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Disseminated scar sarcoidosis may predict pulmonary involvement in sarcoidosis

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Abstract

Sarcoidosis is a chronic, inflammatory, multi-organ disease of unknown origin that is characterized by non-caseating granuloma formation in affected organs. Cutaneous involvement is reported in 25% of patients with sarcoidosis. Scar sarcoidosis is rare but is clinically specific for skin sarcoidosis. Systemic involvement is seen in most patients with scar sarcoidosis. We present a case of scar sarcoidosis in a 30-year-old male that developed infiltrated nodules on old scars, including on his penile shaft, which is rare, and that also had pulmonary involvement. Scar sarcoidosis should be considered in the differential diagnosis of changes in all scar areas and should be investigated for systemic involvement.

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Introduction

Sarcoidosis is a chronic inflammatory and granulomatous disorder. The etiology of this disease is unknown. It affects multiple organs, primarily the skin, lungs, eyes, bones, and lymph nodes (1). Scar sarcoidosis at the sites of old scars is a rare form of cutaneous sarcoidosis (2). Systemic involvement is seen in most patients with scar sarcoidosis (3). If clinicians are unaware of the changes in old scars, scar sarcoidosis may be underdiagnosed. Histopathologic examination of a punch biopsy, taken from an appropriate lesion, usually provides the correct diagnosis.

Case

A 30-year-old male presented with a 1-year history of swelling and redness on numerous old scars from childhood injuries. There were no other constitutional symptoms such as fever, dyspnea, cough, night sweats, or weight loss. A physical examination revealed erythematous, violaceous, firm, non-tender plaques and nodules at old scar sites on the right upper lip, right side of the chin, right palmar area, and penile shaft (Fig. 1). A punch biopsy taken from the lesion on the chin showed numerous, non-caseating naked granulomas with multi-nucleated giant cells (Fig. 2). No foreign body was observed in a microscopic examination with polarized light. Periodic acid-Schiff and Erlich–Ziehl–Neelsen staining were negative. Fungal and mycobacterial cultures of skin biopsies did not demonstrate any fungal organisms or acid-fast bacilli. Routine laboratory measurements, including complete blood count, differential cell count, erythrocyte sedimentation rate, liver and renal function tests, urinalysis, and serum calcium levels were normal. VDRL (Venereal Disease Research Laboratory) and TPHA (*Treponema pallidum* hemagglutination assay) results were negative. The patient's serum angiotensin-converting enzyme (ACE) level was elevated (152 IU/L; normal range 8–52 IU/L). A tuberculin skin test yielded a result of 3 mm. Hand and foot X-rays were normal. Chest radiography showed enlarged hilar lymph nodes in the mediastinum (Fig. 3a). Pulmonary function tests revealed moderate obstructive abnormalities. High-resolution CT (HRCT) showed bilaterally enlarged hilar and mediastinal lymph nodes. Sputum was negative for acid-fast bacilli. An ophthalmoscopic examination revealed normal ocular findings. The patient refused to undergo bronchoscopy. Based on the results of clinical, histopathological, and laboratory evaluations, scar sarcoidosis with pulmonary involvement was diagnosed. High-potency topical corticosteroid was started as a first line-treatment, but no significant change in the lesions was observed. We therefore switched the patient to oral methylprednisolone treatment (40 mg per day). From 1 month, the dose of methylprednisolone was decreased gradually and was stopped after 4 months. We observed complete regression of previously visible induration of the scars (Fig. 4). A chest X-ray showed complete regression in the mediastinal lymph nodes (Fig. 3b). Pulmonary function tests revealed mild obstructive abnormalities.

