CASE REPORT

Scar Sarcoidosis: A Rare Case

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ABSTRACT

Sarcoidosis is an inflammatory granulomatous multisystem disease. Cutaneous symptoms of sarcoidosis are infrequent, and scar sarcoidosis is very rarer. This condition may be caused by mechanical trauma such as skin wound, venipuncture, or scars caused by infection such as herpes zoster, and tattoos. We report a 17-year-old female tattoos who developed scar sarcoidosis following an old large erythematous burn scar on the extensor aspect of her left forearm extending to the hand, of 2 months duration. A large local excision was done. Histological examination was consistent with scar sarcoidosis; revealed typical non-caseating epithelioid cell granulomas.

INTRODUCTION

A 17-year-old girl presented to the dermatology OPD with the complaint of a large, asymptomatic pinkish to skin-colored lesion over her left forearm and hand of 2 months duration. Patient had a history of scalding with boiling water in this region 2 years before, that resolved leaving behind scar. Two months later, patient noticed this raised lesion appearing over the scar that gradually increased in extent.

Physical examination showed, a large, well-defined, irregular bordered erythematous to skin-colored indurated annular plaque measuring approximately 10 x 20 cm on the dorsal aspect of her left forearm and hand with some discrete small plaques (Fig. 1, 2). No tender nodules were noticed, and no regional lymphadenopathy was present. The review of other systems and a routine chest x-ray were unremarkable. Considering the clinical differential diagnosis of a scar sarcoidosis, lupus vulgaris, leishmaniasis, psoriasis, discoid lupus, granuloma annulare and tinea incognito, skin biopsy was taken.

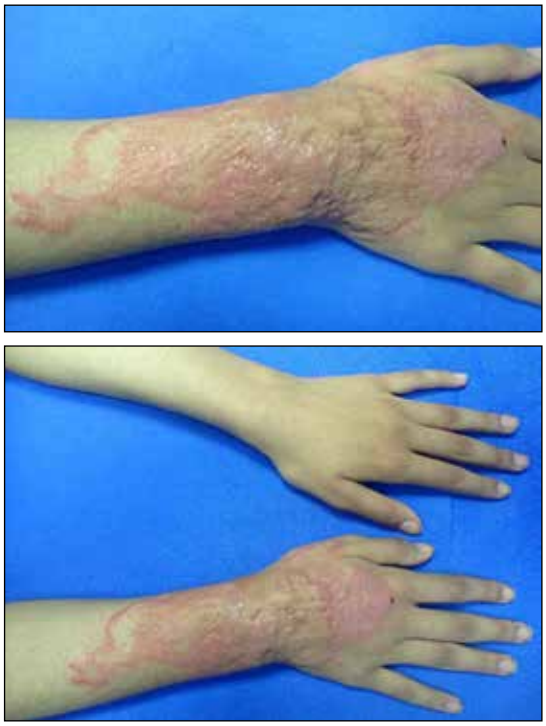

Fig. 1, 2 Erythematous to skin-colored annular plaque on top of old scar measuring approximately 10 x 20 cm on the dorsal aspect of the left forearm and hand with some discrete small plaques

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Serum angiotensin converting enzyme (ACE) levels were normal. A Mantoux test to 5 and 10 tuberculin units was negative. No hypercalcemia or hypercalciuria were detected. Other patients' routine biochemistry and radiological investigations including CBC, CPR, blood sugar, hepatic and renal profile revealed no abnormal findings. Serology for hepatitis B and C and human immunodeficiency virus were nonreactive.

An incisional skin biopsy revealed changes consistent with non-necrotizing, non-caseating granulomatous inflammatory structures comprised of epithelioid cells and Langhans giant cells with few lymphocytic infiltration within papillary and the reticular dermis (Fig. 3, 4). A Clinicopathological correlation was consistent with a diagnosis of scar sarcoidosis.

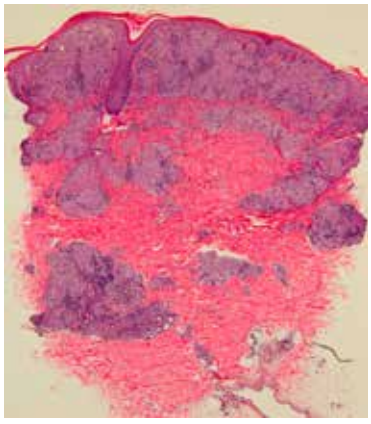

Fig. 3 Lichenoid and nodular granulomatous infiltration in the dermis

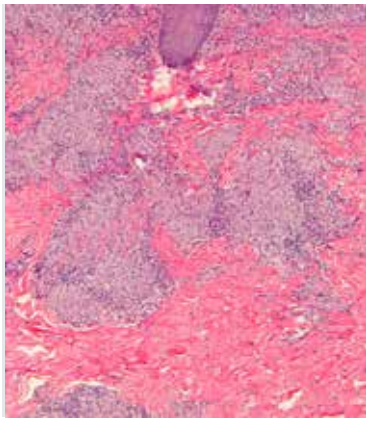

Fig. 4 Non caseatinig naked granuloma formed of epithelioid histiocytes, giant cells and few lymphocytes

The patient was treated with oral prednisolone 30 mg/day for one month, then tapered over the next month together with intralesional corticosteroid and achieved marked improvement.

