World Journal of Biology Pharmacy and Health Sciences
eISSN: 2582-5542
Cross Ref DOI: 10.30574/wjbphs
Journal homepage: https://wjbphs.com/

(CASE REPORT)

Kimura disease presenting with isolated axillary lymphadenopathy and eosinophilia in a young female: A case report and review of diagnostic and management considerations

Talina Fuentes [1], Cindy Almaraz [2], Marcos Domínguez [3], Rikki Johnson [1], Jessica Jahoda [4, 5] and Mohamed Aziz [5, *]

[1] *American University of the Caribbean, AUC, St. Maarten.*
[2] *Ross University School of Medicine, Barbados.*
[3] *Universidad Iberoamericana (UNIBE), Santo Domingo, Dominican Republic.*
[4] *Memorial Healthcare System, Pembroke Pines, FL, USA.*
[5] *Research Writing and Publication (RWP), LLC, NY, USA.*

World Journal of Biology Pharmacy and Health Sciences, 2026, 27(01), 084-090

Publication history: Received on 06 June 2026; revised on 11 July 2026; accepted on 14 July 2026

Article DOI: https://doi.org/10.30574/wjbphs.2026.27.1.0385

Abstract

Kimura disease (KD) is a rare, benign, chronic inflammatory disorder that predominantly affects young Asian men in the head and neck region. Presentation as solitary axillary lymphadenopathy in a female patient is highly uncommon, and diagnostically challenging. Our case follows a 32-year-old Asian woman with a six-month history of erythematous forearm rash, nocturnal pruritus, and progressively enlarging, painless left axillary swelling. Initial assessment revealed peripheral eosinophilia and markedly elevated serum IgE levels. Contrast-enhanced imaging demonstrated isolated left axillary lymphadenopathy without evidence of organomegaly, systemic nodal involvement, or mediastinal extension. Histopathological analysis identified microabscesses, extensive eosinophilic infiltration, and florid germinal center hyperplasia. Molecular and immunohistochemical (IHC) studies confirmed a polyclonal lymphoid pattern, excluding viral etiologies, IgG4-related disease, and lymphoma.

Management with complete surgical excision was performed following multidisciplinary tumor board review, resulting in symptom resolution and normalization of laboratory findings. Sequential multifocal recurrences, initially in the right cervical region at fourteen months and contralateral axilla, required a gradual transition from systemic corticosteroids to low-dose cyclosporine. Sustained remission was maintained after 18 months of follow-up, with stable renal function throughout the disease course.

This case contributes to the limited literature on atypical presentations of Kimura disease, underscores the importance of considering this diagnosis in cases of unexplained eosinophilic lymphadenopathy irrespective of anatomical location or sex, and highlights the chronic, relapsing-remitting nature of the disease, which necessitates individualized, multidisciplinary long-term management.

Keywords: Kimura disease; Eosinophilic lymphadenopathy; Chronic inflammatory disorder; Peripheral eosinophilia

1. Introduction

Kimura disease (KD) is a rare, idiopathic disorder characterized by subcutaneous swelling, lymphadenopathy, peripheral eosinophilia, and elevated immunoglobulin E levels. [1] It most commonly affects young adults, with a higher prevalence in male and Asian populations. [2] KD classically presents as a painless mass in the head or neck region with extension to salivary and lacrimal glands. [3] Other common sites include the periauricular region, groin, orbit, or

* Corresponding author: Mohamed Aziz

eyelids. [2] Diagnosis is made by histological investigation. Therapies for KD include surgical excision for localized cases, steroids, and radiation. [2] Despite treatment, KD has a high recurrence rate, particularly following surgical excision when used as the singular therapeutic modality. [4]

We present the case of Kimura Disease in a 32-year-old Asian female with initial axillary presentation followed by delayed recurrence in the cervical region. We report this case to highlight the atypical presentation of the primary subcutaneous lesion, its occurrence in a female within a male-predominant disease, and its sequential involvement of another anatomical region. Given the rarity of the disease, this report aims to contribute an atypical clinical presentation that further highlights the need for long-term surveillance in KD.

