INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

KIKUCHI'S DISEASE – A TIMELY REMINDER TO GREENWALD'S LAW

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Background: Kikuchi disease (histiocytic necrotizing lymphadenitis) was first described in 1972 by Kikuchi. Historically misdiagnosed as malignant lymphoma, it is a rare lymphohistiocytic disorder.

Observation: A 29 year old female Chinese with a history of systemic lupus erythematosus (SLE) presented with bilateral hand rashes of a month's duration. It spread proximally from her fingertips and was associated with pain. There was associated mucosal ulcers and arthralgia. She was constitutionally well otherwise.
Clinically, differential diagnoses included vasculitis or chilblain lupus.
A skin punch biopsy of the right finger demonstrated mild psoriasiform hyperplasia. The basement membrane was intact and not thickened. No basal vacuolar alteration was seen. There was a superficial and deep perivascular infiltrate of lymphocytes and foamy histiocytes in association with leukocytoclasia. Neutrophils were not a feature. No necrotizing vasculitis was seen. Dermal mucin was slightly increased. Ziehl-Neelsen and Gram stains were negative. This was clinicopathogically consistent with Kikuchi disease.

Discussion / Key Message: Kikuchi disease classically presents with fever and lymphadenopathy. The triad of fever, rashes and bilateral cervical lymphadenopathy is more commonly seen in children. Cutaneous lesions are variable- consisting of papulonodules, facial malar erythema or plaques. No specific laboratory test is diagnostic. Up to 15% of patients will have elevated anti-nuclear antibodies level. Some authors consider Kikuchi disease to be a forme-fruste of SLE.
It is estimated approximately 10-30% of Kikuchi disease cases will go on to develop autoimmune diseases.
Kikuchi disease differentiates itself from SLE in its spontaneous resolution over months and localized adenopathy.
There is no established treatment guideline for Kikuchi disease. Recommended treatments include conservative measures, oral corticosteroids or hydroxychloroquine.
This report highlights the need to maintain an index of suspicion for Kikuchi disease, especially in a closely related setting of underlying SLE.