Scar Sarcoidosis with Pulmonary Involvement in a 44-year-old African woman: Case Report and Review of Literature

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Running Title: Scar Sarcoidosis with Pulmonary Involvement

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ABSTRACT

Sarcoidosis is a rare multisystem granulomatous disease of unknown aetiology that affects virtually all organs and systems, including the lungs, lymph nodes, liver, and the skin. Skin involvement is seen in about 20-35% of the patients with systemic sarcoidosis. Scar sarcoidosis is the most clinically characteristic and a rare cutaneous manifestation of sarcoidosis. It may predict systemic disease, especially pulmonary involvement. We present a case of scar sarcoidosis in a 44-year-old African woman who developed disseminated infiltrated papules, nodules and plaques on old scars from traditional marks on the face and anterior abdominal wall, and also had pulmonary involvement. Scar sarcoidosis should be considered in the differential diagnosis when there are changes to any scar tissue, and such patients should also be adequately evaluated for pulmonary involvement.

Sarcoïdose cicatricielle avec atteinte pulmonaire chez une femme africaine de 44 ans: rapport de cas et revue de la littérature

Abstrait

La sarcoïdose est une maladie granulomateuse multisystémique rare d'étiologie inconnue qui affecte pratiquement tous les organes et systèmes, y compris les poumons, les ganglions lymphatiques, le foie et la peau. Une atteinte cutanée est observée chez environ 20 à 35% des patients atteints de sarcoïdose systémique. La sarcoïdose cicatricielle est la plus caractéristique cliniquement et une manifestation cutanée rare de la sarcoïdose. Il peut prédire une maladie systémique, en particulier une atteinte pulmonaire. Nous présentons un cas de sarcoïdose cicatricielle chez une femme africaine de 44 ans qui a développé des papules, des nodules et des plaques infiltrés disséminés sur de vieilles cicatrices de marques traditionnelles sur le visage et la paroi abdominale antérieure, et qui avait également une atteinte pulmonaire. La sarcoïdose cicatricielle doit être prise en compte dans le diagnostic différentiel lorsqu'il y a des changements dans le tissu cicatriciel, et ces patients doivent également être évalués de manière adéquate pour l'atteinte pulmonaire.

Introduction

Sarcoidosis is a rare multisystem granulomatous disease of unknown aetiology that affects virtually all organ systems, including the lungs and the skin.[1] Skin involvement is seen in about 20-35% of the patients with systemic sarcoidosis.[2,3] However cutaneous sarcoidosis may occur without the systemic disease.[3] Cutaneous lesions in sarcoidosis can have varied morphological presentations, thus termed "the great imitator". Scar sarcoidosis is the most clinically characteristic and rare cutaneous manifestation of sarcoidosis, which may predict systemic invovement.[4,5]

Here, we report a case of a patient with scar sarcoidosis and associated pulmonary involvement.

Case Report

The patient is a 44-year-old female trader who presented to the dermatology clinic with a one-month history of body rashes and cough. Body rashes were first noticed on the abdomen at the site of her traditional scarification marks where her date of birth was written as 11-4-75. The rash progressed to involve her facial tribal marks and other parts of

the trunk as well as the extremities. There was associated mild pruritus but no discharge or ulceration, and the rashes were not worsened by exposure to sunlight. There was no previous history of a similar rash, no known drug allergy, no preceding history of contact with chemical irritants. About the same time, she developed an insidious cough productive of whitish mucoid sputum with no haemoptysis. However, she had been having dyspnoea on moderate exertion over two months with no orthopnoea, paroxysmal nocturnal dyspnoea or leg swelling. There was no associated fever, night sweats or weight loss. A review of the systems was essentially unremarkable. Skin examination revealed mildly scaly hypopigmented papules and plaques over the scarification/tribal marks on the face and upper anterior abdominal wall (Fig. 1). There were other isolated scaly and excoriated papules, nodules and plaques on the palms and lower extremities, especially on the extensor aspects of the knees and the lateral aspects of both feet (Fig. 1). The respiratory system examination showed features of bilateral upper-lobes consolidation. The cardiovascular, gastrointestinal and central nervous systems were essentially normal. Complete blood count and differentials were unremarkable. The erythrocyte sedimentation rate (ESR) was 49mm/hour (Westergren method). Serum biochemistry was essentially normal except for hypercalcaemia with a corrected calcium of 3.8mmol/l (normal range; 2.25-2.75mmol/l). Electrocardiogram (ECG) done was within normal limits and the sputum gene Xpert test for mycobacterium tuberculosis was negative. Chest x-ray (postero-anterior view) showed bilateral hilar lymphadenopathy with widespread peripheral nodular opacities (Fig.3). The histology result of the skin biopsy taken from the scar lesions, show areas of chronic non-caseating granulomatous inflammation within the dermis (Fig.2). Special stains were done to rule out infectious causes (Periodic acid Schiff and Ziehl Needsen tests) which were negative. The overall histological features were consistent with sarcoidosis.