2. Case Presentation

2.1. Clinical presentation

A 32-year-old Asian female presented with a six-month history of progressively enlarging, painless swelling in her left axilla, accompanied by intermittent low-grade fever and generalized pruritus that worsened at night. She had initially managed her symptoms with oral antihistamines and nonsteroidal anti-inflammatory drugs prescribed by her primary care physician, which provided minimal relief. She sought medical attention after noticing that the axillary mass had doubled in size over the preceding 2 months and that she had developed a pruritic, erythematous rash on her forearms. She denied any history of tuberculosis, autoimmune disease, or malignancy, and she had no recent travel, animal exposures, or new medications. Her past medical history was unremarkable, and she was a non-smoker with no significant family history of hematologic or lymphoproliferative disorders.

2.2. Physical examination and initial investigations

On physical examination, the patient was afebrile with stable vital signs. Palpation of the left axilla revealed a single, firm, mobile, non-tender lymph node measuring 3.5 cm in diameter with no overlying skin changes. No other peripheral lymphadenopathy was appreciated. The spleen and liver were not palpable. Laboratory studies showed a high peripheral eosinophil count of 2,100 cells/μL with a normal total white blood cell count. Serum IgE was markedly elevated to 1,850 IU/mL. Renal and hepatic function tests were normal.

2.3. Radiographic findings and differential diagnosis

Ultrasound of the left axilla demonstrated a well-circumscribed, hypoechoic, oval lymph node with preserved hilar architecture and increased vascularity on Doppler imaging. Contrast-enhanced CT of the chest, abdomen, and pelvis showed isolated left axillary lymphadenopathy without mediastinal, retroperitoneal, or pelvic nodes and no organomegaly. The clinical differential diagnosis included reactive lymphoid hyperplasia, drug-induced lymphadenopathy, parasitic infection (especially filariasis or toxocariasis), Kimura disease, angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin lymphoma with eosinophil-rich infiltrate, and T-cell lymphoma with eosinophilia.

The radiological differential similarly favored benign causes due to preserved nodal architecture, but neoplastic processes could not be fully excluded. Axillary lymph node core biopsy was performed, and histopathology reported preserved architecture with markedly hyperplastic germinal centers, massive infiltration of eosinophils extending from the interfollicular zones into the germinal centers and surrounding capsule, with focal eosinophilic microabscesses. (**Figure 1, A, B, C**)

2.4. Multidisciplinary tumor board discussion

The multidisciplinary tumor board, comprising hematopathologists, radiologists, medical oncologists, and surgical oncologists, reviewed the case. The discussion centered on the striking discrepancy between the massive tissue eosinophilia seen on core needle biopsy and the absence of systemic symptoms or disseminated disease. The team systematically excluded parasitic infections through negative serologies, drug-induced causes through medication reconciliation, and neoplastic conditions such as lymphoma through the absence of clonal markers or atypical cells. ALHE was considered but discounted due to the absence of vascular lesions on imaging.

The board concluded that KD was the most likely diagnosis given the demographic triad of young, Asian female with peripheral eosinophilia, elevated IgE, and biopsy showing florid germinal center hyperplasia with dense eosinophilic infiltration. They recommended complete excision of the large axillary node for definitive histologic and immunohistochemical (IHC) confirmation before initiating any systemic therapy.

