Kikuchi's Disease presenting as Lupus Erythematosus

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Kikuchi’s Disease Misdiagnosed as Lupus Erythematosus
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### 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

![Clinical Photos](Image)

### 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.

### 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 for a 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.

### References