A diagnosis of scar sarcoidosis with pulmonary involvement was made and the patient was placed on oral prednisolone 40mg daily for the first one month. There was remarkable clinical improvement within two weeks of commencement of oral prednisolone, as evidenced by reduced pruritus, flattening and clearing of the skin lesions and improvement in her respiratory symptoms (Fig.4). The dose of oral prednisolone was decreased gradually and stopped after four months with no relapse.

Discussion

Sarcoidosis is a rare idiopathic multisystem granulomatous disease that can affect virtually all organ systems, including the lungs, lymph nodes, liver, and, the skin etc.[1] It is a rare disease that commonly affects females around the 3rd to 5th decades of life.[1,2] Cutaneous sarcoidosis is seen in about 20-35% of patients with systemic sarcoidosis, however, cutaneous sarcoidosis may occur without the systemic disease.[2] The index case is a female in her 5th decade of life that presented with both cutaneous and pulmonary involvement. Cutaneous lesions in sarcoidosis are broadly classified as either specific or nonspecific based on the presence or absence of non-caseating granuloma in histology.[2,3] The index case presented with specific skin lesions of scar sarcoidosis that were confirmed by histology. This form of cutaneous manifestation is the most clinically characteristic and a rare cutaneous manifestation of sarcoidosis.[4] The most common types of specific lesions are indurated papules with varying colours ranging from red, reddish-brown, violaceous, translucent, or hyperpigmented, which are commonly seen on the face but may occur anywhere in the body.[3] The index case presented with mainly indurated hypopigmented papules nodules and plaques. The specific sarcoid skin lesions are more chronic and usually resolve with scarring,[6] as seen in the index patient. The non-specific skin lesions are mostly seen with the acute presentation of sarcoidosis and have a good prognosis.[2] Erythema nodosum is the most common non-specific cutaneous sarcoid lesion.[2]

This patient presented with widespread cutaneous sarcoidosis lesions and also symptoms and signs suggestive of pulmonary involvement (cough, dyspnea on exertion and abnormal chest radiograph features of pulmonary infiltrates, hilar and mediastinal lymphadenopathy). Systemic sarcoidosis commonly involves the pulmonary system, and this is seen more with scar sarcoidosis.[5]

Widespread cutaneous sarcoidosis lesions may predict pulmonary involvement,[5] as is the case with our patient. Furthermore, pulmonary involvement is more common in patients with lupus pernio and scar sarcoidosis than in patients with other forms of cutaneous sarcoidosis.[7] The index case has scar sarcoidosis originating from the sites of scarification secondary to skin trauma inflicted 44 years ago. In addition to the reactivation of old scars, our patient had other widespread specific cutaneous lesions of sarcoidosis that include hypopigmented papules, nodules, and plaques. Scar sarcoidosis has been reported in areas of reactivation of traumatic scars, sites of intramuscular injections, tattoos, venepuncture, healed herpes zoster lesions, ritual scarification, and desensitization injections.[5] The predisposing factor reported causing reactivation of previous scars includes previous contamination of old scars with foreign bodies at the time of trauma, with a latency period before the reactivation of old cutaneous scars of between 6 months and 59 years.[8,9] Previous studies have reported that 30% of patients with only cutaneous lesions progress to develop systemic involvement after a period of 1 month to 1 year.[5]

The diagnosis of cutaneous sarcoidosis is made when a compatible clinical or radiologic picture is present, along with histologic evidence of non-caseating granulomas, after excluding other potential causes, such as infections.[2] Histology is the hallmark for making a definitive diagnosis.[3] The index patient in this report had histological features suggestive of sarcoidosis, and other special stains were done to rule out deep fungal and mycobacterial infections. However, other laboratory investigations of value in evaluating patients with sarcoidosis include; serum biochemistry with hypercalcaemia and the elevation of serum transaminases.[5] The index case had hypercalcaemia. The serum angiotensin-converting enzyme (ACE) may also be elevated in sarcoidosis and may be a measure of granuloma volume.[5] However, this was not done in our patient due to unavailability in our setting. The fact that cutaneous sarcoidosis is a "great imitator" and a diagnosis of exclusion, other possible differential diagnoses ought to be excluded such as; infectious skin diseases like mycobacterial, fungal and spirochete infections, Crohn's disease, rosacea, foreign body granuloma, and keloid scarring. Periodic acid Schiff and Ziehl-Neelsen stains were both negative, excluding the possibility of fungal and mycobacterial infections.

