A Curious Case of Resurgence of Old Scars with Pulmonary Involvement

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ABSTRACT

Sarcoidosis is a chronic granulomatous disorder of unknown etiology with multisystemic involvement and myriad clinical manifestations. Scar sarcoidosis is a rare but is a specific form of cutaneous sarcoidosis. Majority of the patients present with systemic disease especially pulmonary involvement. High index of suspicion is required because the diagnosis is often missed. We report a case of a 40-year-old woman presenting with pulmonary involvement and hilar and mediastinal lymphadenopathy with skin lesions that were treated as keloid from elsewhere.

Keywords: Granuloma, keloid, pulmonary involvement, scar sarcoid

Introduction

Sarcoidosis is a chronic granulomatous disorder of unknown etiology characterized by non-caseating granulomas primarily affecting lungs, mediastinal and peripheral lymph nodes, eyes, and skin. Infiltration of old scars with sarcoidal granulomas is a rare but clinically specific manifestation.[1] Most patients with scar sarcoidosis have systemic involvement at the time of diagnosis.[2]

Case

A 40-year-old woman presented with swelling of pre-existing scars for the last 6 months, a 15-year-old ligation scar, and another post-traumatic scar over the left eye acquired 30 years back [Figure 1]. Additional skin lesions were present over alae of nose and left tragus. She had been treated in another clinic with the diagnosis of keloid with intralesional triamcinolone acetonide which improved to a certain extent but gradually she developed new lesions on other sites as well. When she came to us the patient also reported grade 2 dyspnea, hurrying on the level or up a slight hill according to the Medical Research Council dyspnea scale (MRC dyspnea scale).[3] There was no history of fever, cough, night sweats, and weight loss. Cutaneous examination showed skin-colored papules, nodules, plaques overlying old scars, and other sites such as alae of nose and left tragus. Systemic examination was unremarkable. A punch biopsy was taken from a nodule overlying the ligation scar over abdomen, which on hematoxylin and eosin (HE) staining revealed non-caseating epithelioid granulomas with numerous multinucleated giant cells of Langhan's type and foreign body type with occasional lymphocytes at the periphery situated throughout the dermis. There were haphazardly arranged thick collagen bundles in the dermis suggestive of scar tissue [Figure 2]. Periodic acid Schiff and Ziehl Neelsen staining were negative. Based on the clinical and histopathological findings, the diagnosis of scar sarcoidosis was made. To rule out any possible systemic involvement which may occur, further diagnostic workup was done. Routine blood parameters including serum calcium were normal.

