

Rare Scar Sarcoidosis: A Dermatologic Masquerader

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Abstract

Infiltration of sarcoid granuloma in old cutaneous scars is one of the uncommon cutaneous manifestations of sarcoidosis. Here, we report the case of a 32-year-old female who presented with pain and swelling in 9 month old appendectomy scar. An incisional scar biopsy revealed granuloma. On query she had inflammatory polyarthritis and persistent dry cough for last 2 months. High-Resolution CT (HRCT) revealed right paratracheal, both hilar, paraaortic and subcarinal lymphadenopathy with pulmonary nodular densities in both lung fields. FNAC from mediastinal node revealed non caseating granuloma consistent with Sarcoidosis. Successful regression of cutaneous inflammation was achieved using a course of oral steroids, hydroxychloroquine and methotrxate. The onset of scar sarcoidosis is rare and, therefore, not well understood; however, inflammatory alterations in preexisting scars may be important indicators for disease onset.

Keywords: Sarcoidosis; Scar; Granuloma; Fine Needle Aspiration Cytology (FNAC).

Introduction

Sarcoidosis is a systemic inflammatory disorder of unknown origin that can present only cutaneous involvement or affect many organs such as lungs, eyes, lymph nodes and bones [1]. Cutaneous involvement occurs in 25% of cases [2]. Non-caseating granulomas (small inflammatory nodules) are found in the affected organs. Most patients with scar sarcoidosis have a systemic disease. Skin manifestations include erythema nodosum, lupus pernio papules, nodules, and plaques. Infiltration of old cutaneous scars with sarcoid granuloma in the active phase of disease, which is known as scar sarcoidosis, is the most clinically characteristic and an uncommon cutaneous manifestation of sarcoidosis [3, 4]. Cutaneous sarcoidosis is known as a great simulator of other diseases because of the lesional polymorphism⁴ and therefore it represents a major diagnostic challenge [5].

The first description of sarcoidosis, in 1800, was related to its cutaneous manifestations. The term sarcoidosis derives from a report from Boeck, in 1899, and it is due to the clinical similarities of the lesions with benign sarcomas. At the beginning of the year 1900, sarcoidosis was described involving lungs and other internal organs [6]. Sarcoidosis usually occurs in young adults, with two peaks of incidence: between 25 and 35 and 45 and 55 years of age. There is a prevalence of the disease among women [7]. The incidence of this multi-system disease is 10-15 cases per 100,000 each year [8]. Cicatricial onset is rare but clinically characteristic of cutaneous sarcoidosis [9].

Case Report:

A 32-year-old female patient presented painful reactivation of 9 months-old-scar located on right iliac fossa, originally acquired following appendectomy. The patient suffered localized scar pain without itching for two weeks prior to her visit to the outpatient surgery clinic. Systemic symptoms such as weight loss, fever, anorexia, hemoptysis, chest pain, shortness of breath also absent. But on query she complained of symmetrical polyarthritis involving mainly small hand joints with marked inactivity stiffness and resistant dry cough for the last 2 months. Systematic physical examinations for the respiratory, cardiovascular and nervous systems, such as lung and heart auscultation, reported normal results. Ophthalmoscopic examination, including a slit-lamp study, revealed normal findings. However, dermatological physical examinations found erythematous swelling and small papules on localized scar and one subcutaneous indurated nodules, 2×3 cm in size, were found under and below the scar (Figure 1).



Figure 1: Papules over scar with indurated nodule

The patient additionally underwent auxiliary examinations, including a complete blood count and tests to determine erythrocyte sedimentation rate, c-reactive protein and electrolyte levels, and liver and renal function, which revealed Hemoglobin 10.2 gm%, total count 8200 /cmm, lymphocyte 40%, ESR was 18 mm in 1st hour, CRP was 19. ALT was 84, Corrected serum calcium was 8.9 mg/dl. Blood levels of angiotensin-converting enzyme, typically used to aid in the diagnosis of sarcoidosis, was 30 IU/l (reference range 24–65 IU/l), and the urinary analysis was found to be normal. The tuberculosis skin test also reported negative results. ANA and RA tests were negative. However, a biopsy was carried out on the skin lesion and histopathology under light microscopy of the tissue revealed multiple non-caseating granulomas composed of epithelioid cells, numerous multinucleated giant cells and a small number of lymphocytes suggestive of foreign body granuloma (Figure 2).

