Kikuchi-Fujimoto Disease: Decoding the Enigma of Unexplained Symptoms in a Young Indian American Female

Neepa N. Patel MD[1], Aaqib Quadri[2], Aakriti Singla[3], Fred Nora MD[1], Aditi Swami[1], Mohammed Moizuddin MD[1]

[1]Department of Internal Medicine, Pathology, Infectious Disease and Pulmonary/Critical Care, Texoma Medical Center, Denison, Texas, [2]Lake Erie College of Osteopathic Medicine, College of Osteopathic Medicine, Erie, PA, [3]University of Texas at Austin, College of Natural Sciences, Austin, TX

Introduction

- Kikuchi-Fujimoto (KFD) is typically found in young patients of Asian decent, thought it has been reported in all ages and ethnicities worldwide.[1]
- Patients with KFD often recover before diagnosis can be made, as it is an extremely rare disease.[2]
- KFD is associated with systemic lupus erythematosus (SLE) and other conditions, such as lymphoma, organizing pneumonia, and VEXAS syndrome.[3,4,5]

Case Presentation

- A 22-year-old Indian American female with a history of atopic dermatitis and asthma presented with recurrent fever and lymphadenopathy over a three-month period.
- She denied smoking, drinking, and drug usage.
- In addition to chills, headaches, rash, and arthralgia, she also had lymphadenopathy involving bilateral cervical and axillary lymph nodes.
- Mycobacterial, fungal, and autoimmune tests were done, as well as a PET scan and a bone marrow biopsy, which showed no suggestive signs of malignancy.
- She underwent core lymph node biopsy that showed lymphoid tissue without evidence of necrosis or increased histocytes.
- Subsequent excisional lymph node biopsy was done and upon excision, the lymph node architecture was partially effaced by paracortical lymphoid hyperplasia composed of small lymphocytes, immunoblasts, and histocytes.
- Some histocytes expressed myeloperoxidase and the lymphocytes were predominately CD3 positive T-cells.
- Histopathologic findings were consistent with a diagnosis of KFD.
- The patient's symptoms improved with supportive therapy and steroids that were tapered over a period of six weeks.

Hospital Course and Imaging

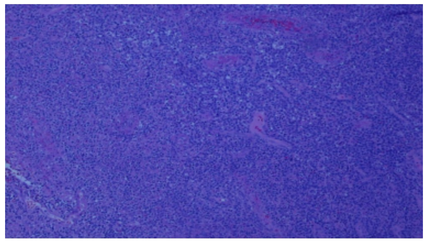

Figure 1: Area of necrosis on left adjacent to population of histiocytes and immunoblasts on right. Necrotic areas exhibit abundant apoptotic bodies, karyorrhexis, and debris. (10x objective)

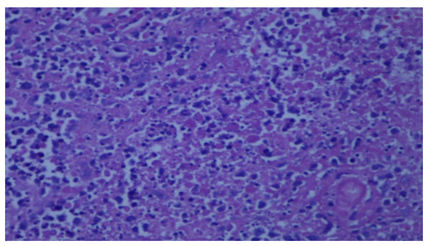

Figure 2: High power view of necrosis with eosinophilic material and nuclear debris. Note absence of inflammatory infiltrate or granulomas. (40X objective)

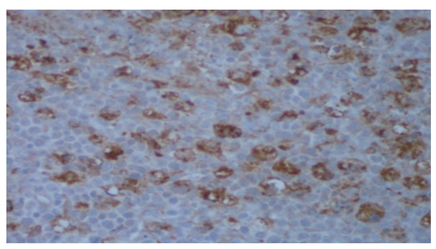

Figure 3: CD68 immunostain highlighting histiocytes and areas surrounding necrosis.

Discussion

- The presentation of KFD in a young Indian American female underscores the diversity of KFD's occurrence, highlighting that KFD is not confined to any specific ethnicity or geographic location.[6]
- KFD is a self-limiting disease with a generally favorable prognosis. However, it can sometimes be associated with SLE, making follow-up crucial.[7]
- KFD has also been associated with various other conditions, including Castleman's disease-like syndrome (CDLS), lymphoma, and VEXAS (Vacuoles, E1 enzyme, X-linked, Autoinflammatory, Somatic) syndrome, requiring a thorough evaluation of symptoms, histopathological examinations, imaging studies, laboratory tests, lymphoma workup, adding to diagnostic challenges.[8]
- Clinicians should be aware of this rare disease that poses a diagnostic challenge to ensure accurate diagnosis and management.

References

[1]Deaver D, Naghashpour M, Sokol L. Kikuchi-fujimoto disease in the United States: three case reports and review of the literature [corrected]. Mediterr J Hematol Infect Dis. 2014 Jan 1;6[1] Erratum in: Mediterr J Hematol Infect Dis. 2014;6[1].
[2]Bosch X, Guilabert A. Kikuchi-Fujimoto disease. Orphanet J Rare Dis. 2006 May 23; 1:18.
[3]Hua F, et al. "Kikuchi-Fujimoto Disease: A Clinical Review." BMC Infectious Diseases. 2010; 10:64.
[4]Yoshino T, et al. "Histopathological Studies on Cervical Lymphadenopathy: Kikuchi-Fujimoto Disease, Glandular Fever, and Toxoplasma Lymphadenitis." Human Pathology. 2000; 31(11):1328.
[5]Yılmaz U, et al. "Clinical and Laboratory Characteristics of Patients with Kikuchi-Fujimoto Disease: A Cross-Sectional Analysis." Clinical Rheumatology. 2022;41(11):3589-3592.
[6]Bosch X, Guilabert A, Miquel R, Campo E. Am J Clin Pathol. 2004 Jul;122 [1]:141-52.
[7]Singh JM, Shermetaro CB. Kikuchi-Fujimoto Disease in Michigan: A Rare Case Report and Review of the Literature. Clin Med Insights Ear Nose Throat. 2019 Feb, 27:12:1179550619828680.
[8]Kumar V, Abbas AK, Aster JC. "Diseases of the lymph nodes." Robbins Basic Pathology. 10th edition. Elsevier. 2017.