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Case Report

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 swelling and irritation in a 9-month-old appendicectomy scar. An incisional scar biopsy was done and it revealed non-caseating granuloma. On query she had inflammatory polyarthritis and persistent dry cough for last two 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 methotrexate. The incidence of scar sarcoidosis is rare and therefore not well-understood. However, inflammatory alterations in preexisting scars may be important indicators of disease onset.

Key Words: *Sarcoidosis; Scar; Granuloma; Fine needle aspiration cytology (FNAC)*

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Introduction

Sarcoidosis is a systemic inflammatory disorder of unknown etiology that can present with only cutaneous involvement or can affect many organs such as lungs, eyes, lymph nodes and bones.[1] Cutaneous involvement occurs in 25% of the 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 is known as scar sarcoidosis which is the clinically characteristic and an uncommon cutaneous manifestation of sarcoidosis.[3] Cutaneous sarcoidosis is known as a great simulator of other diseases because of the lesional polymorphism[4] and therefore it represents a major diagnostic challenge.[5]

Sarcoidosis was described first in 1800 and it was related to cutaneous manifestations. The term sacoidosis 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 found involving lungs and other internal organs.[6] Sarcoidosis usually occurs in young adults with two peaks of incidence at 25–35 years and 45–55 years of age.[3] There is an increased prevalence of the disease among women.[5,7] The incidence of this multisystem disease is 10–15 cases per 100,000 each year.[8] Cicatricial onset is rare but it is clinically characteristic of cutaneous sarcoidosis.[9]

Case report

A 32-year-old female patient presented to the surgery

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outpatient department with painful reactivation of 9-month-old appendicectomy scar located in right iliac fossa. The patient complained of pain in the scar without itching for two weeks prior to her visit. She had no history of weight loss, fever, anorexia, hemoptysis, chest pain and shortness of breath. On query she complained of symmetrical joint pain involving mainly small hand joints with marked inactivity stiffness and persistant dry cough for the last two months. Respiratory, cardiovascular and nervous system examination revealed no abnormal findings. Ophthalmoscopic examination including a slit-lamp study was normal. On examination of the skin there were erythematous swelling and small papules in localized scar and one subcutaneous indurated nodule of 2 cm×3 cm in size under and below the scar (Fig 1).

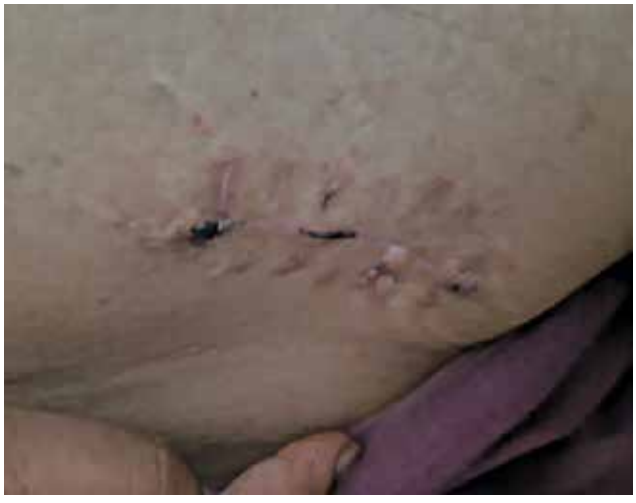

Fig 1. Papules over scar with indurated nodule

Investigation reports are as follows: hemoglobin 10.2 gm%, WBC 8200/cmm, lymphocyte 40%, ESR 18 mm in 1st hour, CRP 19, ALT 84 U/L and corrected serum calcium was 8.9 mg/dL. Serum angiotensin converting enzyme level was 30 IU/L (reference range 24–65 IU/L) and urinary analysis was normal. The tuberculosis skin test was negative. ANA and RA tests were negative. Skin biopsy was done 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 (Fig 2).

