Annals of Clinical Case Reports

Case Report
Published: 11 Nov, 2023

Kikuchi-Fujimoto's Disease: A Clinical Onset of Systemic Lupus Erythematosus

*Garcia-Cerda D[1], Zuno-Silva W[1], Rivas-Casillas NC[1], Vázquez-Camacho M[1], Mendoza-Lujambio I[1,2], Marín-Rosales M[1,3] and Garcia-De la Torre I[1,3]**

[1]Department of Immunology and Rheumatology, Hospital General de Occidente, Mexico

[2]SEPI, Higher School of Medicine, Instituto Politécnico Nacional, Mexico

[3]Universidad de Guadalajara, Guadalajara, Jalisco, Mexico

Abstract

Kikuchi-Fujimoto's Disease (KFD), a benign infrequent idiopathic disorder, is suggested to correlate with viral infections and autoimmune disorders, such as Systemic Lupus Erythematosus (SLE). Cervical lymphadenopathy and constitutive symptoms are the cardinal KFD manifestations that could also be present in other diseases, including SLE.

The overlap diagnosis between KFD and SLE needs high suspicion and a good understanding of both pathologies. This case report presents the case of a female patient presenting with bilateral submandibular adenopathy, right palpebral nodule, weight loss, night fever, node biopsy with histiocytic necrotizing lymphadenitis, and antinuclear antibodies 1:320 with speckled pattern (AC-4) and positive anti-Sm and anti-nRNP/Sm.

Keywords: Kikuchi-Fujimoto disease; Histiocytic necrotizing lymphadenitis; Systemic lupus erythematosus

Introduction

Kikuchi Fujimoto's Disease (KFD), known as histiocytic necrotizing lymphadenitis, is an interspecific regional lymphadenopathy. Due to the inflammatory and immunological phenomena, an association between KFD and SLE has been reported [1,2]. This case report presents the correlation between KFD and SLE and a literature review.

Case Presentation

A 25-year-old female with a non-contributory clinical history presents to the general medical hospital ward. The clinical manifestations started two months prior with painful stony-hard bilateral submandibular adenopathy of rapid growth, with an approximate diameter of 1 cm. Other manifestations were weight loss of 2 kg, arthralgia, night fever, and periorbital edema accompanied by a right conjunctival nodule.

The physical examination showed a mild bilateral exophthalmos with right periorbital edema, painful 1 cm right-sided cervical and 2 cm left infraclavicular adenopathy's, a painful 4 cm left submandibular lymph node, and no evidence of arthritis. The laboratory tests showed hemoglobin 11.8 g/dL, platelets 248 × 10^3/mL, leukocytes 3.81 × 10^3/mL, neutrophils 2.92 × 10^3/mL, lymphocytes 1.51 × 10^3/mL, positive Antinuclear Antibodies (ANA) 1:320 with a fine-speckled nuclear pattern (AC-4) and positive anti-Sm and anti-nRNP/Sm antibodies.

The neck CT scan showed bilateral cervical adenopathy's (1b), while the skull MRI reported orbital infiltrative lesions and extraconal space in lateral angles corresponding to lacrimal glands (1c and 1d). An excisional biopsy documented histiocytic necrotizing lymphadenitis compatible with EKF and ruled out an infectious process and malignancy (1a). KFD and SLE were established as the diagnoses, according to the fever, leukopenia, ANAs positivity, anti-nRNP/Sm, and anti-Sm (ACR/EULAR 11pts). The patient obtained a favorable clinical response with glucocorticoids, antimalarials, and azathioprine.

Discussion

KFD, a rare idiopathic disorder, was described in 1972 with global distribution and a higher prevalence in Asia, principally in the Japanese population [3]. Some reports speculate

OPEN ACCESS

Correspondence:
Ignacio Garcia-De la Torre, Department of Immunology and Rheumatology, Hospital General de Occidente, Mexico

Received Date: *27 Oct 2023*
Accepted Date: *06 Nov 2023*
Published Date: *11 Nov 2023*

Citation:
Garcia-Cerda D, Zuno-Silva W, Rivas-Casillas NC, Vázquez-Camacho M, Mendoza-Lujambio I, Marín-Rosales M, et al. Kikuchi-Fujimoto's Disease: A Clinical Onset of Systemic Lupus Erythematosus. Ann Clin Case Rep. 2023; 8: 2518.

ISSN: 2474-1655.

