam, [4]Allergy & Clinical Immunology Unit, University Medical Center Ho Chi Minh City, Ho Chi Minh City, Vietnam

*Correspondence to Duy Le Pham, Department of Physiology, Pathophysiology & Immunology, University of Medicine and Pharmacy at Ho Chi Minh City, Ho Chi Minh City, 700000, Vietnam; Allergy & Clinical Immunology Unit, University Medical Center Ho Chi Minh City, Ho Chi Minh City, 700000, Vietnam, Email: drduypham@ump.edu.vn; duy.pl@umc.edu.vn

Tel: +84-969-965-278

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

The local ethics committee approval is not required because patient signed informed consent and no study was performed. Written informed consent was obtained from the patient. The authors affirm that the patient provided informed consent for publication of the data and the images in Figs. 1A, B, 2, 3A, B, C, D, 4A, B, and S1.

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Table 1.

Summary table of subclinical results

Subclinical results	Time		Value	Reference value
Complete blood count	January 2018	Total leucocyte count	1,244/mm^3	4,000–10,000/mm^3
		Neutrophil	7,290/mm^3	2,000–7,000/mm^3
		Lymphocyte	1,570/mm^3	1,000–3,000/mm^3
		Eosinophil	2,630/mm^3	20–500/mm^3
		Basophil	6/mm^3	20–100/mm^3
		Red blood cell count	4,090,000/mm^3	4,200,000–5,400,000/mm^3
		Platelet count	226,000/mm^3	150,000–450,000/mm^3
	October 2022	Total leucocyte count	8,890/mm^3	4,000–10,000/mm^3
		Neutrophil	2,347/mm^3	2,000–7,000/mm^3
		Lymphocyte	2,364/mm^3	1,000–3,000/mm^3
		Eosinophil	3,716/mm^3	20–500/mm^3
		Basophil	39/mm^3	20–100/mm^3
		Red blood cell count	4,170,000/mm^3	4,200,000–5,400,000/mm^3
		Platelet count	262,000/mm^3	150,000–450,000/mm^3
IgE test	October 2022	Total serum IgE level	15,403 IU/mL	<100 IU/mL
		Specific IgE (RAST)	Positive with *Dermatophagoides pteronyssinus* (1.0); *Dermatophagoides farinae* (1.2); *Blomia tropicalis* (1.5)	
Ultrasound imaging (Fig. 3)	January 2018	(1) Hypoechoic, oval, heterogeneous diffuse masses in both buccal regions. The mass in the left cheek measured 8 × 15 mm and that in the right cheek 13 × 20 mm.		
		(2) Some poorly echogenic oval structures were observed, as were hilar lymph nodes (grade +) and inflammatory nodules ≤ 10 × 24 mm in size at the angle of the jaw and along both sides of the sternocleidomastoid muscle.		
		(3) Some poorly echogenic oval structures were observed, as were hilar lymph nodes (grade +) and inflammatory nodules ≤ 9 × 23 mm in dimensions behind the left ear.		
Histopathological examination	January 2018	FNA revealed the histopathology of a neck lymph node. Imaging revealed acute soft tissue inflammation.		
	October 2022 (Fig. 4)	Hematoxylin and eosin staining of sections of the left postauricular mass biopsy sample. Imaging revealed follicular hyperplasia with small vessel hyperplasia, diffuse infiltration of eosinophils into lymphoid follicles, and several eosinophilic microabscesses.		
MRI contrast (Supplementary Figure S1; http://links.lww.com/PA9/A21)	January 2018	(1) Plaques associated with abnormal signals were evident in the subcutaneous tissues of both cheeks; the plaques were 25 × 35 × 46 mm in dimensions on the right and 30 × 34 × 50 mm on the left, with small, internal zigzag flow-voids. Contrast enhancement was heterogeneous. No muscle damage was observed.		
		(2) Multiple lymph nodes less than 22 mm in diameter were found under the chin, along the posterior neck, and along the bilateral carotid bundle. The nodules were inflammatory and evidenced homogeneous contrast enhancement.		
		(3) Two lymph nodes of diameter 16 mm were found behind the lateral ear (L). The contrast was not entirely homogenous; the nodes were surrounded by fatty tissue. It was necessary to monitor these nodes in terms of abscesses.		