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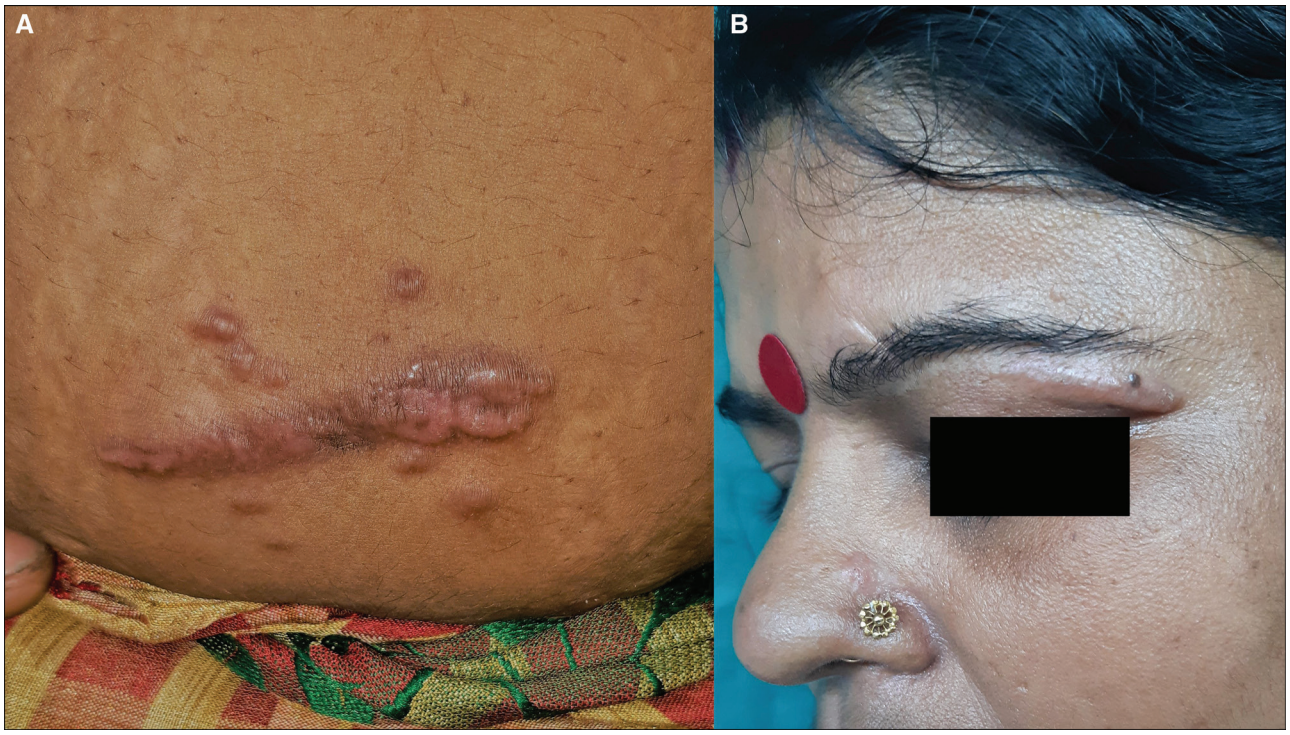

Figure 1: Scar sarcoidosis. (A) Lesion over post ligation scar. (B) Lesion over traumatic scar in left upper eyelid and alae of nose

Serum angiotensin-converting enzyme was elevated, 135 IU/L (normal range, 8–52 IU/L). Mantoux test was negative. Sputum smear examination was negative for acid-fast bacilli. On chest X-ray (CXR), there was left-sided hilar prominence. On high-resolution computed tomography (HRCT), there was subcarinal and left lower paratracheal lymphadenopathy, also parenchymal lesions in the form of bilateral peribronchovascular nodules [Figure 3]. Ophthalmoscopic and slit-lamp examination revealed normal findings. Electrocardiogram and abdominal ultrasound test was normal. The patient was referred to respiratory medicine department for further investigations. She did not give consent for bronchoscopy and transbronchial procedures. According to "Scadding criteria" which are based on chest radiograph findings, the diagnosis of Stage 2 pulmonary sarcoidosis was made as there was hilar lymphadenopathy as well as parenchymal involvement.[4] However, nowadays HRCT is being increasingly used over CXR for initial evaluation of patients as the scans reveal imaging abnormalities not evident on CXR, including prominent lymph nodes, subtle parenchymal involvement, or minimal fibrosis.[5] The patient was initially started on topical clobetasol propionate ointment and hydroxychloroquine 200 mg OD and after 7 days once the diagnosis of pulmonary sarcoidosis was made, prednisolone was added to this existing regimen in a dose of 40 mg daily. There was significant improvement of skin lesions at the end of 4 weeks of therapy [Figure 4] and the patient reported improvement in dyspnea symptoms, which was only with strenuous exercise (MRC Grade 1).

Discussion

Cutaneous sarcoidosis has myriad clinical presentations and majority of them present with lupus pernio, maculopapular eruption, subcutaneous nodules, and erythema nodosum.