Discussion

Sarcoidosis is a chronic multi-organ disorder of unknown etiology that is characterized by non-caseating granulomas in the affected organs (1). Cutaneous manifestations are relatively uncommon (20–35%) (4, 5). They are of two groups: specific and non-specific. Specific cutaneous lesions present as macules, papules, nodules, plaque, subcutaneous nodules, infiltrative scars, and lupus pernio (3). Non-specific lesions are erythema nodosum (EN), calcification, erythema multiforme, prurigo, nail clubbing, and Sweet's syndrome (6). Because skin sarcoidosis has many variants, the diagnosis of cutaneous sarcoidosis is difficult. To establish the diagnosis, all clinical, histopathological, and laboratory findings should be evaluated simultaneously.

In one study, the lesions of 29% of patients with cutaneous sarcoidosis started as cicatricial lesions (7). Yanardag et al. reported that scar lesions were observed in 2.9% of patients that were previously diagnosed with sarcoidosis (8). The main difference between these two studies was whether or not the scar sarcoidosis was accompanied by other cutaneous sarcoidosis lesions. Scar sarcoidosis is very rare, but it is highly specific for skin sarcoidosis in which old scars are infiltrated by non-caseating granulomas (2, 8).

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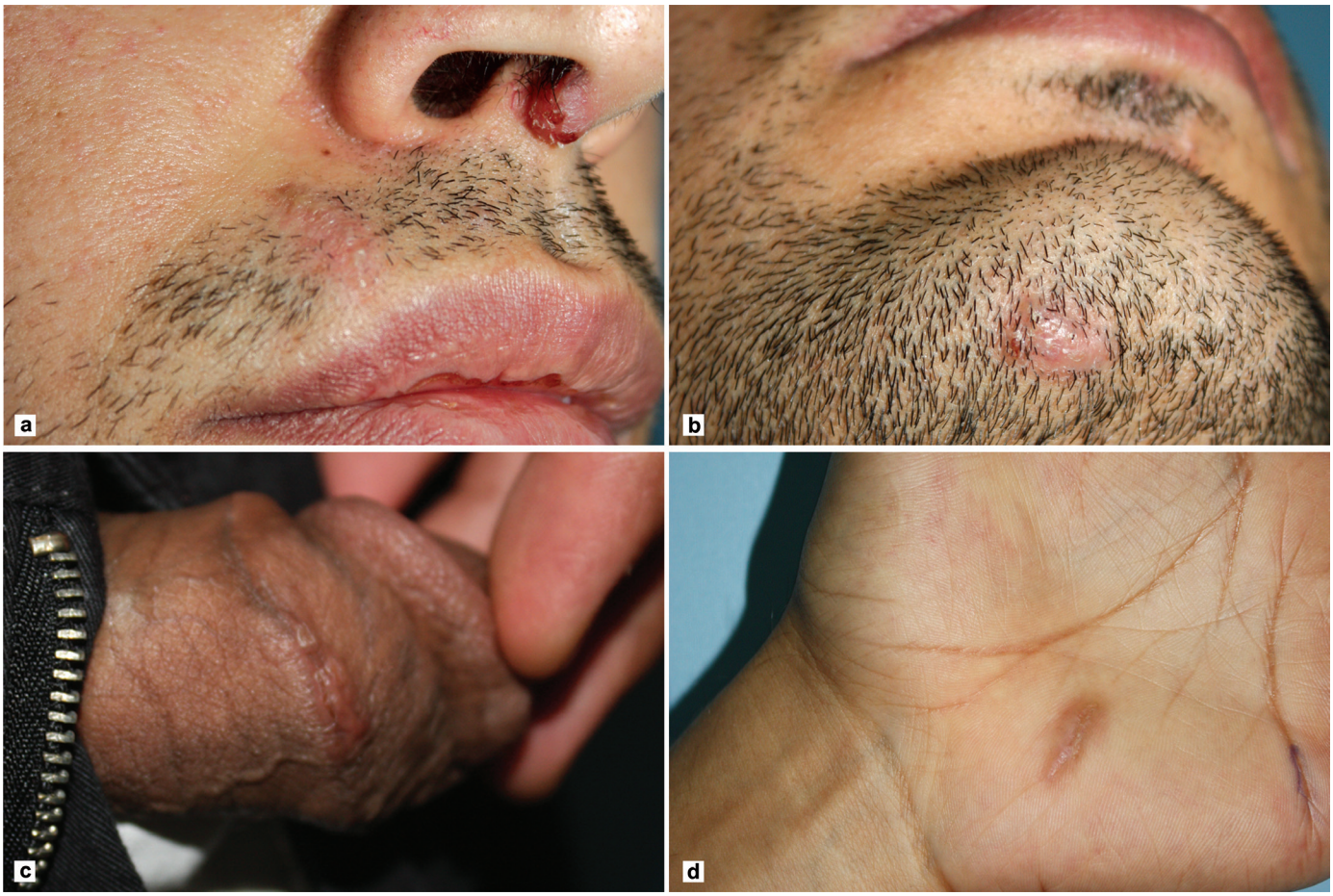

