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Case Report

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Title Page

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ABSTRACT

Kimura disease is a rare chronic inflammatory disorder characterized by painless head and neck masses, peripheral eosinophilia and elevated serum immunoglobulin E levels, with a histopathological finding of eosinophilic microabscesses, follicular hyperplasia and stromal fibrosis.

The diagnostic challenge arises from frequent misdiagnoses such as autoimmune or malignant conditions which delay appropriate management.

It is rarely encountered in the Middle East, as to the best of our knowledge, we present the second documented case of Kimura disease in the United Arab Emirates (UAE), highlighting it prolongs diagnostic complexities and clinical management.

KEYWORDS

Case report, parotid swelling, systemic lupus erythematosus, lymphoproliferative disorder, diagnostic delay, parotidectomy, eosinophilia, immunoglobulin E, histopathological diagnosis.

BACKGROUND

Kimura's disease is a rare, chronic inflammatory disorder characterized by painless subcutaneous masses, peripheral blood eosinophilia, and markedly elevated serum immunoglobulin E (IgE) levels [1]. While the disease frequently affects young men of East Asian descent, its occurrence in non-Asian populations remains exceedingly rare [5]. Because of its characteristic clinical

presentation, it frequently mimics malignant disease, including lymphoma and salivary gland tumors, often leading to initial misdiagnoses by clinicians worldwide [6, 9,14].

This case illustrates a prolonged diagnostic journey spanning a decade, during which the patient underwent multiple investigations and received several alternative diagnoses with chronic steroid intake before definitive histopathological confirmation of Kimura disease was obtained. After which the patient started tapering steroid and steroid-sparing agent. The patient achieved sustained clinical remission without evidence of recurrence.

The extensive delays in diagnosis, prolonged systemic mismanagement and unnecessary surgical interventions are frequently observed and contribute to multiple variables, such as its rarity, distinct ethnic and geographic distribution, clinical overlap with other disorders, and the lack of clinical familiarity outside its endemic regions [2,7,8,12].

This report aims to provide a clear clinical message: clinicians in non-endemic Gulf settings must maintain a high index of suspicion for Kimura disease when evaluating atypical head and neck masses accompanied by eosinophilia, thereby reducing diagnostic delays and optimizing targeted therapeutic interventions [1,7].

Case presentation

A previously healthy Asian male in his late 20s presented to our unit with a nine-year history of a recurrent, progressive, painless enlargement of the left parotid gland. The mass had gradually increased in size over time, notably without associated fever, constitutional symptoms, skin rash, or facial nerve weakness. Medical, social, and family history were otherwise unremarkable. Initial histopathological and radiological assessment in his home country suggested obstructive

or inflammatory parotid gland disease. While surgical intervention was recommended at that stage, the patient and his family opted for conservative management.

Two years later, the persistence of the mass alongside elevated systemic inflammatory markers prompted a rheumatological evaluation. A diagnosis of systemic lupus erythematosus was made based on these chronic inflammatory manifestations. Treatment with oral prednisolone tablets and hydroxychloroquine tablets was initiated, which the patient continued for approximately one year.

Following the patient's relocation to the Middle East and due to an inability to confirm systemic lupus erythematosus upon subsequent reviews, these immunomodulatory medications were discontinued. During this period, the patient developed chronic headaches, which neurology teams suspected were secondary to the localized chronic inflammation surrounding the enlarging parotid lesion.

Continued progression of the mass eventually prompted another ultrasound-guided biopsy demonstrated lymphoid hyperplasia as shown on (Figure 1), a referral for oncological evaluation raising clinical suspicion of lymphoproliferative disorder, specifically lymphoma or Castleman disease.

Due to this persistent diagnostic uncertainty and progressive mechanical enlargement of the lesion, surgery has been decided.

The patient presented to our health facility with a 9x6x12 cm firm, rubbery, non-tender left parotid swelling as shown in (Picture 1).

A magnetic resonance image (Image1) demonstrated left parotid gland heterogenous enhancement diffuse mass involved both the superficial and deep lobes associated with multiple ipsilateral inflammatory lymphadenopathies and left mastoid effusion which was inconclusive.

The patient underwent a definitive left parotidectomy with preservation of the facial nerve as shown in (Picture 2).