Reactivation of old scars in the form of scar sarcoidosis is a rare but specific manifestation of cutaneous sarcoidosis.[6] Although the exact incidence is unknown, it is reported to vary from 2.9% to 13.8% in series of adult cutaneous sarcoidosis.[7] Scar sarcoidosis may present with pruritus, erythema, elevation, induration, or increase in size of an older quiet scar. In addition to involvement of old scars, skin lesions may be present on other sites as well. Other than post-traumatic and postsurgical scars, scar sarcoidosis has been reported to occur in previous tattoo scars, intramuscular injection sites, venepuncture sites, scars of herpes zoster, following hyaluronic acid injection and laser surgery.[8] The age of scar is not that important as it has been reported to occur in relatively new onset (6 months) as well as very old scars (59 years).[9] Scar sarcoidosis may predict systemic involvement, especially pulmonary involvement.[10] It may be the presenting symptom and also a sign of recurrence of systemic disease in patients on remission. Pulmonary involvement is known to be more frequent in patients with lupus pernio and scar sarcoidosis than in patients with other skin sarcoidosis variants. Scar sarcoidosis may also be associated with erythema nodosum, hilar and generalized lymphadenopathy.[11] In our case also scar sarcoidosis paved way for the diagnosis of internal disease of which she was unaware of despite

