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Kikuchi's Disease presenting as Lupus Erythematosus

Olabola Awosika

Chauncey A. McHargue

Kikuchi's Disease Misdiagnosed as Lupus Erythematosus

Olabola Awosika, MD, MS, Zoë Smith, BS, and Chauncey McHargue, MD

Department of Dermatology, Henry Ford Health System, Detroit, MI

History

- A 34-year-old Bengali man with past medical history of hyperthyroidism presented to Dermatology with a red asymptomatic rash affecting the ears, face, and upper body for 1 year.
- He endorsed fever prior to onset of his rash and chronic muscle aches, joint pain, fatigue, and 10-pound weight loss over the past 2 months.
- He had previously been treated with oral clindamycin and triamcinolone ointment with no improvement and prednisone taper with recurrence.

Examination

- On the glabella, eyebrows, cheeks, nose, chin, and back, there were erythematous indurated plaques and papules and hyperpigmented brown patches.
- On the right plantar toes, there were tender and thin desquamative plaques with petechiae.
- The distal half of all toenails were remarkable for transverse brown bands.

Histopathology

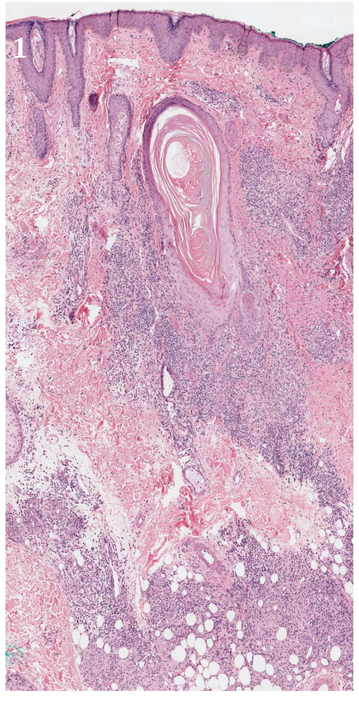

Figure 1: Punch biopsy from the left medial cheek and left mid-back revealed a dense lymphohistiocytic infiltrate in the superficial and deep dermis. Lymphocyte karyorrhexis was also observed. CD4 and CD8 staining revealed a predominance of CD8 lymphocytes and Granzyme B positivity. T-cell clonality studies failed to detect a dominant clonal population.

Clinical Photos

Figure 2: A) Superior forehead, glabella, eyebrows, nasal dorsum and tip, and bilateral cheeks with erythematous indurated papules and plaques.

B) Scattered across the entire back there are numerous follicular based erythematous papules, small plaques, and hyperpigmented brown macules and patches.

Course and Therapy

- At time of presentation, the patient had a biopsy done by an outside dermatologist demonstrating interface dermatitis with superficial and deep perivascular chronic inflammatory infiltrate concerning for lupus erythematosus.
- He was empirically started on a prednisone taper and fluocinonide cream.
- Due to lack of improvement, repeat biopsy was performed showing features concerning for Kikuchi-Fujimoto's Disease.
- Topical steroids were discontinued and he was referred to general surgery for a lymph node biopsy to rule out lymphoma.
- However, due to resolution of lymphadenopathy at presentation, surgical exploration was not recommended.
- During this time, the patient had spontaneous improvement of his skin and associated symptoms.

Discussion

- Kikuchi-Fujimoto Disease (Kikuchi's disease or histiocytic necrotizing lymphadenitis) is a rare, benign condition presenting with fever and cervical lymphadenopathy.
- It is most commonly seen in people of Asian descent and adults under 40 years of age.
- The etiology of the disease is unknown but an association with viral infections (including EBV, parvovirus B-19, HIV, and HHV 6) has been proposed.
- Skin manifestations develop in up to 40% of patients with Kikuchi's disease and can be indicative of a more severe clinical course.
- Common findings include erythematous macules, papules, and plaques affecting the face, trunk, and upper extremities.
- Systemic lupus erythematosus (SLE) has been reported in association with Kikuchi's disease in up to 25% of cases.
- Patients with positive serologies, arthralgias, skin manifestations, and weight loss are most at risk for the development of SLE.
- Thus, long term monitoring for SLE is required in patients with Kikuchi's disease.
- Diagnosis of Kikuchi's disease can be challenging as it is frequently mistaken for malignant lymphoma or SLE.
- Accurate diagnosis requires excisional lymph node biopsy demonstrating focal areas of necrosis surrounded by lymphocytes, immunoblasts, and histiocytes.
- Dermatopathology can also reinforce these findings with cutaneous biopsies typically showing karyorrhectic debris with surrounding lymphohistiocytic infiltrate.
- Treatment for Kikuchi's disease is primarily supportive as it tends to resolve within 1- 4 months.
- In severe protracted disease course, oral corticosteroids or hydroxychloroquine may lead to improvement.

References

1. Mathew LM, Kapila R, Schwartz RA. Kikuchi-Fujimoto disease: a diagnostic dilemma. Int J Dermatol. 2016;55(10):1069–1075.
2. Williams EE, Patel N, Guo H, Hurley MY, Abate MS. Kikuchi-Fujimoto disease diagnosed by correlating skin and lymph node biopsies. 2019;5(5):416-418.