Darier-Roussy Variant of Sarcoidosis

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Abstract

Darier-roussy disease (subcutaneous nodular sarcoidosis) is a rare variant of sarcoidosis. Classically, lesions present as asymptomatic, firm, mobile, subcutaneous nodules without epidermal changes.

We report a case of darier-roussy disease and pulmonary sarcoidosis that presented with lesions involving the hands and extremities. A biopsy of the left arm was the biopsy revealed a normal epidermis with a deep dermal inflammatory infiltrate comprised of well-demarcated "naked granulomas" embedded in a dense fibrotic stroma.

Our case represents the clinicopathologic spectrum on which sarcoidosis can present and underscores why sarcoidosis is often known as the great imitator.

Case Report

58 year old Caucasian female presented to the dermatology office with complaints of masses over her hands and upper and lower extremities. The lesions developed over the past 4.6 weeks. The patient complained of mild to moderate shortness of breath. All other review of systems was negative. Patient denied recent travel or new medications. Past history is significant for asthma, COPD, atrial fibrillation, hypothyroidism and breast cancer.

Exam

Bilateral mildly erythematous deep seated nodules and plaques involving the posterior lateral upper arms and extensor elbows (Fig. 1). Multiple Nodules were also found on the anterior lateral lower extremities. Deep nondiaphragm firm plaques and nodules over bilateral distal and proximal volar hands several overlying interphalangeal joints (Fig. 2).

Examination of the skin over the chest, back, face, mouth, hair, and nails was noncontributory.

Diagnostic Testing

A four millimeter punch biopsy from the left lateral forearm was performed with special attention to include the underlying subcutaneous fat. The results revealed a normal epidermis and a deep dermal inflammatory infiltrate comprised of well-demarcated granulomas embedded in a dense fibrotic stroma (Fig. 3). The subcutaneous tissue showed "naked" granulomatous dermatitis (Fig. 4). The biopsy was consistent with subcutaneous sarcoidosis, also known as darier-roussy disease.

Discussion and Treatment

The constellation of clinical findings together with the histopathologic changes represented the darier-roussy disease variant of sarcoidosis.

The patient was referred for a pulmonology consult given her respiratory symptoms and started on prednisone 60mg daily. Her symptoms promptly responded to therapy. The steroids were tapered with the plan to transition to a steroid sparing regimen therapy such as hydroxycloroquine

Discussion

Sarcoidosis is a disease that can affect all ages, gender, and ethnicities. With peak incidence distribution in a bimodal pattern; age 25-35 and 45-65. The most common presentation is in African American women in their fourth decade. While the pathogenesis of sarcoid is unknown, it is believed to be immune-mediated. The noncaseating granulomas seen in sarcoid are made up of CD4 helper T cells. Patients are also noted to have elevated levels of IFN-gamma and IL-2 with a TH1 type immune response.

Cutaneous sarcoidosis can present clinically with a variety of cutaneous scenarios and is often known as a "great imitator". Approximately 25% of patient with sarcoidosis will have cutaneous findings. A majority of patients have systemic manifestations that can affect the lungs, peripheral lymph nodes, heart, kidneys, gastrointestinal tract, nervous system, liver, spleen, endocrine glands, muscle, and bone. One of the most frequent findings is lung involvement, affecting approximately 90% of patients.

This patient presented with darier-roussy disease also known as sarcoidal panniculitis. It is characterized by asymptomatic subcutaneous firm mobile nodules without epidermal changes.

References