CASE STUDIES

A case of Darier-Roussy sarcoidosis

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Abstract

Subcutaneous sarcoidosis, a rare form of cutaneous sarcoidosis, is seen in approximately 4.3-12.1% of patients with systemic sarcoidosis. These lesions are often associated with less severe systemic disease and usually appear at the beginning of the disease. The present study discusses an unusual case of subcutaneous sarcoidosis with interferon-γ release assay (IGRA) positivity. The case report highlights the increased chances of misdiagnosis of Darier-Roussy sarcoidosis as tuberculosis based on positive IGRA, especially in countries with increased prevalence of TB.

Keywords: subcutaneous sarcoidosis, tuberculosis, Interferon-γ release assay, IGRA

Introduction

Subcutaneous sarcoidosis (previously known as Darier-Roussy sarcoidosis), restricted to the subcutaneous tissue, is a specific form of cutaneous sarcoidosis characterized by typical sarcoidal granulomas. The prevalence of subcutaneous sarcoidosis in patients with systemic sarcoidosis is approximately 4.3-12.1%.[1, 2] The disease has a female predilection with a peak incidence during the fourth decade. Typical skin finding is the presence of painless, firm, mobile nodules (measuring 0.5-2 cm in diameter), without overlying epidermal involvement. The nodules can occur over single or multiple sites and are mainly distributed in the extremities (78.8%), followed by the trunk (28%), buttocks (10%), and forehead (5%).[3]

Case report

A 60-year-old female patient presented with non-itchy, reddish-brown skin rashes over the periorbital area (Fig. 1), and multiple painless nodular lesions over extensor surfaces of fingers (Fig. 2), hands, forearms (Fig. 3) and shins (measuring around 5-25 mm). She had no fever, cough, dyspnea, weight loss, eye pain, and blurring of vision. Medication history revealed that the patient was on telmisartan, amlodipine, glimepiride, metformin and voglibose, for hypertension and diabetes. Ophthalmic examination using Schirmer test showed complete absence of tears in the right eye and 15 mm in the left eye. She also had left eye vitritis. Systemic examination was normal.

Her blood investigations revealed: normal blood counts, ESR: 22 mm/hr, CRP: 11.30 mg/L, positive anti-nuclear antibody (1+) with speckled pattern at 1:40 dilution, negative rheumatoid factor (24.5 IU/ml), elevated angiotensin converting enzyme (ACE) (68U/L [normal-52U/]) and positive QuantiFERON gold (interferon-γ assay, IGRA). Chest x-ray showed hilar widening with normal lung parenchyma. High-resolution computed tomography (HRCT) chest was normal. Biopsy from the nodule showed multiple discrete granulomas composed of epithelioid cells, multinucleate giant cells surrounded by fibrosis and a few lymphocytes with no necrosis involving the dermis and subcutis (Fig. 4). The Ziehl-Neelsen and modified Fite-Faraco staining were negative for *M. tuberculosis* and *M. leprae* respectively. Based on the biopsy finding and other clinical investigations, the final diagnosis was concluded as sarcoidosis.

The patient was treated with oral prednisolone 0.25 mg/kg, methotrexate 10 mg/week and hydroxychloroquine 400 mg/day. The nodules almost regressed in 2 months. Methotrexate was gradually increased over 4 months to 25 mg/week and steroids were tapered. There was no relapse of disease activity.

