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Case Report

Case of Cutaneous Sarcoidosis with Multiple Morphologies and Review of the Clinical Spectrum

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Abstract

Sarcoidosis is a multi-organ disease that commonly presents with cutaneous manifestations. However, it can clinically mimic innumerable skin conditions, posing a diagnostic challenge for the clinician. Here, we describe a case of a 51-year-old female with cutaneous sarcoidosis exhibiting multiple morphologic patterns. Physical exam showed erythematous to violaceous indurated, large plaques and deep firm nodules on the upper and lower extremities. Two months later, she developed a diffuse papular eruption on extremities and involving a prior scar. Biopsies showed granulomatous dermatitis consistent with sarcoidosis. Imaging revealed pulmonary involvement. This case highlights some of the cutaneous manifestations of sarcoidosis. We propose that in the presence of multiple co-existing morphologies, a single process such as sarcoidosis should be strongly considered.

Introduction

Cutaneous findings are observed in an estimated 30-40% of patients with sarcoidosis [1]. On exam, brown, red, or skin-colored infiltrative papules and plaques without overlying scale are the most often encountered lesions. However, a host of other clinical presentations exist either individually or in combination. As such, cutaneous sarcoidosis can often mimic a different disease process. We report a case of a 51-year-old female with cutaneous sarcoidosis with different clinical presentations.

Case Report

A 51-year-old Caucasian female originally presented to the rheumatology clinic with a three-week history of painful swelling and "Knots" on her bilateral forearms and hands. The patient was referred to dermatology for evaluation. Her past medical history was significant for type II diabetes mellitus, Hashimoto's thyroiditis and retinoblastoma of the right eye requiring enucleation as a child. On clinical examination, she was noted to have erythematous to violaceous indurated, large edematous plaques on the bilateral extensor arms and distal lower legs as well as deep firm nodules (Figure 1). The differential included morphea vs panniculitis vs eosinophilic fasciitis vs other deep dermal inflammatory process.

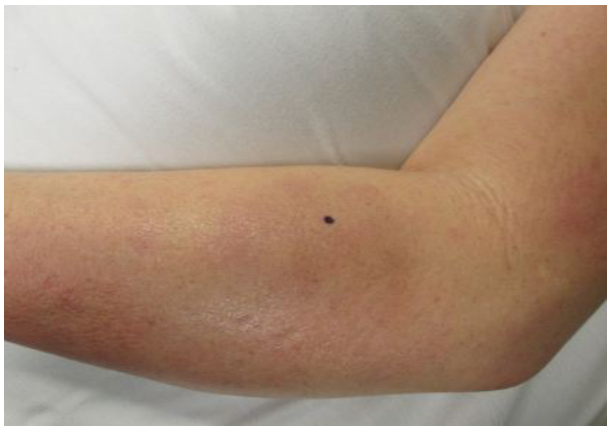

Figure 1: View of an erythematous to violaceous plaque on the left proximal forearm with peaud'orange epidermal change. The biopsy site is marked in ink.

A punch biopsy was performed on the left proximal forearm and revealed granulomatous dermatitis with minimal surrounding inflammation (Figure 2). Fungal and mycobacterial stains were negative, and no polarizable foreign material was appreciated. Subsequently, a tissue culture was performed and was negative for organisms.

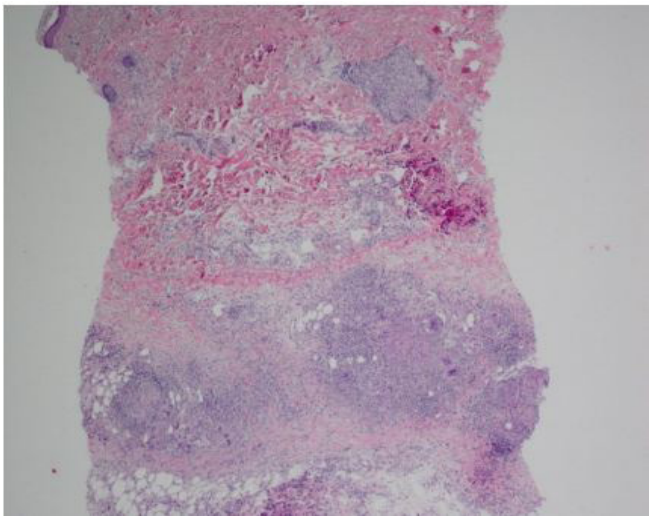

Figure 2: 4x views of deep granulomatous inflammation extending into subcutaneous tissue present on initial biopsy of sclerodermoid plaque from patient's left forearm. Subsequent stains and tissue cultures were negative for infection.