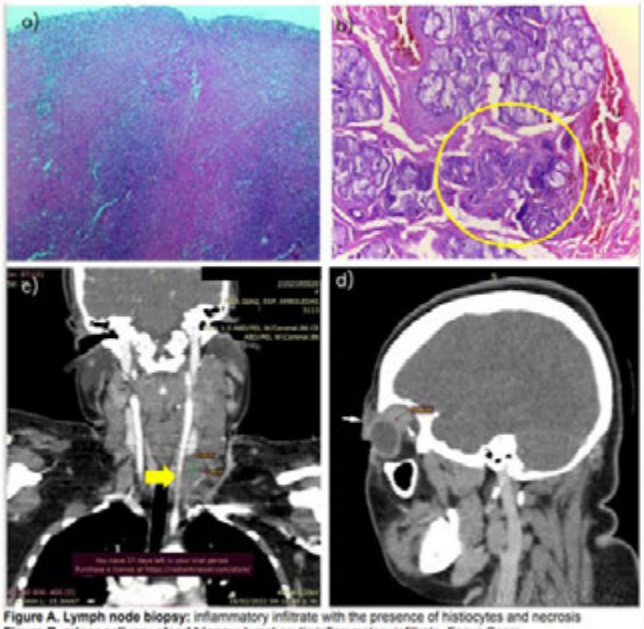

Figure 1: Imaging and histopathological findings.
Lymph node biopsy revealed parenchymatous tissue with regularly circumscribed paracortical necrosis and irregular areas accompanied by cartographic appearance, abundant tissue elements of karyorrhexis, and diffuse reactive histiocytic lymphoproliferation with a starry sky image, without multinucleated giant cells (a). Minor salivary gland biopsy showed lymphocytic inflammatory infiltrate, Focus Score 2 (b). Cervical adenopathy (2.49 cm × 1.89 cm) perivascular carotid (c). The cranial MRI scan shows periorbital infiltrative lesions (d).

an association between infectious processes (toxoplasmosis, infectious mononucleosis, tuberculosis, among others) and systemic autoimmune diseases such as SLE [3,4].

The primary clinical manifestation is a variable size (0.5-6 cm) cervical lymphadenopathy of 3 weeks of evolution, commonly accompanied by fever predominantly at night, weight loss, vomiting, and leukopenia. In some cases, it could also affect extra nodal organs, including various ophthalmologic manifestations.

The most common histopathological features to establish the diagnosis by histopathological assessment of the lymph node are necrotizing lymphadenitis, histiocytes, foamy histiocytes, plasmacytoid dendritic cells, and karyorrhexis. According to the clinical phase, the histopathological findings vary and are classified into proliferative, necrotizing, and xanthomatous [4,5].

Lymphadenopathy prevalence in SLE is 12% to 59% [6]. Up to 47% of cases reported a simultaneous onset and coexistence between KFD and SLE [7]. ANAs are positive 90% when suspicion of lupus overlaps with necrotizing lymphadenopathy is present. In addition, immunospecificity was detected against anti-Sm and anti-RNP/Sm antibodies with a presentation frequency of 14% and 22%, respectively [1].

Conclusion

KFD, as the initial manifestation of SLE, is a challenging diagnosis. Both diseases share similar clinical, histological, and pathophysiological characteristics. It is not yet defined if the association is the coexistence of both pathologies or simply the spectrum of one disease. This case report concluded the SLE diagnosis based on the symptoms, ANAs, and immunospecificity against anti-Sm and anti-RNP/Sm antibodies.

References

1. Sharma V, Rankin R. Fatal Kikuchi-like lymphadenitis associated with connective tissue disease: A report of two cases and review of the literature. Springer plus. 2015;4(1):167.
2. Kucukardali Y, Solmazgul E, Kunter E, Oncul O, Yildirim S, Kaplan M. Kikuchi-Fujimoto Disease: Analysis of 244 cases. Clin Rheumatol. 2007;26(1):50-4.
3. Bosch X, Guilabert A. Kikuchi-Fujimoto disease. Orphanet J Rare Dis. 2006;1(1):18.
4. Deaver D, Horna P, Cualing H, Sokol L. Pathogenesis, diagnosis, and management of Kikuchi-Fujimoto disease. Cancer Control. 2014;21(4):313-21.
5. Jamal AB. Kikuchi Fujimoto Disease. Clin Med Insights Arthritis Musculoskelet Disord. 2012;(5):63-6.
6. Bosch X, Guilabert A, Miquel R, Campo E. Enigmatic Kikuchi-Fujimoto disease: A comprehensive review. Am J Clin Pathol. 2004;122(1):141-52.
7. Londhey VA, Buche AS, Kini SH, Rajadhyaksha GC. Kikuchi Fujimoto disease and systemic lupus erythematosus - A rare association. J Assoc Physicians India. 2010;58(10):642-3.