Fig. 1: Maculopapular, red-brown skin rashes over the periorbital area

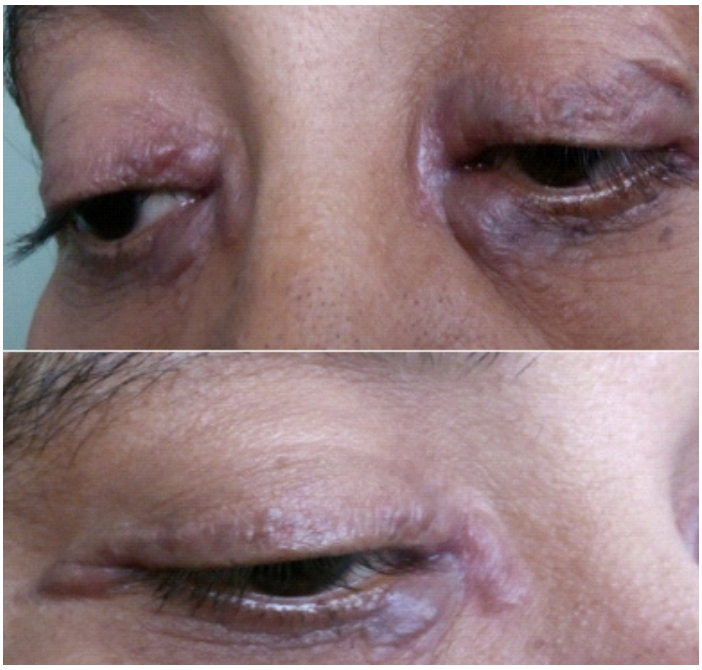


Fig. 2: Subcutaneous nodules over dorsum of hands and fingers

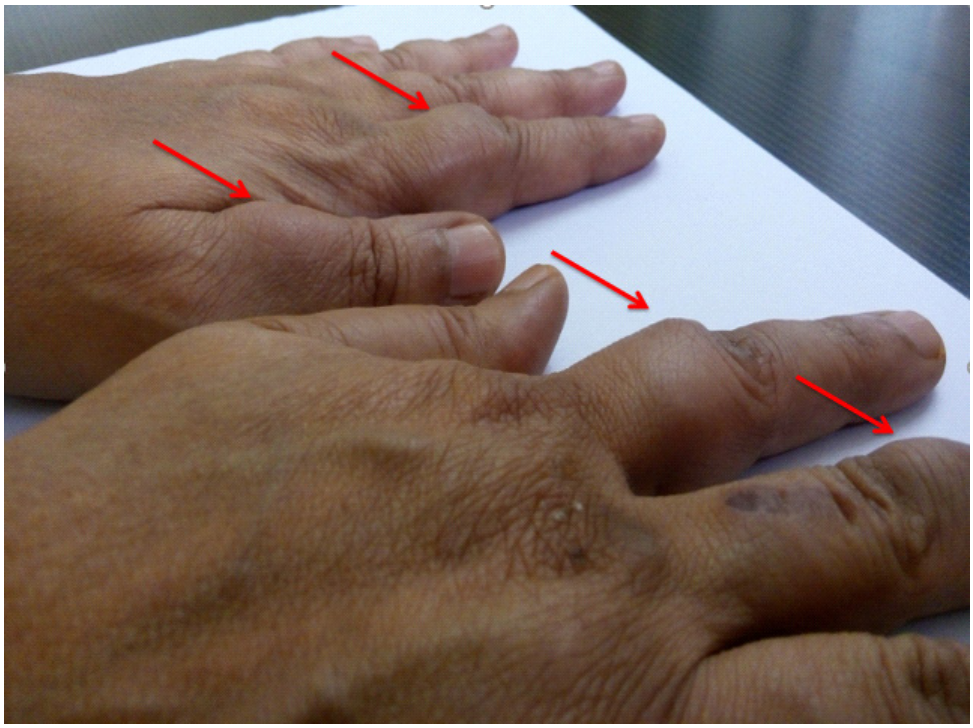


Fig. 3: Subcutaneous nodules over extensor surface of forearms

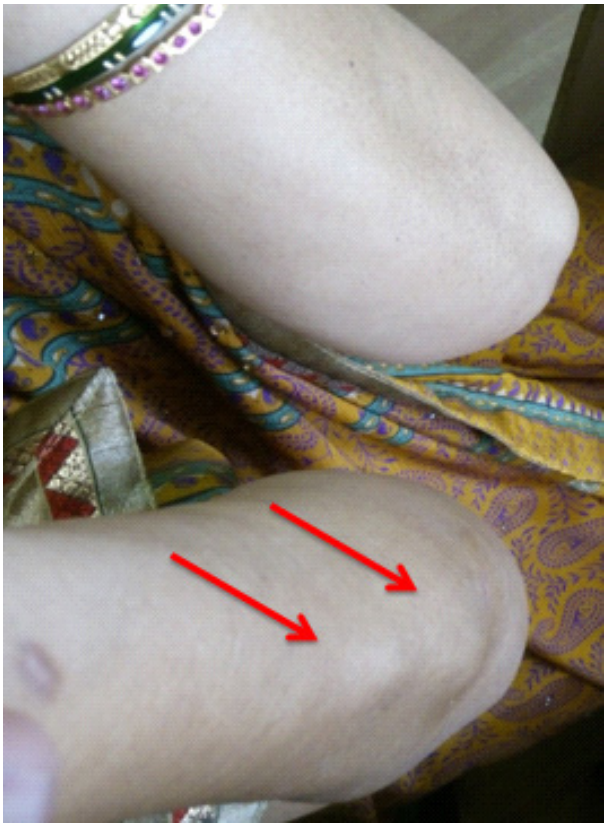
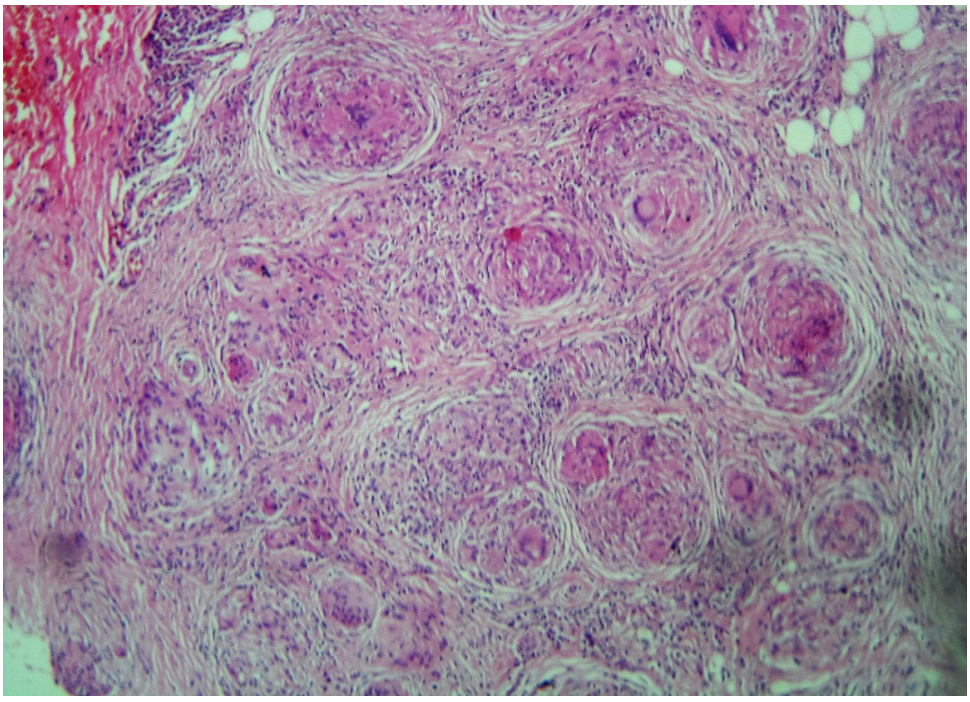


Fig. 4: Biopsy from the nodule showing multiple discrete epithelioid cell granulomata involving the dermis and subcutis (H & E stain, 10 x 10 magnification)



Discussion

Patients with subcutaneous sarcoidosis often have less severe systemic disease. The disease is usually associated with extracutaneous systemic disease involvement, especially hilar adenopathy. However, the present case did not have hilar lymphadenopathy. It may sometimes present as the initial manifestation of systemic sarcoidosis. The diagnosis of subcutaneous sarcoidosis depends on identifying subcutaneous sarcoidal or epithelioid granulomas with minimal lymphocytic inflammation.[1, 3, 4] In addition to the occurrence of subcutaneous nodules over extremities, the present patient had maculopapular lesions around periorbital area, which is the most common form of specific cutaneous sarcoidosis. Co-existence of other types of specific skin lesions with subcutaneous sarcoidosis, including plaques, papules, and scar sarcoidosis, was noted in 71% of the patients in a study by Ahmed *et al*.[3]