FINAL DIAGNOSIS

Scar Sarcoidosis

DISCUSSION

Sarcoidosis initially was described by Sir Jonathan Hutchinson in 1875, and cutaneous sarcoidosis (lupus pernio) was described by Besnier in 1899. Sarcoidosis is a multisystem inflammatory disorder of unknown etiology with formation of non-caseating granulomas. The incidence of this disease is 10-15 cases per 100.000 per year. Broad American studies show that sarcoidosis is 10-17 times more common in African American than in Caucasians.[1] In most cases lymphadenopathy and other organs such as lung and liver are also often involved. Even destruction and deformation of bones have so far been described.[2] Cutaneous sarcoidosis is known as one of the 'great imitators' in dermatology because of different morphologies of the lesions. It occurs in up to one third of patients with systemic sarcoidosis, that is why recognition of cutaneous lesions is important because the lesions provide a visible clue to the diagnosis and are an easily accessible source of tissue for histologic examination.[3]

Scar sarcoidosis is a more rare, benign and specific variant of cutaneous sarcoidosis, representing 29% of the patients with cutaneous sarcoidosis, it has been considered to be a systemic autoimmune disease.[4-6] It arises in the area of older scars after mechanical insult that lead to a loss of skin function of the affected area, such as the sites of previous intramuscular injections, Kveim

or tuberculin test sites, sites that have received hyaluronic acid injection for wrinkles, sites of cosmetic tattoos and sites of previous laser surgery.[7,8] Scar sarcoidosis also has been reported following herpes zoster infection.[9]

Information regarding the appearance of the lesions of scar sarcoidosis differs among races, as well as among individual cases affected. It is characterized by the onset of erythematous or red-purplish swelling and the development of papules, nodules or plaque within the original scars. In cases of cutaneous or subcutaneous swelling in the area of an old scar or beside a scar, a scar sarcoidosis is a possible differential diagnosis.[2] Differential diagnoses of scar sarcoidosis include lupus vulgaris, necrobiosis lipoidica, leprosy, leishmaniasis, psoriasis, and discoid lupus. Important other differential diagnoses like a sarcoma or a keloid should be considered.[10]

Diagnosis of sarcoidosis might be challenging requires a combination of: i) Supporting clinical-radiological findings such as hilar and/or paratracheal lymph node enlargement with or without pulmonary infiltrates; ii) histological evidence of non-caseating epithelioid granulomas at disease sites; and iii) exclusion of alternative causes for the granulomatous inflammation and local sarcoid-like reactions.[11]

In case of scar sarcoidosis further investigation on systemic involvement have to be performed. It is recommended for all patients with cutaneous sarcoidosis a periodic clinical evaluation, including complete anamnesis, physical exam, serum calcium, renal and hepatic functions, radiography of the chest, pulmonary function tests, eletrocardiogram (ECG) along with ophthalmological evaluation.[12]

Histopathological examination of scar sarcoidosis revealed non-caseating epithelioid cell granulomas. Typical sarcoid lesions are characterized by the presence of circumscribed granulomas of epithelioid cells with little or no necrosis. Granulomas usually are in the superficial dermis but may involve the full thickness of the dermis and extend to the subcutaneous tissue. Islands of epithelioid cells may have a few Langerhans giant cells.[13] Giant cells may contain asteroid or Schaumann bodies; that usually are calcified at the periphery. Granulomas are referred to as naked because they have only a sparse lymphocytic infiltrate at the margins of the granulomas. Fibrosis, if present, usually starts at the periphery and advances toward the center.[14]

The cicatricial sarcoidosis typically resolves slowly and spontaneously. In the case of pure cutaneous sarcoidosis, high-potency intralesional and topical corticosteroids can be beneficial.[1] When there are disfiguring lesions non responsive to the initial topic therapy or in cases with systemic involvement, oral prednisone, hydroxychloroquine, methotrexate and allopurinol can be used.[15] In some cases, the above-mentioned treatments result in an incomplete clinical response or adverse events. In such situations tumor necrosis factor –alpha inhibitors, infliximab and adalimumab have been tried.[16] Laser therapy is a recent therapeutic approach that may give a rapid and non-invasive solution for cutaneous sarcoidosis.[15] The prognosis of cutaneous sarcoidosis depends on the systemic involvement. The course is variable, ranging from self-limited acute episodes to a chronic debilitating disease that may result in death. Spontaneous remissions occur in nearly two thirds of patients, but 10% to 30% of patients have a more chronic or progressive course.[17] Cutaneous sarcoidosis usu-

ally has a prolonged course. Papules and nodules tend to resolve over months or years, though plaques may be more resistant. As treatment is withdrawn, relapses are frequent, especially in black patients who tend to have more severe and prolonged symptoms.[18]

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