Figure 1 | Scar sarcoidosis. a) upper lip, b) chin, c) penile shaft, d) palmar area.

In addition to reactivation of the scars in traumatic scar areas, scar sarcoidosis has been reported at the sites of intramuscular injections, tattoos, venepuncture, healed herpes zoster lesions, ritual scarification, and desensitization injections (9–11). It has been suggested that previous contamination of the old scars with foreign bodies at the time of trauma may be a cause of scar sarcoidosis (12). Foreign body granuloma might be considered in the differential diagnosis of scar sarcoidosis. The search for foreign bodies by microscopic examination with polarised light is important for the diagnosis of scar sarcoidosis.

Pulmonary involvement is known to be more frequent in patients with lupus pernio and scar sarcoidosis than in patients with other skin sarcoidosis variants (8). Scar sarcoidosis may predict systemic involvement, especially pulmonary involvement. Scar sarcoidosis without other skin lesions is uncommon (8, 13). The reported latency period before reactivation of old cutaneous scars is between 6 months and 59 years (14).

Differential diagnosis of scar sarcoidosis includes infectious skin diseases such as mycobacterium and spirochete infections, Crohn's disease, rosacea, foreign body granuloma, and keloid scarring. Our patient had no cutaneous manifestations of sarcoidosis other than reactivation of old scars. Syphilis is an important entity in differential diagnosis because our patient had an erythematous, indurated raised scar on his penile shaft. The patient stated that the lesion was a traumatic scar that appeared after a childhood injury. In addition, there was reactivation of all old cutaneous scars. Serologic tests for syphilis were negative.

Mana et al. reported that 30% of patients with only cutaneous lesions developed systemic involvement after a period of 1 month to 1 year (15). Scar sarcoidosis is often associated with erythema nodosum and hilar and generalized lymphadenopathy (14, 15). We detected skin findings of scar sarcoidosis along with hilar and mediastinal lymphadenopathy in our patient.

Sarcoidosis has an extremely heterogeneous clinical picture, and is thus defined as "the great imitator." Awareness of alterations of old scar areas is important. If scar sarcoidosis is suspected and a punch biopsy is taken from the reactivated old scar lesion, it may be easily diagnosed by histopathological examination.

The treatment and prognosis of skin sarcoidosis are primarily related to the degree of systemic involvement (16). Topical steroid therapy may sometimes be effective for isolated skin sarcoidosis (17). For resistant lesions that are unresponsive to topical therapy and cases of systemic involvement, systemic corticosteroids, hydroxychloroquine, methotrexate, tetracyclines, isotretinoin, pentoxifylline, allopurinol, vitamin D, thalidomide, azathioprine, cyclophosphamide, mycophenolate mofetil, and tumor necrosis factor (TNF)-α inhibitors may be given (6, 17, 18). Our patient responded well to systemic corticosteroid treatment. In spite of the fact that the patient did not undergo bronchoscopy with transbronchial biopsy and histopathological examination of the biopsy, we concluded that there was pulmonary involvement because of complete regression in the mediastinal lymph nodes in chest X-ray and improvement in pulmonary function test results after systemic corticosteroid treatment.

