SKIN MANIFESTATIONS OF INTERNAL DISEASE

SARCOID-LIKE DERMATITIS IN THE SETTING OF CHRONIC VARIABLE IMMUNODEFICIENCY: SHOULD SARCOID PATIENTS BE SCREENED FOR CVID?

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Abstract: Chronic variable immunodeficiency (CVID) is a disorder of unknown etiology that presents with recurrent infections, decreased immunoglobulins of mainly IgG type, and variable T cell deficits. Patients can also develop diffuse granulomatosis of not only the skin, but the lungs (granulomatous and lymphocytic interstitial lung disease), and other organs.1 With so many similarities to sarcoidosis, should patients with sarcoid be screened for CVID?

Background: CVID is a heterogeneous group of immunodeficiencies that is relatively common. It has variable signs and symptoms, with recurrent respiratory infections being the most common. Granulomatous and lymphocytic interstitial lung disease (GLILD) occurs in up to 20% of patients. It is an aggressive disorder that histologically is very similar to pulmonary sarcoidosis. These patients can have elevated ACE levels as well as a positive Keim test similar to sarcoidosis.

Observation: A 53-year-old female with a PMHx of CVID with GLILD presented with a recurrent, slightly pruritic of 6 months duration. Exam revealed multiple excoriated, annular plaques on the extremities, neck, and back. A biopsy revealed epithelioid granulomatous dermatitis most consistent with sarcoidosis; however, with the patient's history, a diagnosis of CVID associated granulomatous dermatitis was favored.

Key Message: The described rash of CVID is often variable and no distinguishing features between it and sarcoidosis either clinically or histologically have been identified. The rash of CVID, however, is often associated with granulomatous disease elsewhere including lungs, lymph nodes, spleen, and liver.
The most reliable way to separate these two disorders is by circulating immunoglobulin levels as well as functional T cell studies. If a history of recurrent infections is elicited amongst sarcoid patients, these tests should be performed, as therapy and prognosis for these two disorders is very different. Lastly, patients with CVID have a less favorable

prognosis and can suffer from lymphomas, bronchiectasis, and enteropathy5.