2.5. Pathology, ancillary studies, and final diagnosis

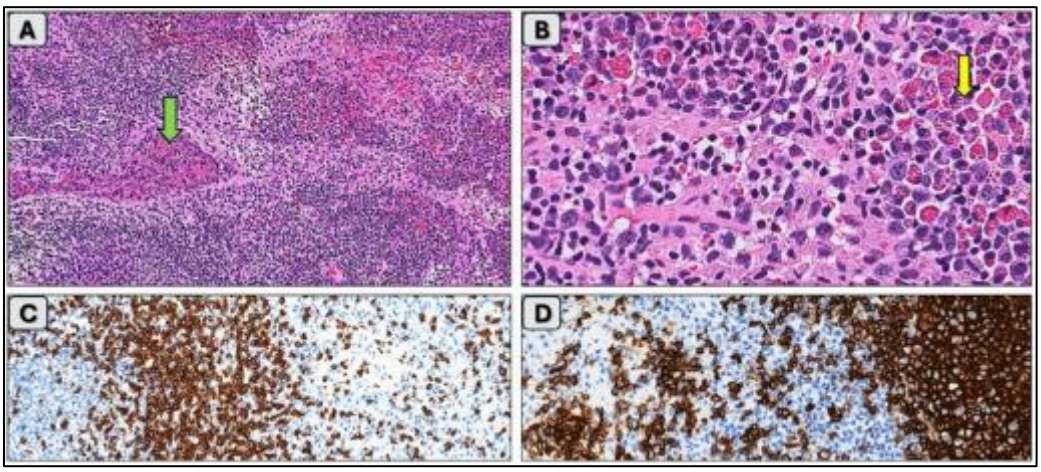
The excised lymph node measured 3.8 cm, and the microscopic examination confirmed the findings noted in the core biopsy. IHC demonstrated CD3-positive T cells and CD20-positive B cells with a polytypic pattern (kappa and lambda light chains), excluding clonality. CD21 highlighted intact follicular dendritic cell meshworks. No CD30-positive or CD15-positive large, atypical cells were seen, ruling out Hodgkin lymphoma. IgG4 immunostaining showed scattered positive plasma cells but it did not meet the criteria for IgG4-related disease. Special stains for fungi and acid-fast bacilli were negative. In situ hybridization for Epstein-Barr virus-encoded RNA was negative. Molecular studies via PCR for T-cell receptor gamma and immunoglobulin heavy chain gene rearrangements showed a polyclonal pattern. Peripheral blood flow cytometry revealed no evidence of a clonal T-cell population or circulating lymphoma cells. These findings confirmed the diagnosis of Kimura disease.

2.6. Treatment, outcome, follow-up and recurrence

Initial treatment consisted of complete surgical excision of the involved left axillary lymph node, which the patient tolerated well. Postoperatively, her pruritus resolved, and her peripheral eosinophil count decreased from 2,100 cells/μL to 450 cells/μL within three months. She remained asymptomatic for twelve months, with surveillance ultrasounds showing no axillary or regional lymphadenopathy.

At month fourteen, however, she returned reporting a new, painless swelling in the right cervical region. Physical examination revealed a 2 cm mobile, non-tender lymph node in the right posterior cervical chain, without fever or systemic symptoms. Laboratory studies showed re-emergence of peripheral eosinophilia to 1,900 cells/μL and an elevated serum IgE of 1,200 IU/mL. Ultrasound of the neck demonstrated a hypoechoic, oval node with preserved architecture but marked cortical thickening. An excisional biopsy of the cervical node was performed, and histopathology again revealed florid germinal center hyperplasia with a massive eosinophilic infiltrate, identical to the original axillary specimen. IHC and clonality studies confirmed a polyclonal pattern, and all neoplastic and other non-neoplastic differential diagnoses were again excluded. The diagnosis of recurrent Kimura disease was therefore established.

Given the recurrence at a different site, the multidisciplinary tumor board recommended transitioning from local therapy to systemic immunomodulation. The patient received oral corticosteroids (prednisone 0.5 mg/kg/day tapered over eight weeks), which led to complete regression of the cervical node and normalization of eosinophil counts. However, when corticosteroids were tapered below 5 mg daily, she developed a second recurrence in the contralateral axilla. The board then initiated low-dose cyclosporine (3 mg/kg/day) as a steroid-sparing agent, resulting in sustained remission.



1A**:** Intermediate power view showing hyperplastic germinal centers, infiltration of eosinophils, and eosinophilic microabscesses (Green arrow) (H&E X40); 1B**:** High power view showing massive infiltration by eosinophils (Yellow arrow) (H&E X40); 1C**:** Lymphoid cells positive for CD3; 1D: Lymphoid cells positive for CD20

Figure 1 Microscopic features of Kimura disease (KD)

At 18 months after the initial diagnosis (and 6 months after starting cyclosporine), the patient remained free of lymphadenopathy, with peripheral eosinophils of 210 cells/μL and IgE of 150 IU/mL. She was advised to follow up every 3 months with a physical examination, complete blood count, renal function monitoring (given cyclosporine's potential nephrotoxicity), and an annual ultrasound of all nodal basins.