We report this case because it is a rare case of scar sarcoidosis with pulmonary involvement that developed on an old penile scar and other scar areas. We conclude that scar sarcoidosis should be considered in the differential diagnosis of alterations in traumatic and/or scar areas and should be investigated for systemic involvement, especially pulmonary involvement, after histopathological confirmation of the disease.

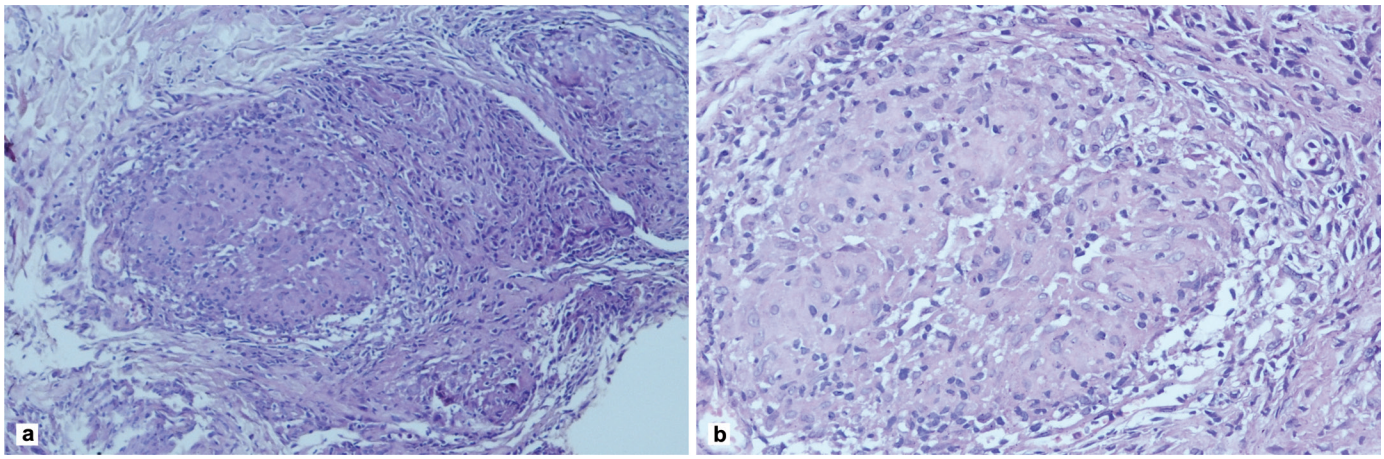

Figure 2 | Naked granuloma located in the dermis. (HE × 100).