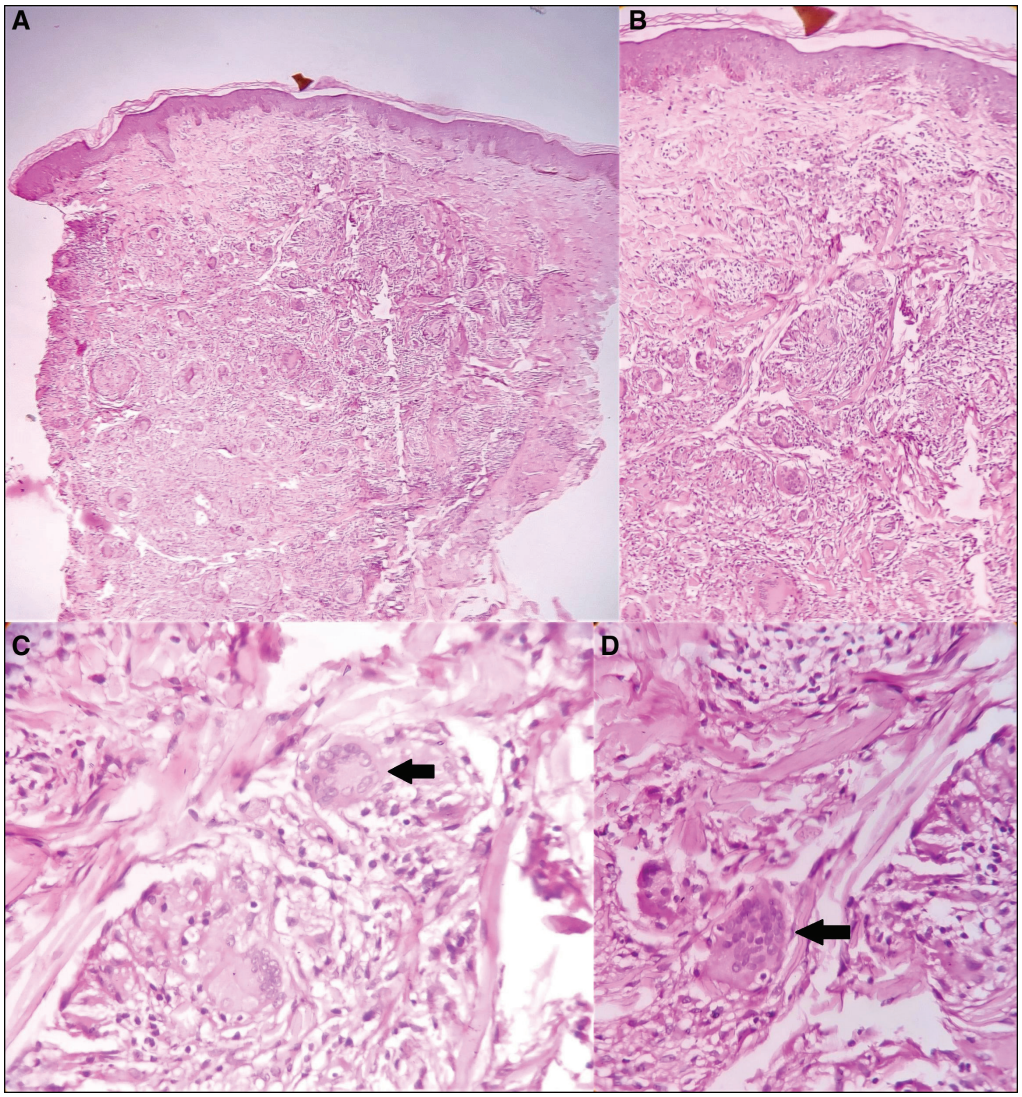

Figure 2: Multiple non-caseating epithelioid granulomas throughout the dermis. (A) 40× HE view. (B) 100× HE view of upper and mid dermis. (C) Langhans giant cell. (D) Foreign body giant cell and haphazardly arranged collagen fibers in the dermis, 400× HE view

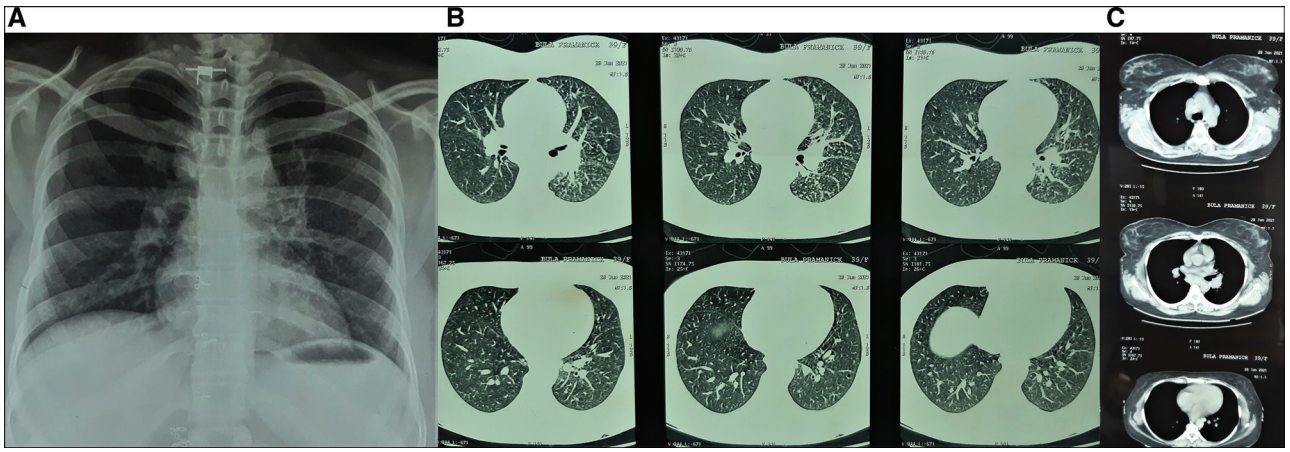

Figure 3: (A) A chest X-ray showing hilar widening on left side. (B) A high-resolution computed tomography (HRCT) (parenchymal view) showing peribronchovascular nodules. (C) HRCT showing hilar and mediastinal lymphadenopathy