This recurrence underscores that Kimura disease, while benign, follows a chronic relapsing-remitting course, and complete surgical excision alone is often insufficient for long-term control when multifocal or recurrent disease develops.

3. Discussion

3.1. Background (History, epidemiology, risk factors, and WHO classification):

Kimura disease is a rare, benign, chronic inflammatory disorder of unknown etiology, first described by Kimm and Szeto in 1937 as an "eosinophilic hyperplastic lymphogranuloma" and later systematically characterized by Kimura et al. in 1948. [5,6] Epidemiologically, the disease is endemic to East and Southeast Asia, predominantly affecting young adult males in their second to fourth decades of life, with a male-to-female ratio ranging from 3:1 to 9:1. [2,7] The exact pathogenesis remains poorly understood. However, current evidence suggests an aberrant immune response driven by a T-helper 2 (Th2) cytokine profile (including interleukin -4, -5, and -13), leading to elevated immunoglobulin E (IgE) production and dense eosinophilic infiltration. [8] While potential triggers such as parasitic infections, trauma, or autoimmune processes have been hypothesized, none have been definitively proven. [9]

Clinically, Kimura disease typically presents as indolent, painless subcutaneous masses in the head and neck region, often accompanied by regional lymphadenopathy and salivary gland involvement. [10] Extranodal and extra-cervical presentations, such as isolated axillary or inguinal lymphadenopathy, are exceedingly rare. [11] The World Health Organization (WHO) Classification of Tumors recognizes Kimura disease as a non-neoplastic haematolymphoid disorder, distinguishing it from malignant lymphoproliferative processes. [12] Despite its benign nature, the disease follows a chronic, relapsing-remitting course. It is associated with renal complications, most notably nephrotic syndrome, in up to 20% of patients, underscoring the need for careful systemic evaluation and long-term surveillance. [13]

3.2. Pathogenesis, Pathophysiology

KD is often accompanied by regional lymphadenopathy, peripheral eosinophilia, and elevated serum IgE levels. [4] Although its pathogenesis remains incompletely understood, proposed mechanisms include trauma, infection, IgE-mediated hypersensitivity, and chronic immune dysregulation, leading to a Th2-predominant immune response. [14,15] Activation of CD4+ T helper cells toward a Th2 phenotype promotes immunoglobulin switching to IgE, leading to elevated serum IgE concentrations, expanded IgE-positive B-cell populations, and characteristic reticular IgE deposition within germinal centers. [15]

Eosinophilic infiltration is a prominent feature, and eosinophil-derived mediators, including major basic protein, eosinophilic cationic protein, and eosinophilic peroxidase, contribute to tissue injury, fibrosis, and the formation of eosinophilic abscesses and germinal center eosinophilic deposits. [4,14]

3.3. Comparative Analysis of Our Case with Existing Literature.

The clinical presentation of this case diverged significantly from the classic epidemiological and anatomic profile of KD, presenting unique diagnostic challenges. While the literature establishes a strong predilection for young Asian males presenting with head and neck subcutaneous masses, our patient was a young female who presented with isolated axillary lymphadenopathy. [2,7] A review of the literature reveals that isolated axillary involvement is exceptionally rare. For instance, a 2017 case report described a 74-year-old Japanese female with a solitary left axillary mass diagnosed as Kimura disease, which similarly required excisional biopsy to exclude occult breast malignancy and lymphoma. [16] In contrast to that older patient, our patient fell within the typical age range but lacked the characteristic head and neck involvement at initial presentation.