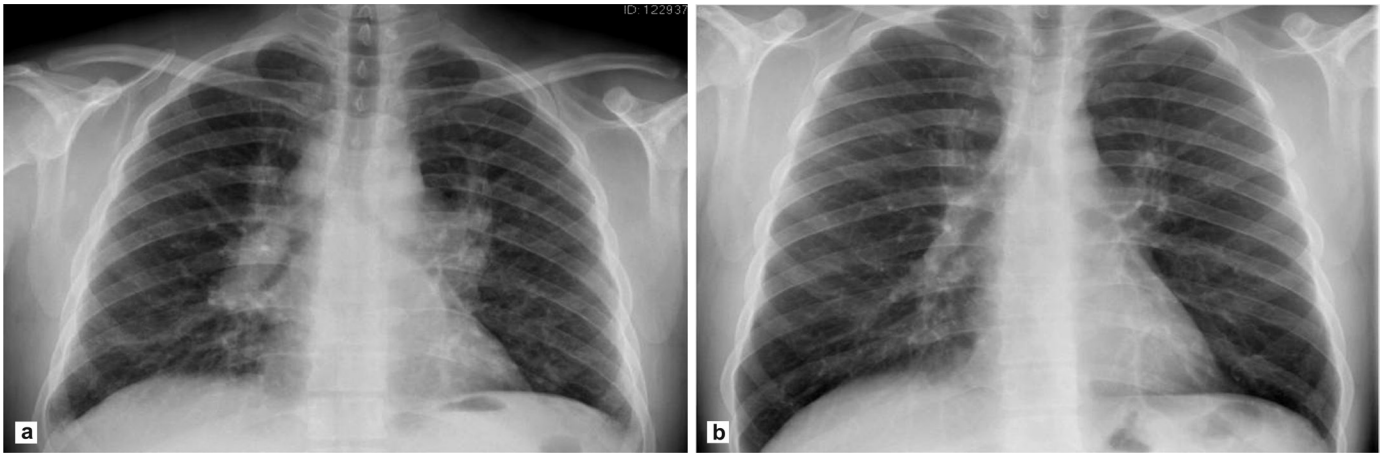

Figure 3 | a) Chest radiogram showing bilateral hilar enlargement (before treatment). b) Chest radiogram showing regression of hilar lymph nodes (after treatment).