Treatment of cutaneous sarcoidosis is often difficult with a high rate of recurrence as well as many refractory lesions.[2] The choice of treatment and prognosis of cutaneous sarcoidosis depends on the degree of systemic involvement.[5] Both topical and systemic corticosteroid therapy may be used.[4] Topical therapy is usually reserved for isolated skin lesions.[5] When the skin lesions are widespread or unresponsive to topical therapy or cases with systemic involvement, systemic immunosuppressive agents are employed. These include systemic corticosteroids, hydroxychloroquine, methotrexate, tetracyclines, isotretinoin, pentoxifylline, allopurinol, vitamin D, thalidomide, azathioprine, cyclophosphamide, mycophenolate mofetil, and tumour necrosis factor (TNF)-α inhibitors (e.g. etanercept).[10] Systemic corticosteroids, antimalarials (hydroxychloroquine), and methotrexate are the most widely accepted standard therapies for sarcoidosis.[11] Systemic corticosteroids are considered to be the most effective agents,[3] and are commonly used at slow, tapering dosages, starting from 20 to 40 mg of oral prednisone daily for four to six weeks.[3] However, its efficacy is limited by the adverse effects of chronic steroid use coupled with the relapse of symptoms after discontinuation of therapy.[3] The index patient was treated with systemic corticosteroids and good clinical response was noticed within two weeks. We are yet to notice any flare of symptoms in this patient since the discontinuation of systemic corticosteroid therapy.

Conclusion

Scar sarcoidosis that develops on scarification marks is a rare finding and a predictor of systemic disease, especially pulmonary sarcoidosis. Thus, all individuals presenting with scar sarcoidosis should be adequately evaluated for systemic involvement, especially pulmonary disease.

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Conflicting Interest: None

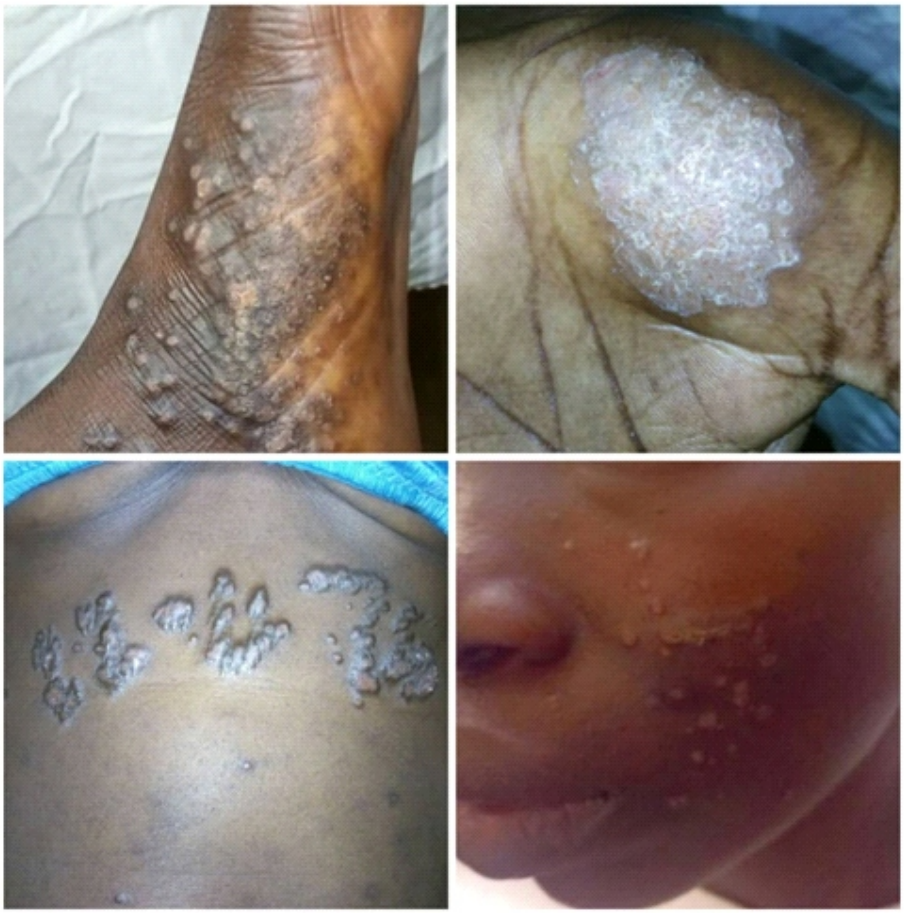

Fig.1: Showing mildly scaly hypo/hyperpigmented papules and plaques on the scarification/tribal marks, palms and lower extremities before treatment