Further work up included a chest x-ray which showed mild bibasilar atelectasis. She did have a slight elevation in her angiotensin converting enzyme level. A chest computed tomography scan was performed and showed mild septal thickening with 1-2 mm ground glass nodular opacities scattered throughout the bilateral lungs. There were also numerous enlarged mediastinal, hilar, and periportal lymph nodes. She was later evaluated by pulmonology who recommended pulmonary function tests. She was started on Plaquenil 200 mg twice daily.

Two months later at clinic follow up, she developed the appearance of new erythematous papules diffusely scattered on her dorsal hands, forearms, and upper arms (Figure 3). Also noted, of new onset, were erythematous papules along an old scar on the left frontal hairline (Figure 4). She continued to complain of fatigue and joint pain in the wrists, knees, and hands, although reported to have improvement in the indurated plaques and nodules that she presented with initially. Biopsy of the new eruption also showed a granulomatous dermatitis with negative infectious stains.

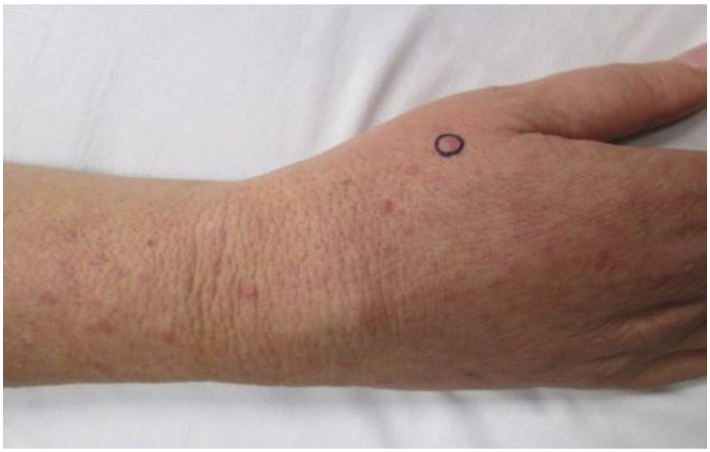

Figure 3: Diffuse papular eruption on bilateral forearms and dorsal hands. Biopsy site marked in ink.

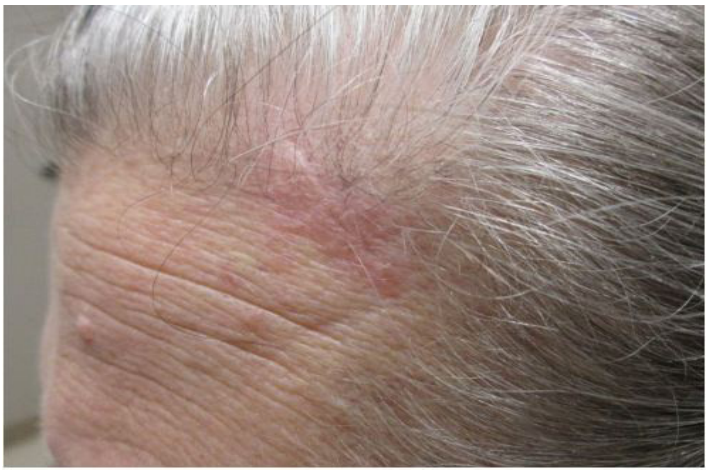

Figure 4: Small eruption of papules on the left frontal hairline around previous traumatic scar.

The patient was continued on Plaquenil and also was given a three-week taper of prednisone. This almost completely cleared the papular eruption on the forearms. She continues to use topical Clobetasol for the remaining areas of induration.

Discussion

The vast spectrum of morphologies associated with cutaneous sarcoidosis remains a diagnostic challenge (Table 1). The most common type of lesions is maculopapular with reddish-brown hyperpigmentation. Lupus pernio is characterized by chronic, violaceous papules or plaques affecting the central face. Lesions may

also manifest as an enlarging, previously inactive scar. Plaque-type sarcoidosis tends to be symmetric on extensor surfaces, often exhibiting thick scale that mimics psoriasis. Hypopigmented patches are seen with a central red papule giving them a "Fried-egg" appearance. Flat-topped, lichenoid lesions are rare, but predominant in children. Ulcerative forms may cause previous lesions to ulcerate or new lesions to appear on the lower extremities. Additionally, subcutaneous nodules, ichthyosiform, morpheaform, and erythrodermic manifestations have been reported [2]. The presence of non-caseating granulomas on histopathology will help confirm the diagnosis [3].