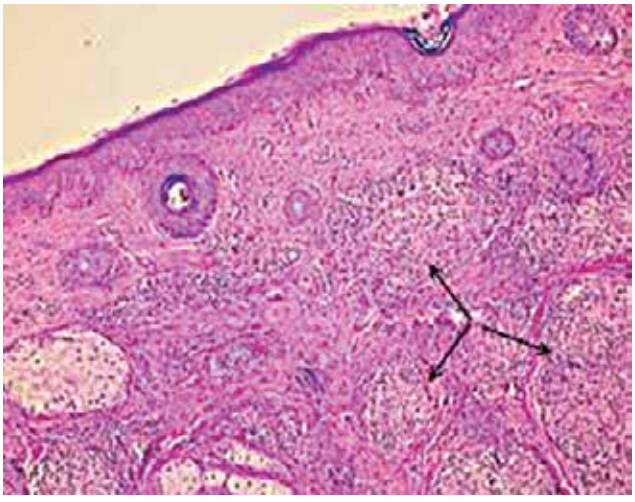

Fig 2. Histopathology from scar tissue showing multiple granuloma

Chest radiography showed bilateral hilar lymphadenopathy (Fig 3) and CT scan of chest revealed two obscure nodules (largest one ~10 mm in size) in the lungs (Fig 4) and swollen lymph nodes in the hilum pulmonis and mediastinum (paratracheal, both hilar, para-aortic and subcarinal lymphadenopathy) (Fig 5). CT-guided FNAC was done from mediastinal lymph node and non-caseating granuloma was found in cytological study (Fig 6).

