

Case Report

Scar sarcoidosis: additional involvement of *de novo* sites and update on immunopathogenesis

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ABSTRACT

Cutaneous sarcoidosis is an immune-mediated multisystem granulomatous inflammatory disease of unknown etiology. Noncaseating granuloma in pre-existing scars is an uncommon but specific cutaneous manifestation of sarcoidosis. This great imitator may be an under-diagnosed disease in developing countries like India probably because of the diverse range of clinical presentations. Herein we present a 45-year old female with cutaneous sarcoidosis involving scars in addition to de-novo sites.

Keywords: Sarcoidosis, Noncaseating granuloma, Great imitator, Scar

INTRODUCTION

Besnier reported the first case of sarcoidosis in 1889 while the first case in India was reported by Rajam et al. in 1957.¹ The origin of the word "Sarcoid" is Gr. Sark=Flesh, oid=like. In India, the exact prevalence of sarcoidosis is not known. It has been estimated that sarcoidosis constituted 10 to 12 cases per 1,000 new registrations in a respiratory unit at Kolkata and 61.2/100,000 new cases at a center in New Delhi in a report published in 2002.² Sarcoidosis is an immune-mediated inflammatory disease that manifests as non-caseating epithelioid granulomas, the development of which is attributed to a number of environmental factors and genes. The disease predominantly affects the lungs, lymph nodes, also eyes and skin, peripheral lymph nodes and liver. It is more common in middle-aged females and was earlier it was found to be predominant in the western countries.³ Herein we report a rare presentation of sarcoidosis occurring in old and recent scars as well as *de novo* and discuss various current clinico-immunological

aspects along with possible outcomes and appropriate management.

CASE REPORT

A 45-year-old female presented with multiple asymptomatic erythematous lesions over the face, neck and external genitalia, gradually increasing in number and size since 2 months. She denied history of fever, constitutional symptoms, musculoskeletal symptoms, cough, dyspnea etc. History of atrophic scar at sutured contused lacerated wound over philtrum area (post road traffic accident 3 years ago) was elicited. Similar lesions appeared on a 7 year old posterolateral episiotomy scar over perivaginal area. She had been previously treated with emollients and flucinolone acetonide cream 0.025% twice daily with minimal improvement. General examination was unremarkable. Dermatological examination showed erythematous, non-tender, indurated and infiltrated papules and plaques over philtrum, upper back, labia majora and perianal area (Figures 1-3). Rest

of the cutaneous and systemic examination was unremarkable. Skin biopsy was performed considering differentials of hypertrophic scars, sarcoidosis, lupus vulgaris, leprosy, tumid lupus erythematosus. Histopathological examination revealed dermis showing abundant non-caseating naked granulomas comprising epithelioid cells, lymphoplasmacytes and Langhans giant cells; extending till subcutaneous fat, clinching diagnosis of sarcoidosis (Figure 4). Routine haematological evaluation including serum calcium and urine examination was within normal limits. HIV serostatus was negative. Serum Angiotensin converting enzyme (Sr. ACE) level was significantly raised to 114.77 U/litre (Biological reference interval 8.0 to 52.0 U/litre), further supporting our diagnosis. Patient did not have any other co-morbidities. Radiological investigations including chest X-ray and ultrasonography of abdomen and pelvis showed no abnormality. Electrocardiogram was normal. The patient was started on oral hydroxychloroquine 200 mg twice daily after routine workup (ruling out color vision abnormalities and fundoscopy, slit lamp examination). Additionally, intralesional injections of Triamcinolone acetonide 10 mg/ml were administered every three weeks along with Betamethasone valerate 0.1% cream application once daily at night. Visible improvement was noted within nine weeks of treatment initiation. Currently patient is under monitoring with regular follow-up after four intralesional injection sessions.



Figure 1: Infiltrated erythematous plaque and few papules over philtrum area of nose.



Figure 2: Skin colored grouped papules over upper back.



Figure 3: Similar shiny papules and plaques over external genitalia at site of old posterolateral episiotomy scar.