Comprehensive histopathological examination of the surgical specimen established the diagnosis of nodal and extranodal Kimura disease which revealed significant eosinophilia and eosinophilic microabscesses (Figure 2a), interfollicular expansion with polymorphous inflammatory cell infiltrates, increased vascularity, and fibrosis (Figure 2b), involvement of parotid parenchyma, perinodal fatty tissue, and skeletal muscle (Figure 2c).

Laboratory investigations later demonstrated peripheral eosinophilia of $1.82 \times 10^3/\mu L$ (normal value: $0.00–0.50 \times 10^3/\mu L$) and elevated serum immunoglobulin E levels of 591 IU/mL (normal value: <100 IU/mL).

Following surgery, the patient received a tapering course of prednisolone. Owing to concern regarding recurrence and persistent immune dysregulation, steroid-sparing therapy with azathioprine tablets 50 mg twice daily was subsequently initiated under supervision. The patient tolerated the treatment well without adverse effects.

The complete chronological sequence of the patient's multi-year diagnostic journey is summarized in (Table 1).

Table 1: Chronological timeline of clinical presentations, diagnostic shifts, and interventions.

Timeline Phase	Clinical Presentation & Interventions	Working Diagnosis / Suspicions	Rationale & Diagnostic Pitfalls
IN HOME COUNTRY **2014-2019**			
Initial Onset	Left parotid region painless swelling. No constitutional symptoms or focal neurological deficits.	Early localized painless mass	Typical insidious presentation of early-stage major salivary gland Kimura disease.
Initial assessment	Biopsy and computed tomography imaging performed. Surgery recommended but conservative management chosen by the patient's family	Obstructive or inflammatory parotid gland disease	Mimicked structural sialadenitis due to diffuse glandular capsular enlargement.
Few months later	Second computed tomography imaging performed. Refer to Rheumatology clinic. persistent parotid	Systemic lupus erythematosus	Overlapping inflammatory markers led to an unconfirmed autoimmune diagnosis.

	mass with elevated inflammatory markers.		Managed with prednisolone and hydroxychloroquine for 12 months.
RELOCATION TO THE MIDDLE EAST **2020 – till this date**			
Pre-operative	Persistent parotid mass.	Unconfirmed Autoimmune Disease	Rheumatologic medications discontinued
	Neurology teams' assessment for chronic headaches.	Chronic inflammation of the mass.	
	progression of the swelling. Ultrasound-guided biopsy and imaging. Oncology referral.	Lymphoma or Castleman disease	Follicular hyperplasia. Necessitating surgical intervention.

Operative and post-operative	Underwent left parotidectomy with facial nerve preservation.	Kimura disease (Definitive Histopathological Diagnosis)	Kimura disease was confirmed. Initiated on combination maintenance protocol of tapering systemic corticosteroids and azathioprine.

OUTCOME AND FOLLOW-UP

The patient demonstrated significant clinical improvement following combined surgical and immunosuppressive management. During follow-up there was no evidence of recurrent parotid swelling (Picture 3) and the patient remained clinically stable on azathioprine therapy with substantial improvement in quality of life as reported by the patient. No associated allergic, respiratory, gastrointestinal or dermatological manifestations developed during follow-up.

Despite complete surgical resolution and sustained structural stabilization of the parotid swelling under Azathioprine maintenance, the patient's absolute eosinophil count and serum IgE levels remained consistently above the upper reference limits as shown in (Table 2 and 3). The absolute eosinophil count, while dramatically improved from its pre-operative peak of $1.82 \times 10^3/\mu L$ (normal value: $0.00–0.50 \times 10^3/\mu L$), remain at a chronic fluctuating between 0.50 and $0.80 \times 10^3/\mu L$. Concurrently, serum IgE levels remained elevated, reaching 381 IU/mL (normal value: <100 IU/mL) at the 9 months mark.

Long-term surveillance was planned as recurrence has been frequently reported in Kimura disease even after surgical excision.

DISCUSSION AND CONCLUSION

Kimura disease is defined as a rare, benign, idiopathic chronic lymphoproliferative and fibroinflammatory disorder [1–3]. It is characterized by deep subcutaneous lymphoid masses predominantly localized to the head and neck, hyperplasia, marked peripheral blood eosinophilia, and elevated serum Immunoglobulin E (IgE) levels [1–3].