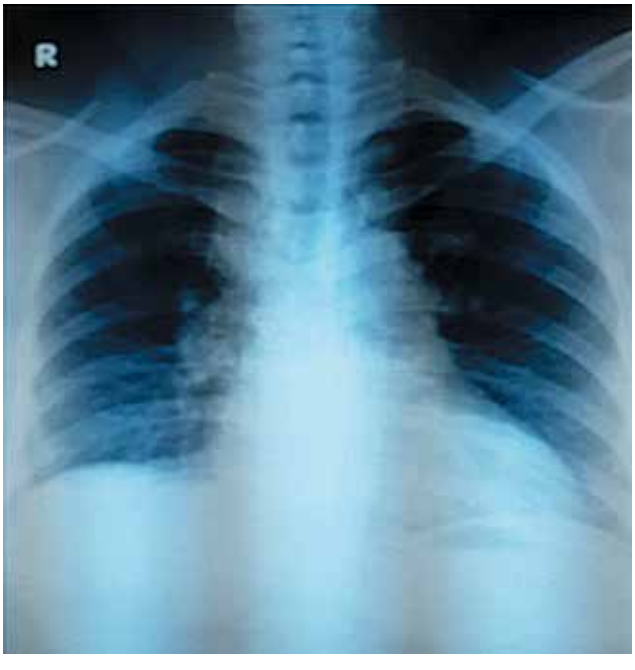

Fig 3. Chest X-ray showing mediastinal lymphadenopathy

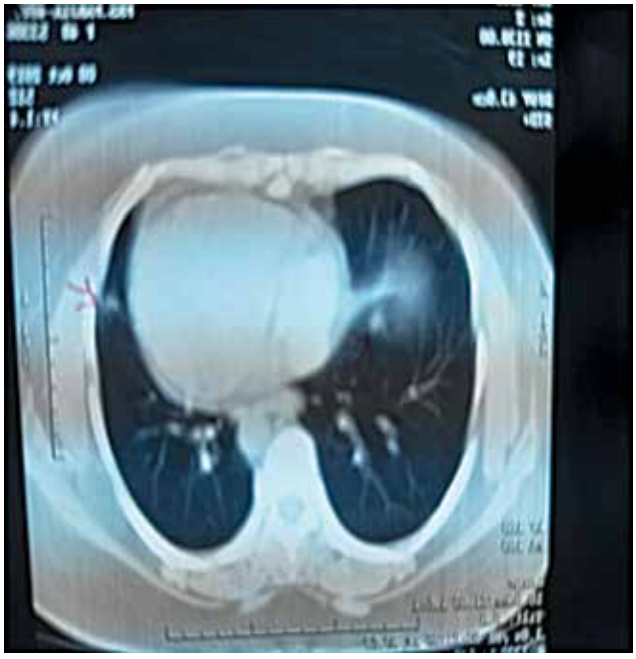

Fig 4. CT scan of chest showing two pulmonary nodules

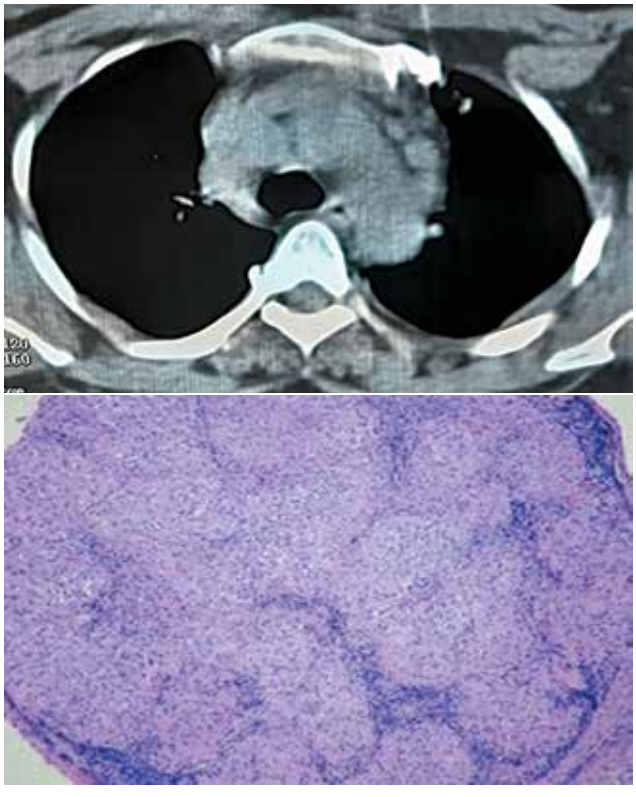

Fig 5. CT scan of chest showing mediastinal lymphadenopathy and histopathology from mediastinal node showing multiple non-caseating granuloma

Acid-fast bacilli were not found in the biopsy sample. Pulmonary function tests were not done and patient refused to undergo bronchoscopy. Following the diagnosis of scar sarcoidosis, the patient was treated with prednisolone 30 mg/day for four weeks then gradually tapered, hydroxychloroquine 200 mg twice daily and methotrexate weekly 10 mg and folic acid. After treatment the visible inflammation in the scar regressed. She remained stable without recurrence for a follow-up period of six months. However, hilar lymphadenopathy on chest radiography persisted.

Discussion

Sarcoidosis is a multisystem disease of unknown etiology characterized by the formation of non-caseating granulomas in the affected organs.[9] 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.[5] 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.[7]

The diversity of clinical presentations means that sarcoidosis is difficult to diagnose. Disease diagnosis requires a combination of following criteria: i) supporting clinical-radiological findings such as hilar and/or paratracheal lymph node enlargement with or without pulmonary infiltrates, ii) histological evidence of non-caseating epithelioid granulomas at disease sites and iii) exclusion of alternative causes for the granulomatous inflammation and local sarcoid-like reactions.[11] 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.[6,12]

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, tatoos, venepuncture and cutaneous manifestation of herpes zoster after a period of time which varies from six months to 59 years.[9,13,14] The previous contamination of these scars with foreign material has been suggested as a possible cause of

epithelioid granuloma.[9,15] The patient in study showed nodules on scars nine 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.[8] 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.[13] Alterations, such as further damage or stress to the existing scars, often prompt worsening of sarcoidosis.[16]

Our case of scar sarcoidosis was a middle-aged woman who had no other health problems including tuberculosis and hepatitis. The patient presented with development of painful nodules and erythematous swelling on preexisting scars located on right iliac region, which formed following appendectomy 9 months back. She also had symmetrical polyarthritis involving mainly small hand joints with marked inactivity stiffness and resistant dry cough for the last two months. Chest radiography showed stage II pulmonary involvements. As chest radiography or CT scan was not performed 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.[14] Standard therapies for sarcoidosis include the administration of corticosteroids, antimalarials and methotrexate.[17] However, scar sarcoidosis often resolves slowly and spontaneously.[6] Our patient was treated with standard protocol and improved. For patients with progressive cutaneous sarcoidosis or refractory cases, monoclonal antibodies are novel therapeutic options. For example etanercept that target tumor necrosis factor-α have been demonstrated to be beneficial in treating recalcitrant sarcoidosis.[12,18] In the treatment of scar sarcoidosis, injection of the corticosteroid triamcinolone acetonide into the skin lesions is also effective.[14] Furthermore, it is necessary to continue follow-up the patients with regular pulmonary examination and chest radiography every two months, as well as periodic monitoring for other systemic manifestations.

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