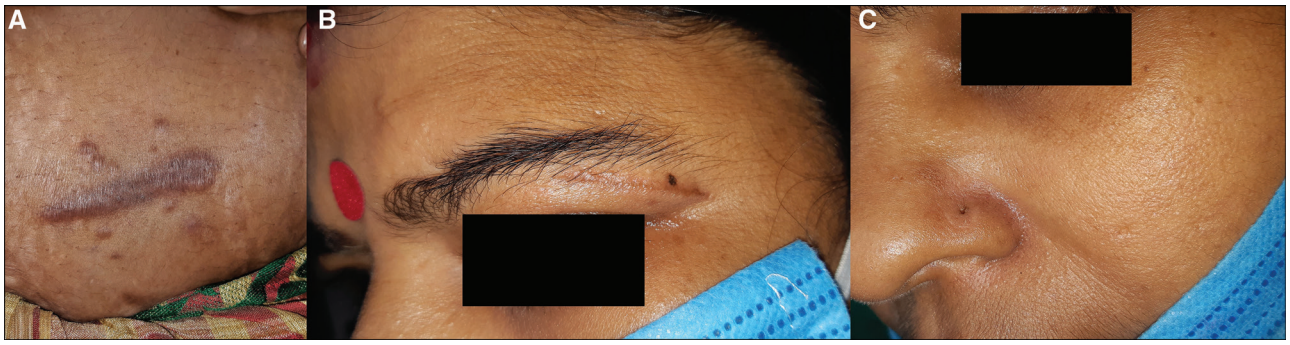

Figure 4: Four weeks after treatment images. (A) Flattening of lesion over abdomen. (B and C) Disappearance of lesions over left upper eyelid and alae of nose

Table 1: Therapeutic options for the management of sarcoidosis[14]

Organ	Manifestations	Treatment	
		Drug of choice	Alternatives
Cutaneous	Localized lesion(s)	Intralesional corticosteroids Potent topical corticosteroids	Oral corticosteroids (prednisolone 20–40 mg/day) Methotrexate, azathioprine, hydroxychloroquine, mycophenolate mofetil, infliximab, adalimumab, and tetracyclines.
	Diffuse lesions	Oral corticosteroids (prednisolone 20–40 mg/day)	Methotrexate, Azathioprine, Hydroxychloroquine, Mycophenolate mofetil, Infliximab, Adalimumab, Tetracyclines.
	Lupus pernio: disfiguring facial sarcoidosis	Oral corticosteroids	Infliximab, methotrexate, azathioprine, hydroxychloroquine, mycophenolate mofetil, and adalimumab.
Extracutaneous		Oral corticosteroids	Infliximab, methotrexate, azathioprine, hydroxychloroquine, mycophenolate mofetil, and adalimumab.

having exertional dyspnea. Thus every patient with scar sarcoidosis requires an initial workup followed by periodic screening for systemic involvement.

The principal differential diagnoses of scar sarcoidosis are hypertrophic scar/keloid and foreign body granuloma. In our case also the diagnosis was misleading as keloid therefore further investigations were not performed in previous clinic and internal organ involvement was not revealed. Other diagnoses to be considered are mycobacterial infection, Crohn's disease, and deep fungal infections like sporotrichosis.[12] Although presence of scar tissue along with non-caseating tuberculoid granulomas is quite characteristic of scar sarcoidosis, other histopathological differential diagnosis to be considered are noninfectious granulomas like foreign body granuloma, interstitial granulomatous dermatitis, and infectious granulomas such as cutaneous tuberculosis, atypical mycobacterial infection, and Hansen's disease.[13]

Like other types of skin sarcoidosis, treatment and prognosis of scar sarcoidosis is dependent on presence of systemic involvement [Table 1]. For isolated cutaneous lesions ultrapotent class 1 topical corticosteroid is the first-line treatment. For lesions not responding to topical therapy and cases having systemic involvement, systemic corticosteroids, hydroxychloroquine, methotrexate, tetracyclines, isotretinoin, thalidomide, azathioprine, cyclophosphamide, mycophenolate mofetil, and tumor necrosis factor α inhibitors may be given.[14] Our patient responded well to hydroxychloroquine and prednisolone and there was appreciable improvement of skin lesions and exertional breathlessness (from MRC grade 2 to grade 1) after 4 weeks of therapy and she is currently under the same treatment.

Conclusion

High index of suspicion is required for the diagnosis of scar sarcoidosis as the diagnosis is often missed. In the case of reactivation of any old scar, diagnosis of scar sarcoid should be kept in mind. It also provides us a visible clue of systemic involvement and allows an easily accessible site for biopsy and HPE which negates the need for invasive investigations like bronchoscopy and transbronchial biopsy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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