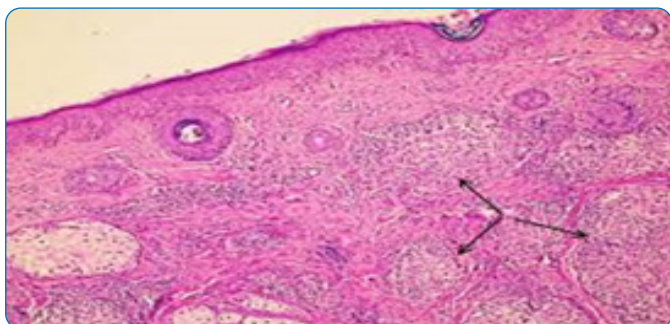


Figure 2: Histopathology from scar tissue showing multiple granuloma

The patient's chest was also examined. A chest x-ray revealed bilateral hilar lymphadenopathy (Figure 3) while computer tomography of the chest revealed two obscure nodules (largest one ~10 mm in size) in the lungs (Figure 4) and swollen lymph nodes in the hilum pulmonis and mediastinum (paratracheal, both hilar, paraaortic, and subcarinal lymphadenopathy) (Figure 5). CT guided FNAC was done from mediastinal lymphnodes which revealed non caseating granuloma (Figure 6).

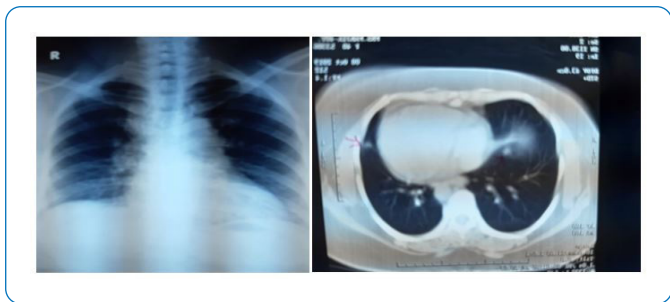


Figure 3&4: Chest X-ray showing mediastinal lymphadenopathy and CT chest showing two pulmonary nodules

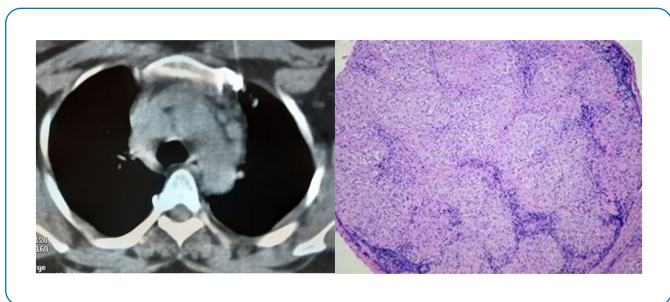


Figure 5&6: CT chest showing mediastinal lymphadenopathy and histopathology from mediastinal node showing multiple non caseating granuloma Detection of acid-fast bacilli was negative in the

biopsy sample. Pulmonary function tests were not done and patient refused for bronchoscopy. Following the diagnosis of scar sarcoidosis, the patient was treatment with prednisolone, hydroxychloroquine 200 mg twice daily and weekly 10 mg methotrexate with folic acid. After a short course of oral prednisolone (30 mg per day for 4 weeks and gradually reduced over the next month), the visible inflammation in the scars regressed. She remained stable without recurrence for a follow-up period of 6 months. However, hilar lymphadenopathy on chest radiography persisted.