While the precise underlying etiology remains unclear, the core pathophysiology of Kimura disease is driven by dysregulated T-helper 2 (Th2) cell-mediated immune responses, through the release of interleukins, specifically interleukin 4, 5 and 13 (IL-4, IL-5, IL-13). IL-4 and IL-13 stimulate B lymphocytes to undergo class-switching, promoting systemic IgE production. While IL-5 promote proliferation, recruitment, activation, and prolonged tissue survival of peripheral eosinophils with release of eosinophilic granular proteins into the affected salivary glands or subcutaneous stroma, resulting in tissue damage and progressive local fibrosis [1–3,15].

Kimura disease first described in 1948 [4]. It remains an uncommon but increasingly recognized in both Asian and non-Asian populations. Literature demonstrates a wider global distribution, including Europe, North America, and the Middle East [5–7]. This shifting pattern strongly suggests that Kimura disease is more likely underdiagnosed globally rather than strictly geographically restricted [5–7].

Although individuals of Asian descent constitute a significant portion of the Gulf population, a comprehensive review of regional medical literature reveals Kimura disease remain rare across the region, with our case is the second documented case of Kimura's disease in the United Arab Emarat (UAE).

literature from the United Arab Emarat only documented a female in her 30s presented with epiglottic mass, suspected as carcinoma or lymphoma with a risk of airway obstruction. She underwent localized surgical excision of the mass. After histopathological examination of the mass Kimura disease was confirmed. The patient did not start on medical therapy, and she lost subsequent follow up [11].

Published data from the Middle East and Gulf region demonstrated consistent diagnostic challenges where Kimura disease is frequently misdiagnosed as lymphoma or salivary gland disorders. Definitive diagnosis is frequently delayed until histopathological confirmation following surgical excision [6,7, 12-14].

The principal diagnostic difficulty in Kimura disease lies in its non-specific clinical and radiological presentation with fluctuation responses to steroid, which closely mimics autoimmune conditions and malignant condition as lymphoma or salivary gland neoplasms [8–10].

In the present case, a prolonged history of recurrent unilateral parotid swelling with a partial corticosteroid response led to an initial working diagnosis of systemic lupus erythematosus (SLE). Such transient steroid responsiveness has been previously reported in fibroinflammatory lesions and can misleadingly support an autoimmune etiology despite being non-specific [1,5].

Radiological findings in Kimura disease are typically non-specific, often demonstrating enhancing, infiltrative parotid or subcutaneous masses with associated regional lymphadenopathy [9,10]. Similarly, fine-needle aspiration cytology (FNAC) is frequently inconclusive, mostly showing reactive lymphoid hyperplasia with prominent eosinophils but without malignant features [9].

These diagnostic limitations consistent with the prior reports [6,9] and similarly in our case as shown in (Image1) and (Figure1) which led to delay in the diagnosis and mismanagement.

Definitive diagnosis of Kimura disease which distinguishes it from lymphoma can only be achieved by histopathological examination. The classic diagnostic triad includes the preservation of nodal architecture, florid follicular hyperplasia, dense infiltration of eosinophils with occasional eosinophilic microabscesses, and prominent post-capillary vascular proliferation [4,8,11].

In our case, a definitive diagnosis was only established following an excisional parotidectomy, which is highly consistent with literature supporting an excision biopsy as the most reliable diagnostic and therapeutic modality for major salivary gland involvement [4,8].

There is no standardized treatment protocol for Kimura disease. Globally to induce effective initial remission in Kimura disease is receiving systemic corticosteroids, but it has a 60–80% relapse rate on tapering, and on extended maintenance doses leads to glucocorticoid adverse effects. [5,6,8]. Consequently, the introduction of a steroid-sparing agent is vital to maintain long-term clinical remission while minimizing adverse effects of steroid [6,7,12-14,15].

In our case a successful remission was achieved when used a combination maintenance protocol of tapering systemic steroids and Azathioprine as steroid-sparing agent to provide effective maintenance and prevent recurrence. Although most of the data showed the efficacy of other steroid-sparing agents with hardly considering azathioprine as alternative. The outcome in our patient provides valuable support for using Azathioprine as a reliable, reproducible steroid-sparing agent to avoid the high recurrence rates typical of Kimura disease.