The disease course in our patient mirrored the high recurrence rates documented in broader case series. Recurrence is common, affecting up to 40% of patients following initial surgical excision. [17] The transition from localized surgical management to systemic immunomodulation with corticosteroids and eventually cyclosporine aligns with recent treatment algorithms for recurrent KD. [18] Cyclosporine has demonstrated significant efficacy as a steroid-sparing agent, achieving rapid and sustained remission in refractory cases by targeting the underlying Th2-mediated immune dysregulation. [19] This case corroborates the literature indicating that while complete excision is optimal for localized disease, multifocal or recurrent presentations necessitate systemic therapies to control the chronic inflammatory process. [20]

4. What Have We Learned from This Case?

This case provided several critical insights into the diagnosis and management of atypical KD. Diagnostically, it emphasized that Kimura disease must be maintained in the differential diagnosis for unexplained, persistent lymphadenopathy, even when presenting in atypical demographics (females) and uncommon anatomic locations (the axilla). The primary diagnostic lesson was the necessity of integrating clinical findings with histopathology; the striking discrepancy between the massive tissue eosinophilia seen on biopsy and the absence of systemic symptoms or clonal markers was the key to unlocking the diagnosis. Clinicians should recognize that marked peripheral eosinophilia and elevated serum IgE levels, when paired with florid germinal center hyperplasia and eosinophilic microabscesses, are strong indicators of this condition.

From a management perspective, this case reinforced the understanding that KD is a chronic, systemic inflammatory process rather than a strictly localized structural anomaly. While complete surgical excision was effective for initial local control, the subsequent contralateral and distinct anatomic recurrence highlighted the limitations of surgery alone in altering the natural history of the disease. The successful transition to low-dose cyclosporine as a steroid-sparing agent demonstrated the practical utility of targeted immunomodulation in achieving sustained remission. Consequently, long-term surveillance is imperative, not only to monitor disease recurrence across all nodal basins but also to screen for potential systemic complications, such as renal dysfunction, which can emerge insidiously over the course of the disease.

Abbreviations

- Kimura disease (KD)
- Hyperplasia with eosinophilia (ALHE)
- immunohistochemical (IHC)

5. Conclusion

This case of Kimura disease presenting as isolated axillary lymphadenopathy in a young female expanded the recognized clinical spectrum of this rare chronic inflammatory disorder. We presented this case to underscore the diagnostic pitfalls of atypical presentations, in which the condition can easily masquerade as a malignant lymphoproliferative process or an infectious disease. The value of this report to the medical community lies in its demonstration of the critical role of multidisciplinary collaboration and histopathological confirmation in securing an accurate diagnosis. Furthermore, it highlighted the chronic, relapsing nature of KD, illustrating that while surgical excision is valuable for initial diagnosis and local control, recurrent or multifocal disease requires a paradigm shift toward systemic immunomodulatory therapies, such as cyclosporine, to achieve long-term remission.

Compliance with ethical standards

Acknowledgments

Special thanks to Hamzah Alnajjar and Grace Durham for their assistance in reviewing the final manuscript. Additionally, we appreciate the assistance of Grammarly's language editor and the Manus 1.6 program, which provided valuable writing support by identifying and correcting errors in grammar, spelling, punctuation, and writing style, ultimately enhancing the manuscript.

Disclosure of conflict of interest

All authors make the following declarations:

- Payment/services information: All authors have declared that they received no financial support from any organization for the submitted work.
- Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might be interested in the submitted work.

Statement of informed consent

The patient was lost to follow-up, and all attempts to reach the patient were unsuccessful. Therefore, the paper has been sufficiently anonymized to maintain patient confidentiality.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

References

[1] Buder K, Ruppert S, Trautmann A, Bröcker E, Goebeler M, Kerstan A. Angiolymphoid hyperplasia with eosinophilia and Kimura's disease – a clinical and histopathological comparison. JDDG: Journal der Deutschen Dermatologischen Gesellschaft. 2013 Dec 19;12(3):224–8.

[2] Dhingra H, Nagpal R, Baliyan A, Alva SR. Kimura disease: case report and brief review of literature. Medicine and pharmacy reports. 2019 Apr 25;92(2):195.

[3] Sun Q, Xu D, Pan SK, Jeak Ling Ding, Xue Z, Miao CS, et al. Kimura disease: review of the literature. Internal Medicine Journal. 2008 Aug 1;38(8):668–72.

[4] Loperfido A, Cavaliere C, Fionda B, Bellocchi G, Masieri S, Caminati M. Narrative Review of Genetic and Immunological Mechanisms Involved in the Pathogenesis of Kimura's Disease: New Therapeutic Targets. Genes. 2025 Feb 4;16(2):194.