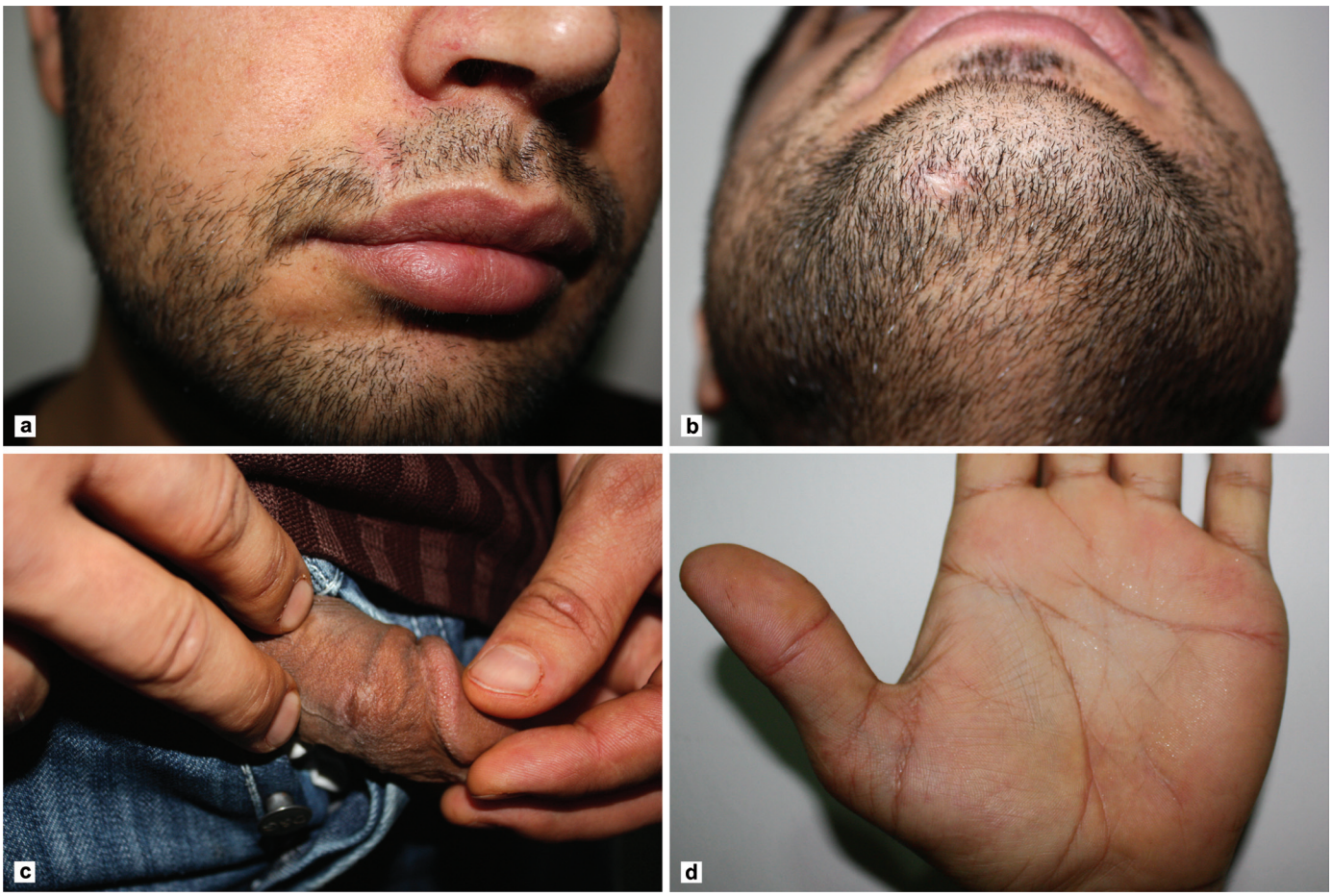

Figure 4 | Scar sarcoidosis after treatment. a) upper lip, b) chin, c) penile shaft, d) palmar area.

References

1. Mahajan VK, Sharma NL, Sharma RC, Sharma VC .Cutaneous sarcoidosis: clinical profile of 23 Indian patients. Indian J Dermatol Venereol Leprol. 2007;73:16-21.
2. Manz LA, Rodman OG. Reappearance of quiescent scars. Sarcoidosis. Arch Dermatol. 1993;129:105-8.
3. Chudomirova K, Velichkova L, Anavi B, Arnaduova M. Recurrent sarcoidosis in skin scars accompanying systemic sarcoidosis. J Eur Acad Dermatol Venereol. 2003;17:360-1.
4. Hanno R, Needelman A, Eiferman RA, Callen JP. Cutaneous sarcoidal granulomas and the development of systemic sarcoidosis. Arch Dermatol. 1981;117:203-7.
5. Samtsov AV. Cutaneous sarcoidosis. Int J Dermatol. 1992;31:385-91.
6. Lodha S, Sanchez M, Prystowsky S. Sarcoidosis of the skin: a review for the pulmonologist. Chest. 2009;136:583-96.
7. Marchell RM, Judson MA. Chronic cutaneous lesions of sarcoidosis. Clin Dermatol. 2007;25:295-302.
8. Yanardag H, Pamuk ON, Karayel T. Cutaneous involvement in sarcoidosis: analysis of the features in 170 patients. Respir Med. 2003;97:978-82.
9. Corazza M, Bacilieri S, Strumia R. Post-herpes zoster scar sarcoidosis. Acta Derm Venereol. 1999;79:95.
10. Alabi GO, George AO. Cutaneous sarcoidosis and tribal scarifications in West Africa. Int J Dermatol. 1989;28:29-31.
11. Healsmith MF, Hutchinson PE. The development of scar sarcoidosis at the site of desensitization injections. Clin Exp Dermatol. 1992;17:369-70.
12. Caro I. Scar sarcoidosis. Cutis. 1983;32:531-3.
13. Sorabjee JS, Garje R. Reactivation of old scars: inevitably sarcoid. Postgrad Med J. 2005;81:60-1.
14. Payne CM, Thomas RH, Black MM. From silica granuloma to scar sarcoidosis. Clin Exp Dermatol. 1983;8:171-5.
15. Mana J, Marcoval J, Graells J, Salazar A, Peyri J, Pujol R. Cutaneous involvement in sarcoidosis. Relationship to systemic disease. Arc Dermatol. 1997;133:882-8.
16. Hong YC, Na DJ, Han SH, Lee YD, Cho YS, Han MS. A case of scar sarcoidosis. Korean J Intern Med. 2008;23:213-5.
17. English JC 3rd, Patel PJ, Greer KC. Sarcoidosis. J Am Acad Dermatol. 2001; 44: 725-43.
18. 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.