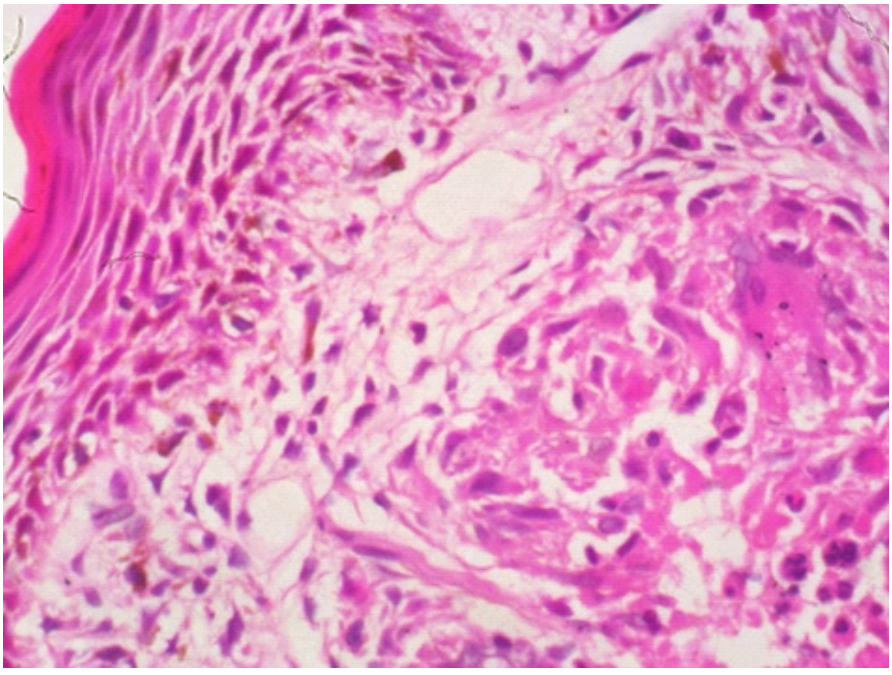

Fig.2: Showing chronic non-caseating granulomatous inflammation within the dermis, H and E, ×200

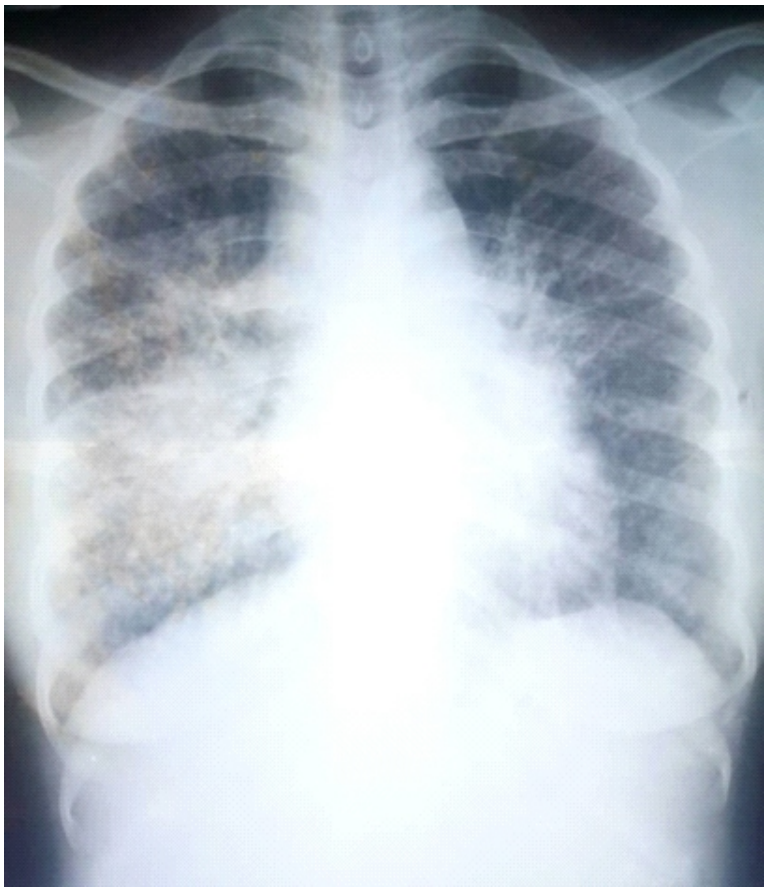

Fig.3: Chest x-ray (posterior anterior view) showing bilateral hilar lymphadenopathy with widespread peripheral nodular opacities