[5] Kimm HT, Szeto C. Eosinophilic hyperplastic lymphogranuloma, comparison with Mikulicz's disease. Chin Med J. 1937;25:699-700.

[6] Kimura T, Yoshimura S, Ishikawa E. On the unusual granulation combined with hyperplastic changes of lymphatic tissues. Trans Soc Pathol Jpn. 1948;37:179-180.

[7] Lagerstrom IT, Danielson DT, Muir JM, Foss RD, Auerbach A, Aguilera NS. A comprehensive review of Kimura disease. Head and Neck Pathology. 2025 Jun 23;19(1):75.

[8] Kim WJ, Kim HK. Current concepts of Kimura disease: pathophysiology and evolution of treatment. Archives of Craniofacial Surgery. 2022 Dec 20;23(6):249.

[9] Zhao F, Zhou M, Mao A, Zhang Y, Chen Y. Kimura disease: a detailed analysis of clinical and radiological manifestations in a retrospective case series. Journal of Inflammation Research. 2024 Dec 31:3371-81.

[10] Akinosoglou K, Tsoupra S, Rigopoulos A, Schinas G, Polyzou E, Akrida I, Labropoulou V, Symeonidis A, Kaiafa G, Kourea E. Kimura's Disease: A Case Report. Cureus. 2025 Aug 18;17(8):e90446.

[11] Syafawani N. Kimura disease: A rare case affecting the inguinal lymph node. International Journal of Surgery Case Reports. 2025 Jun 1;131:111332.

[12] Arfa A, Yang Y, Sandoval-Sus JD. Benign Lymphadenopathy. InNon-Neoplastic Hematologic Disorders: A Quick Review of Modern Diagnostic and Therapeutic Approaches 2024 Oct 24 (pp. 657-680). Cham: Springer Nature Switzerland.

[13] Cordeil S, Hermine O, Hot A. Diagnostic challenges and updated therapeutic strategies of Kimura's disease: A case report successfully treated by dupilumab and review. Medicine. 2023 Nov 24;102(47):e34191.

[14] Wang X, Ng C, Yin W. A comparative study of Kimura's disease and IgG4-related disease: similarities, differences and overlapping features. Histopathology. 2021 Aug 11;79(5):801–9.

[15] Maehara T, Munemura R, Shimizu M, Kakizoe N, Kaneko N, Murakami Y, Masafumi M, Kiyoshima T, Kawano S, Nakamura S. Tissue-infiltrating immune cells contribute to understanding the pathogenesis of Kimura disease: a case report. Medicine. 2019 Dec 1;98(50):e18300.

[16] Kuroda K, Kashiwagi S, Teraoka H, Kinoshita H, Nanbara M, Noda E, Chikugo T, Hirakawa K, Ohira M. Kimura's disease affecting the axillary lymph nodes: a case report. BMC Surgery. 2017 May 26;17(1):63.

[17] Lee CC, Chang SY, Teng WC, Wu CJ, Liu CH, Huang SW, Wu CE, Yu KH, Chan TM. Coexisting nodular sclerosis Hodgkin lymphoma and Kimura's disease: a case report and literature review. International Journal of Molecular Sciences. 2023 Apr 21;24(8):7666.

[18] Lee CC, Feng IJ, Chen YT, Weng SF, Chan LP, Lai CS, Lin SD, Kuo YR. Treatment algorithm for Kimura's disease: A systematic review and meta-analysis of treatment modalities and prognostic predictors. International Journal of Surgery. 2022 Apr 1;100:106591.

[19] Gupta D, Kumari R, Rajesh NG, Manobalan K, Thappa DM. Low-dose cyclosporine for rapid remission and maintenance in recurrent Kimura's disease. Indian Journal of Dermatology, Venereology and Leprology. 2017 Mar 1;83:262.

[20] Liu Y, Liu S, Xu J, Xu X, Wang M. An unusual case of systemic lymphadenopathy-Kimura's disease. Journal of Inflammation Research. 2023 Dec 31:701-5.