Discussion

Sarcoidosis is a multisystemic disease, of unknown etiology, characterized by the formation of non-caseating granulomas in the affected organs. The involvement of the skin occurs in 25% of the cases and it can develop in any phase of the disease although it is more common at the beginning [10]. The relation between cutaneous and systemic sarcoidosis has been evaluated. About 30% of the patients with isolated cutaneous lesions will develop systemic sarcoidosis, after a period of time which varies from one month to one year.

The diversity of clinical presentations mean that sarcoidosis is difficult to diagnose.

Disease diagnosis requires a combination of:

- Supporting clinical-radiological findings such as hilar and/or paratracheal lymph node enlargement with or without pulmonary infiltrates [11].
- Histological evidence of non-caseating epithelioid granulomas at disease site
- Exclusion of alternative causes for the granulomatous inflammation and local sarcoid-like reactions [12].

Skin lesions in sarcoidosis may appear as maculae, papules, plaques, nodules, ulcers, localized alopecia, ichthyotic areas, subcutaneous nodules, lupus pernio, scar sarcoidosis, psoriasiform and even pustules. Among these, scar sarcoidosis is rare, accounting for 5.4–13.8% of sarcoidosis cases [13].

Cicatricial onset has been found in 29% of the patients with cutaneous sarcoidosis. Besides the reactivation of scars originated from previous wounds, cicatricial sarcoidosis has been found in areas of intramuscular injections and hyaluronic acid, tattoos, venepuncture and cutaneous manifestation of herpes zoster, after a period of time which varies from six months to 59 years. The previous contamination of these scars, with strange material, has been suggested as a possible subjacent cause. The patient in study showed nodules on scars 9 months after appendicectomy, without report of trauma or infections.

Scar sarcoidosis is characterized by the onset of erythematous swelling and the development of papules and nodules 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. Excluding skin damaged by mechanical injury, scar sarcoidosis can

occur on skin sites damaged by a range of factors, including venipuncture, intramuscular injections, inoculations, tattoos and infections such as herpes zoster [14]. It is suggested that foreign material within the scar, deposited by external factors including those stated above, is a possible cause of epithelioid granuloma [15]. The specific skin lesions that occur and the resulting sarcoidosis may be associated with the severity and duration of the disease, with scar sarcoidosis often being accompanied by systemic involvement. Alterations, such as further damage or stress to the existing scars, often prompt worsening of sarcoidosis [16].

The case of scar sarcoidosis presented here is an middle aged woman, displaying no other health problems, including tuberculosis and hepatitis. The patient visited the clinic due to the development of painful nodules and erythematous swelling on preexisting scars located on right iliac region, which formed following appendectomy 9 months back. On query patient complained of symmetrical polyarthritis involving mainly small hand joints with marked inactivity stiffness and resistant dry cough for the last 2 months. Although the patient did not report any systemic symptom, On HTCT chest it was found that she had stage II pulmonary involvements, while all other organs were not involved except for asymptomatic biochemical hepatitis. As chest X-ray or computed tomography scan examination was not performed on the patient within several years, it could not be determined whether the onset of pulmonary involvement occurred before or after scar sarcoidosis; however the majority of patients with scar sarcoidosis develop systemic disease. Therefore, it is necessary for patients who are presenting with painful inflammation on or around their existing scars to be examined systematically and followed up. Standard therapies for sarcoidosis include the administration of corticosteroids, antimalarials and methotrexate [17]. However, scar sarcoidosis often resolves slowly and spontaneously. Thus, the patient in the present case refused treatment. Alternative second-line drug treatments include methotrexate and hydroxychloroquine; however, these agents are not 100% effective [18]. For patients with progressive cutaneous sarcoidosis or refractory cases, monoclonal antibodies are a novel therapeutic option. For example, etanercept antibodies [18-20]. That target tumor necrosis factor- α have been demonstrated to be beneficial in treating recalcitrant sarcoidosis. In the treatment of scar sarcoidosis, injection of the corticosteroid triamcinolone acetonide into the skin lesions is also effective. Furthermore, it is necessary to continually survey patients via pulmonary examination and chest radiography every two months, as well as periodic monitoring for other systemic manifestations.