A highly instructive and unique feature of our case is the distinct dissociation between clinical remission and persistent laboratory abnormalities during the long-term follow-up period. Despite complete surgical resolution and sustained structural stabilization of the parotid swelling under Azathioprine maintenance, the patient's absolute eosinophil count and serum IgE levels remained consistently above the upper reference limits as shown in (Table2 and 3). The absolute eosinophil count, while dramatically improved from its pre-operative peak of $1.82 \times 10^3/\mu L$ (normal value: $0.00–0.50 \times 10^3/\mu L$), remain at a chronic fluctuating between 0.50 and $0.80 \times 10^3/\mu L$. Concurrently, serum IgE levels remained elevated, reaching 381 IU/mL (normal value: <100 IU/mL) at the 9 months mark.

In Kimura disease, peripheral eosinophilia and elevated IgE levels act as direct reflection of underlying Th2 cell hyperactivity and cytokine production [3,5,7,15]. The persistence of these markers in our patient who is otherwise completely free of detectable masses or systemic symptoms suggests that the underlying immunological drivers are suppressed but not entirely eradicated by the current maintenance dosing which highlights the chronic, relapsing-remitting characteristic feature of Kimura disease.

This observation emphasizes a critical clinical lesson for regional practitioners: treatment success should be primarily measured by the stabilization or regression of the masses and the prevention of structural damage with tolerates the current maintenance medication, rather than aggressively escalating immunosuppressive therapy to achieve a normal laboratory reference range.

This case highlights the importance of considering Kimura disease in the differential diagnosis of patients presenting with chronic painless parotid swelling, with early excisional biopsy and histopathological examination being essential for establishing a definitive diagnosis, avoiding misdiagnosis and preventing unnecessary treatment.

As Kimura disease predominately affects individuals of Asian ethnicity, increased awareness in non-endemic regions, particularly the Gulf region, is essential given its large expatriate population of Asian origin. Clinicians should maintain a high index of suspicion for Kimura disease when evaluating Asian expatriates presenting with painless head and neck lymphadenopathy or salivary gland masses.

DECLARATIONS

Ethics and consent for participate

The manuscript was reviewed and consent was obtained from Mohammed Bin Rashid University Institutional Review Board (MBRU-IRB). The reference Number: MBRU-IRB-2026-905

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor in Chief of this journal.

Availability of data and materials

Not applicable

Competing interests

The authors declare that they have no competing interests

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Authors' contributions

All authors read and approved the final manuscript

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FIGURES AND IMAGES

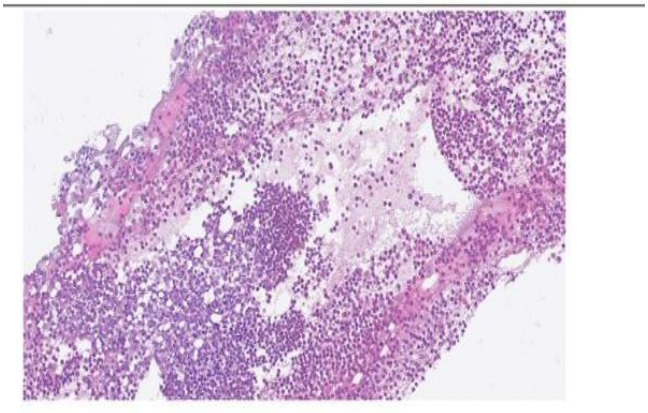


Figure1: Ultrasound guided core biopsy from upper deep cervical enlargement lymph node revealed lymphoid hyperplasia.

Source: original image obtained from laboratory records.

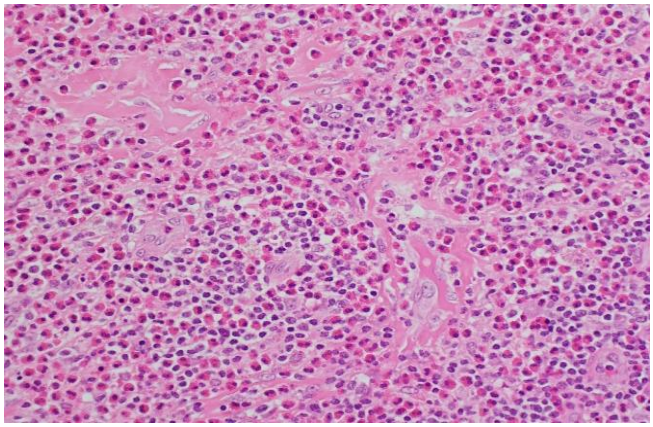


Figure 2a**:** Heavy diffuse inflammatory eosinophilic infiltrates with eosinophilic microabscesses intermixed with background lymphoplasmacytic inflammatory infiltrates, fibrosis and prominent venules H&E x400.