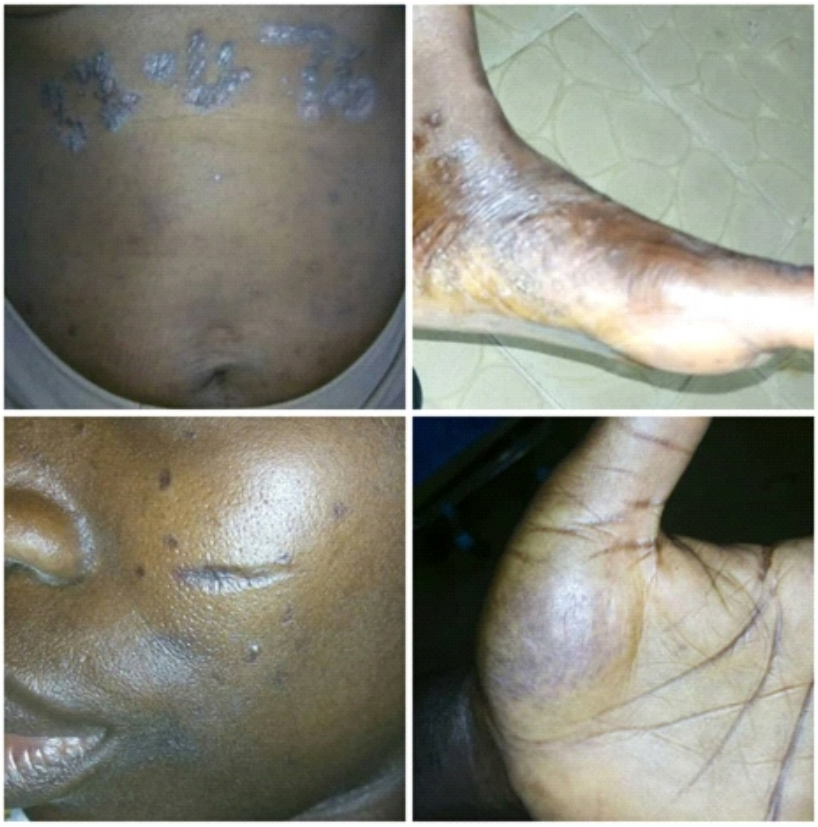

Fig.4: Showing the lesions after treatment

References

1. Éksarenko OV, Kharlap SI, Safonova TN. Sarcoidosis. Vestn Oftalmol 2012,128(3):42–48.
2. Kang MJ, Kim HS, Kim HO and Park YM. Cutaneous Sarcoidosis Presenting as Multiple Erythematous Macules and Patches. Ann Dermatol 2009;21(2):168–70.
3. Katta R. Cutaneous Sarcoidosis: A Dermatologic Masquerader. Am Fam Physician 2002;65:1581-4.
4. Hong YC, Na DJ, Han SH, Deok lee Y, Cho YS, Han MS. A case of scar sarcoidosis. Koren J Intern med 2008;23(4):213-5.
5. Su O, Onsun N, Topukçu B, Özçelik HK, Çakıter AU, Büyükpınarbaşılı N. Disseminated scar sarcoidosis may predict pulmonary involvement in sarcoidosis. Acta Dermatovenerol APA 2013;22:71-4.
6. Marchell RM, Judson MA. Chronic cutaneous lesions of sarcoidosis. Clin Dermatol 2007;25:295–302.
7. Yanardag H, Pamuk ON, Karayel T. Cutaneous involvement in sarcoidosis: analysis of the features in 170 patients. Respir Med. 2003;97:978-82.
8. Caro I. Scar sarcoidosis. Cutis. 1983;32:531-3.
9. Payne CM, Thomas RH, Black MM. From silica granuloma to scar sarcoidosis. Clin Exp Dermatol. 1983;8:171-5.
10. Beegle SH, Barba K, Gobunsuy R, Judson MA. Current and emerging pharmacological treatments for sarcoidosis: a review. Drug Des Devel Ther 2013;7:325- 38.
11. Doherty CB, Rosen T. Evidence-based therapy for cutaneous sarcoidosis. Drugs. 2008;68:1361-83.