References

1. Hiram Larangeira de Almeida Jr, Heitor Alberto Jannke. "Sarcoidose em cicatrizes prévias." *An Bras Dermatol* (2004):79-82.
2. Joseph C. English, Purvisha J. Patel, Kenneth E. Greer. "Sarcoidosis." *J Am Acad Dermatol* (2001):725-743.
3. N Milman, AL Hoffmann, K-E Byg. "Sarcoidosis in Children: Epidemiology in Danes, Clinical Features, Diagnosis, Treatment and Prognosis." *Acta Paediatr* (1998):871-878.

4. Katta, Rajani. "Cutaneous Sarcoidosis: a Dermatologic Masquerader." *Am Fam Physician* (2002):1581-4.
5. Esteban Fernandez-Faith, Jonelle McDonnell. "Cutaneous Sarcoidosis: Differential Diagnosis." *Clin Dermatol* (2007):276-87.
6. Richard M. Marchell, Marc A. Judson,. "Chronic Cutaneous Lesions of Sarcoidosis." *Clin Dermatol*. (2007):295-302.
7. Athol Wells, V. Kouranos. "Sarcoidosis." *Am Fam Physician* (2004):312-22.
8. Marcel-Philipp Henrichs, Arne Streitbürger, Georg Gosheger, Carsten Surke, Christian Dierkes, et al." Scar Sarcoidosis on a Finger Mimicking a Rapidly Growing Soft Tissue Tumour: A Case Report." *BMC Res Notes* (2012):545-549.
9. Young Chul Hong, Dong Jib Na, Sang Hoon Han, Yang Deok Lee, et al. "A case of scar sarcoidosis." *Korean J Intern Med* (2008):213-5.
10. W.-S. Chong, H.-H. Tan, S.-H. Tan. "Cutaneous Sarcoidosis in Asians: a Report of 25 Patients From Singapore." *Clin Exp Dermatol* (2005):120-4.
11. D. Dal Sacco, E. Cozzani, Aurora Parodi, A. Rebora. "Scar Sarcoidosis After Hyaluronic Acid Injection." *Int J Dermatol* (2005):411-2.
12. Spagnolo, Paolo. "Sarcoidosis: A Critical Review of History and Milestones." *Clin Rev Allergy Immunol* (2015):1-5.
13. Mañá, Juan. "Cutaneous Involvement in Sarcoidosis. Relationship to Systemic Disease." *Arch Dermatol* (1997):882-888.
14. S. Y. Choi, M. Y. Hyun, Y. A. No, K. Y. Park, et al. "Scar Sarcoidosis on a Hypertrophic scar." *Clin Exp Dermatol* (2014):945-947.
15. I. Cervigón, A. Palomo, L.M. Torres. "Cutaneous Lesions on Scars." *Actas Dermosifiliogr* (2007):433-434.
16. J. Marcoval, J. Mañá, M. Rubio. "Specific Cutaneous Lesions in Patients With Systemic Sarcoidosis: Relationship to Severity and Chronicity of Disease." *Clin Exp Dermatol* (2011):739-744.
17. Christy B Doherty, Ted Rosen. "Evidence-Based Therapy for Cutaneous Sarcoidosis." *Drugs* (2008):1361-1383.
18. Adele Haimovic, Miguel Sanchez, Marc A. Judson, Stephen Prystowsky. "Sarcoidosis: A Comprehensive Review and Update for the Dermatologist: Part I. Cutaneous Disease." *J Am Acad Dermatol* (2012):699.e1-e18.
19. Spagnolo, Paolo. "Sarcoidosis: A Critical Review of History and Milestones." *Clin Rev Allergy Immunol* (2015):1-5.
20. Papapit Tuchinda, Matthew Bremmer, Anthony A. Gaspari. "Etanercept for chronic progressive cutaneous sarcoidosis." *J Drugs Dermatol* (2006):538-540.

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