Source: original image obtained from laboratory records.

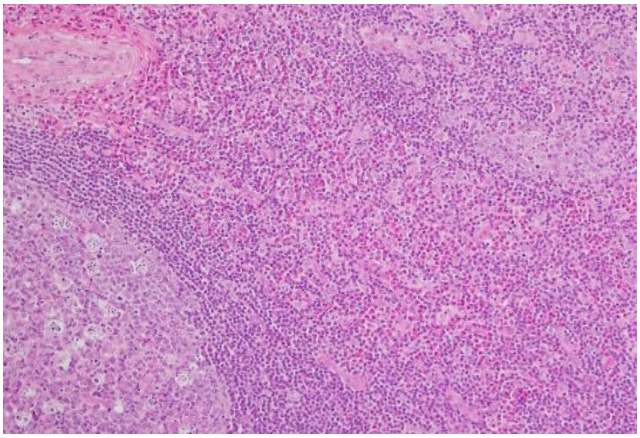


Figure 2b: Reactive lymphoid follicles infiltrated by eosinophils with interfollicular eosinophilic infiltration and prominent blood vessels H&E x200.

Source: original image obtained from laboratory records.

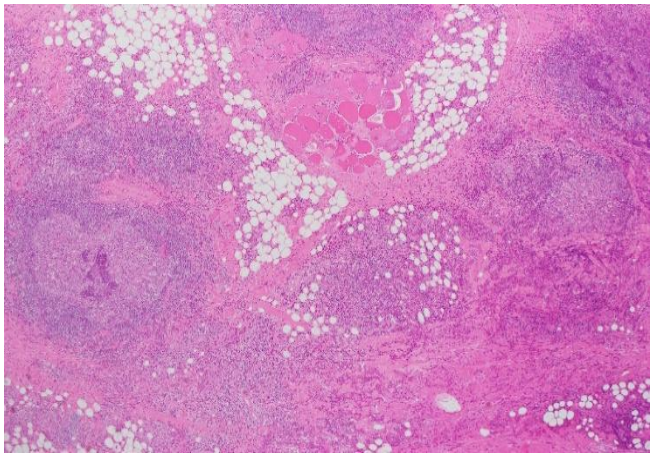


Figure 2c: Tissue replaced by reactive lymphoid follicles, with fibrosis and numerous eosinophilic inflammatory infiltrates in soft skeletal tissue and salivary tissue H&E x40.

Source: original image obtained from laboratory records.



Image 1: MRI demonstrated left parotid gland heterogenous enhancement diffuse mass measuring 9 x 6 x 12 cm at maximum diameter involved both superficial and deep lobes.

Source: original image obtained from radiology records.



Picture 1: Pre-surgery image shows left parotid swelling.

Image obtained with patient consent.



Picture 2: surgery site for left parotidectomy with preservation of the facial nerve**.**

Image obtained with patient consent.



Picture 3: post-surgery and during treatment with Azathioprine tablet showing sustained remission of the mass.

Image obtained with patient consent.

Table 2: Absolute Eosinophil Count (AEC) showing values pre and post immunotherapy (Azathioprine) with consistent high level despite the remission of the mass size.

Date	Results (Reference Range 0.00 - 0.50 × 10^3/μL)
19/05/2023	1.82 × 10^3/μL
08/06/2023	0.00× 10^3/μL
11/07/2024	0.80× 10^3/μL
13/09/2024	1.60 × 10^3/μL
28/10/2024	0.88× 10^3/μL*
20/12/2024	0.60× 10^3/μL
04/02/2025	0.60× 10^3/μL
27/03/2025	0.60× 10^3/μL
23/06/2025	0.50× 10^3/μL
04/09/2025	0.50× 10^3/μL
22/10/2025	0.60× 10^3/μL
180/3/2026	0.80× 10^3/μL
22/04/2026	0.50× 10^3/μL
19/05/2026	0.60× 10^3/μL

*4 weeks on Azathioprine tablets

Source: obtained from patient's laboratory records.

Table 3: Immunoglobulin E level pre and post immunotherapy (Azathioprine)

Date	Results (Reference range <100 IU/ml)
4/9/2025	591 IU/ml (pre)
19/5/2026	381 IU/ml (post)

Source: obtained from patient's laboratory records.