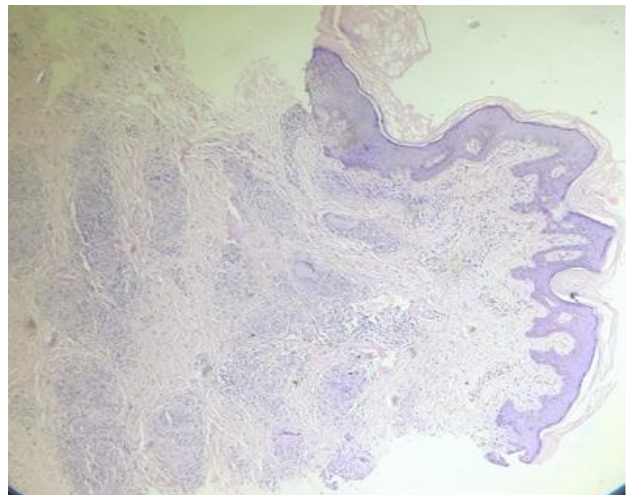


Figure 4: Skin biopsy with histopathological examination H & E (10X).

DISCUSSION

Morphological variants of cutaneous sarcoidosis include papules, subcutaneous nodules, plaques, lupus pernio, ulceration, scarring, erythema nodosum, alopecia. Rare presentations like follicular, verrucous, ichthyosiform, psoriasiform and annular lesions hypopigmented patches also has been described.⁴ Sarcoid granulomas in pre-existing scars can occur secondary to mechanical trauma such as venipuncture, intramuscular injections, surgery, post-herpes zoster, and tattoos, previous pseudo-folliculitis and sites of ritual scarifications.⁵ Interestingly, our case had developed sarcoidal lesions at sites of old (episiotomy) scar as well as recent (sutured contused lacerated wound) scars over philtrum, left upper lip and lower lip in addition to de-novo lesions over back. It is crucial that patients complaining of inflammation at the sites of old scars should be thoroughly examined.

Reactivation of old scars appears to be highly specific for sarcoidosis and hence early skin biopsy can diagnose scar sarcoidosis.

The development of sarcoidosis as a reaction to an environmental antigen, in addition, also depends on host's genetic polymorphisms and immune status. Majority of immunopathogenesis studies mainly focus on Th1/Th2 model highlighting immune dysregulation in sarcoidosis. However, role of Th17, a novel CD4⁺ effector T-cell population needs to be elucidated. High levels of IL-17⁺/CD4⁺ T lymphocytes have been found in sarcoid granulomas of disease-active patients.⁶ Recently, Richmond and colleagues verified the specificity of Th17 cells for mycobacterial antigens, a commonly implicated antigen for sarcoidosis, suggesting their role in disease progression.⁷ Serum angiotensin converting enzyme (ACE) levels have relatively lower sensitivity and specificity. Various novel biochemical markers in parallel to disease activity have been reported. These include detection of T-bet protein, IFN- γ and MicroRNA-29 (miR-29). However, ease of availability of these supportive tests is debatable.⁸ Our patient did not show any extracutaneous or systemic involvement. However involvement of *de novo* sites (without any scars or preceding trauma) points to disease activity, supporting above theory.

Spontaneous resolution is seen in most of sarcoidal cutaneous lesions. In case of progressive systemic involvement oral corticosteroids, methotrexate, azathioprine, mycophenolate mofetil, infliximab and etanercept, alefacept are reserved for patients. Local treatment modalities include high potency topical corticosteroids, tacrolimus, intralesional triamcinolone injections, cryotherapy, radiotherapy, PUVA therapy. Newer recent advances like laser, ultraviolet radiation and photodynamic therapies are also useful for treating cutaneous lesions. Antimalarial drugs are also beneficial for skin and joint sarcoidosis.⁹ As the patient had isolated cutaneous involvement she was started on oral Hydroxychloroquine 200 mg twice daily, intralesional injections of Triamcinolone acetonide, along with Betamethasone valerate 0.1% cream application once daily. Furthermore, surveillance of the behavior of scars form an important part of patient examination with suspected or proven sarcoidosis, as the prevalence scar sarcoidosis is rare, accounting for 5.4–13.8% of all cutaneous sarcoidosis cases.¹⁰

CONCLUSION

Despite nearly 130 years of extensive research, etiology, pathogenesis and management of sarcoidosis remains a challenge and constantly under scan. Sarcoidosis may be under-diagnosed due to lack of clinical suspicion. Better understanding of this disease and its varied manifestations mimicking common dermatologic

conditions like scars, psoriasis, lichen planus with availability of investigative modalities like histopathology and serum calcium and Sr. ACE levels can facilitate early diagnosis and timely management thereby preventing further complications.

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