Type	Findings	Type	Findings
Maculopapular	• *Most common type* • *Red, brown slight infiltration, no epidermal change* • *Often spontaneously resolve*	*Hypopigmented*	• *Well demarcated, round, hypopigmented patches* • *Some with Central red papule (fried egg appearance)*
Subcutaneous Nodules	• *Darier-Roussy disease* • *~ 1 - 6% of cases* • *Painless mobile nodules, no epidermal change*	*Lichenoid*	• *1-3mm flat topped, skin colored papules* • *~ 1-2% of cases* • *Reported more often in kids*
Lupus pernio	• *Violaceous plaque-like infiltration of the nose, cheeks and earlobes* • *Uveitis, pulmonary fibrosis, bone cysts* • Chronic course	*Ulcerative*	• *Located mainly on lower legs* • *Evolve into/from atrophic or papulonodular lesions* • *Traumatic activation of atrophic plaque*
Erythema Nodosum	• *Non-specific lesions* • *Seen in Lofgren's syndrome (acute)* • *Good prognostic sign*	*Ichthyosiform*	• *Rare; 23 reported cases* • 0.1 cm to 1 cm irregular a*dherent, greyish plaque with heavy scale* • *Lower legs*
Scar/Tattoo	• *Common; ~ 10% of cases* • *Often associated with polarizable foreign bodies* • *Possible sign of reactivation*	*Erythrodermic*	• *Diffuse erythema, induration and scaling > 90% of body* • *Can have areas of sparing*
Plaques	• *Infiltrated brown/red lesions, usually confluent papules* • *Extensor surfaces* • *Indicates chronic course; need for corticosteroids*	*Morpheaform*	• *Indurated, atrophic plaques* • *Linear distribution (usually on thighs)* • *Granulomas and dermal sclerosis on histopathology*
Angiolupoid	• *Type of plaque sarcoidosis* • *Large telangiectasias* • *Female predominance*	*Oral Sarcoidosis*	• *Firm nodular lesions* • *Buccal mucosa (rare on the lips)* • *Must r/o systemic sarcoidosis as this may be 1st sign*

Table 1: Select Types of Cutaneous Sarcoidosis [1,2].

Prognosis, severity, and chronicity of sarcoidosis changes markedly depend on the type of lesion. Marcoval, Mana, and Rubio analyzed 506 patients with systemic sarcoidosis [4]. 80% of the patients had skin findings prior to or at the time of systemic disease diagnosis. Persistent, life threatening illness and increased need for systemic corticosteroid therapy was more likely involved with plaque type sarcoidosis. The patients that presented with papules in the study resolved in a mean 5.44 months, most clearing spontaneously. Interestingly, although their study had low power, there was a noticeable association between lupus pernio and bone involvement seen in 50% of cases. Additionally, patients with subcutaneous sarcoidosis showed a relatively good prognosis usually resolving in a mean of 10.10 months. Erythema nodosum, although non-specific, is associated with good prognosis.

There are also specific constellations of findings that are seen in certain clinical variants of sarcoidosis. Chronic lung involvement, uveitis, and bone cysts must be considered with the appearance of violaceous infiltrative lesions of lupus pernio on the nose, cheeks and earlobes. Lofgren's syndrome, linked with acute sarcoidosis, must be considered when erythema nodosum occurs simultaneously with hilar adenopathy, acute iritis, migrating polyarthritis and fever [1]. Blau syndrome, an autosomal dominant form of sarcoidosis caused by mutations in the NOD2 gene, initially presents with a scaly or nodular granulomatous dermatitis, but continues on with a clinical course featuring arthritis, uveitis, synovial cysts and camptodactyly [5]. In conclusion, sarcoidosis can present with several clinical morphological presentations and, as such, may pose a diagnostic challenge. Herein, we reviewed the clinical cutaneous presentations

of this condition. Histopathology reveals non-caseating "naked" granulomas and is helpful in confirming the diagnosis. Given its systemic nature, diagnosis of cutaneous sarcoidosis is very important and may facilitate and guide further diagnostic workup and therapeutic planning.

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