FNA, fine-needle aspiration; MRI, magnetic resonance imaging.

Figure 1. Masses in both cheeks and behind the left ear.

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.

3. 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% to 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 US, computed tomography, and magnetic resonance 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.

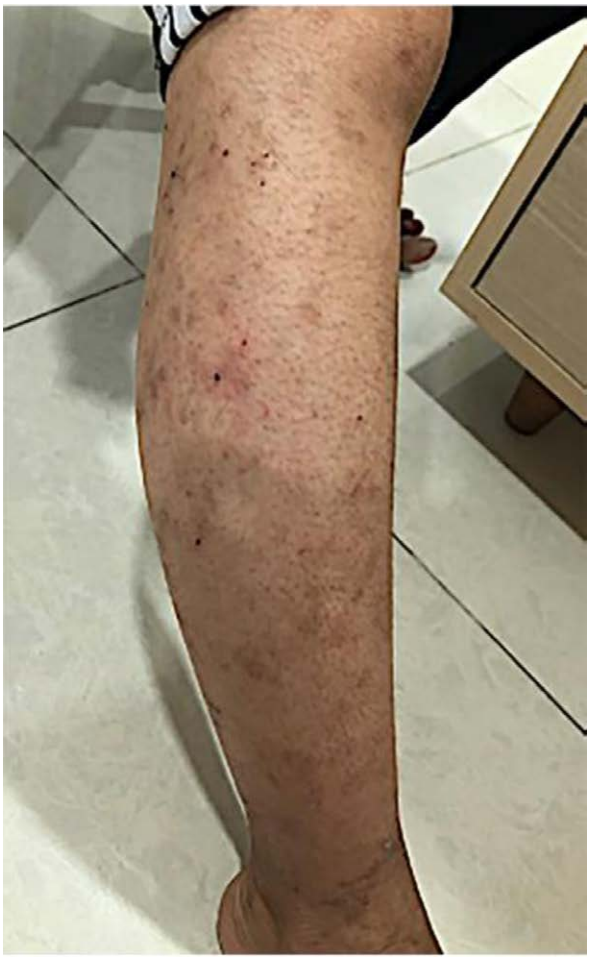

Figure 2. Eczematous lesions (erythema, vesicles) on the right lower leg.

Sometimes, several eosinophilic microabscesses and fibrosis are observed [9, 15]. Histologically, angiolymphoid hyperplasia with eosinophilia (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].

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.

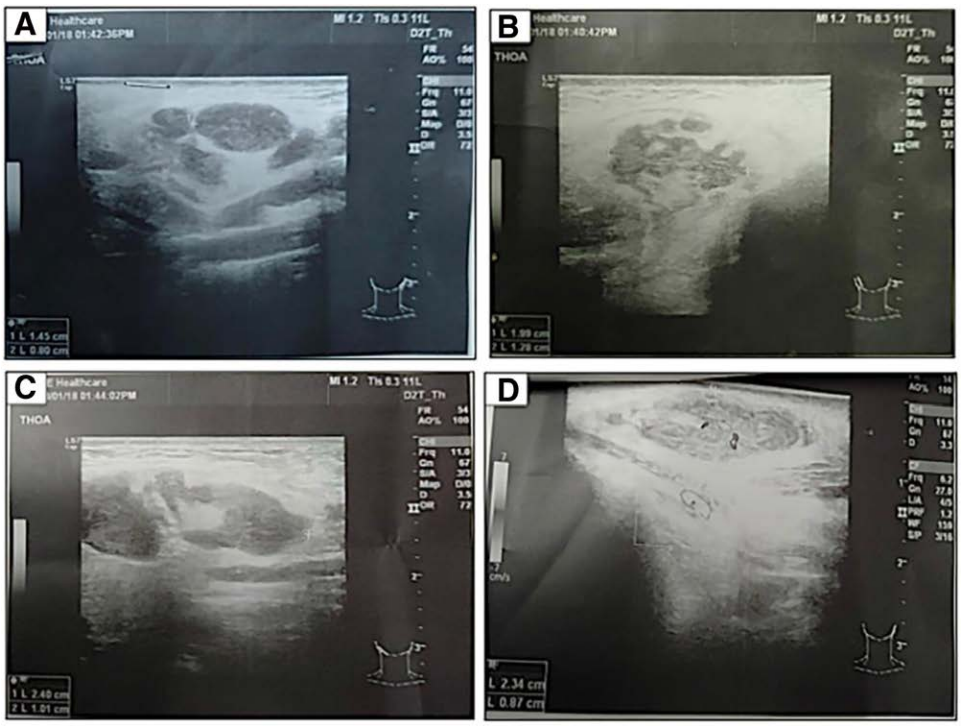

Figure 3. Ultrasonography: ultrasound revealed (A) a mass in the left cheek; (B) a mass in the right cheek; (C) swollen lymph nodes along both sides of the sternocleidomastoid muscle; and (D) a left postauricular mass.

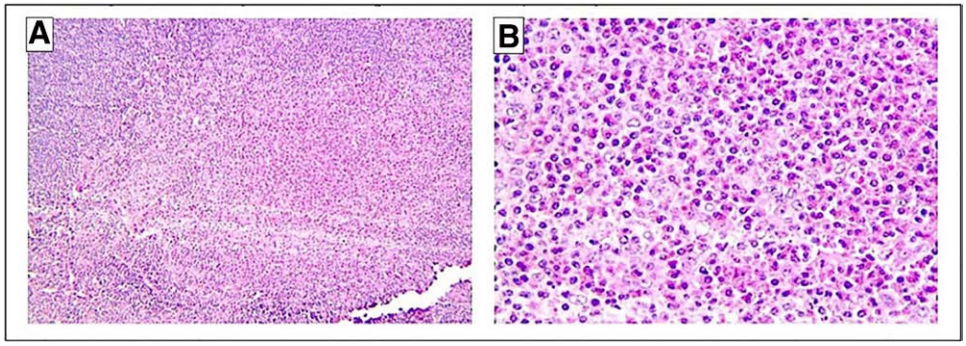

Figure 4. Histopathological examination of the left postauricular mass: (A) Follicular hyperplasia with small vessel hyperplasia and diffuse infiltration of eosinophils into lymphoid follicles (H&E staining ×100); (B) Eosinophilic microabscesses (H&E staining ×400). H&E, hematoxylin and eosin.

Table 2.

Comparison of angiolymphoid hyperplasia with eosinophilia and Kimura disease [3, 6, 7, 9, 10, 16]

	Kimura disease	Angiolymphoid hyperplasia with eosinophilia
Clinical features		
Age	Young (20–30 years)	Middle age (30–50 years)
Gender	Male > female	Female > male
Race	Prominent in Asians	Any races
Symptom	Asymptomatic	Pruritus, pain
Lesion		
Number	Single or multiple	Usually multiple
Size	Larger (up to 20 cm in diameter)	Smaller (average of 1 cm in diameter)
Overlying skin	Usually normal	Usually erythematous skin
Location	Subcutaneous	More superficial
Lymphadenopathy	Frequent	Rare
Laboratory findings		
Eosinophilia	Common	Uncommon
Elevated serum IgE level	Common	Rare
Histopathological		
Location	Subcutaneous, muscle	Dermis, subcutaneous
Lymphoid follicles	Prominent (with germinal centers)	Uncommon
Vascular proliferation	Unremarkable	Prominent
Fibrosis	Prominent	Absent or limited
Complications	Allergic skin disease, nephropathy (present in up to 20%)	Rare

4. Conclusions

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.

Conflicts of interest

The authors declare no conflicts of interest.

Author contribution

All authors contributed to the study conception and design. Case preparation was performed by Linh Ngoc Tuong Tran and Duy Le Pham. The first draft of the manuscript was written by Linh Nguyet Le and all authors commented on previous versions of the manuscript. Linh Nguyet Le was involved in the correction of the manuscript after review process. Duy Le Pham supervised all the process. All authors read and approved the final manuscript.

Supplementary material

Supplementary Figure S1 can be found via 10.5415/apallergy.2022.12.e38.
Supplementary Figure S1
Click here to view

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