The current patient was positive for IGRA assay. Due to similar clinical, histological and radiological findings, differential diagnosis of tuberculosis (TB) and sarcoidosis is difficult, especially in countries with high burden of TB prevalence. *M. tuberculosis* is considered as a possible etiological agent in sarcoidosis. The cumulative evidence suggests that mycobacterial antigens are responsible for initiating and/or maintaining granulomas in some patients with sarcoidosis. A meta-analysis by Gupta *et al.* has reported the presence of mycobacterial DNA and RNA in 48% of sarcoid biopsy tissues using nucleic acid amplification tests.[5] Patients with positive IGRA are sometimes misdiagnosed as TB. A recent study from north India by Gupta *et al.* showed positive IGRA in 34.2% patients with sarcoidosis. In the present case, the diagnosis was concluded on the basis of high ACE levels and the biopsy showing non-caseating granulomas, in addition to positive IGRA. Thus, in countries with high TB prevalence, it is necessary to consider other diagnostic factors, in addition to positive IGRA, for confirming the diagnosis of TB.[6]

The nodules resolve spontaneously in certain cases. The mainstay of treatment for subcutaneous sarcoidosis is oral glucocorticoids (prednisone, 20-40 mg/day), with responses noted within 4 to 8 weeks after initiation of therapy.[3] The most common steroid-sparing drug used for subcutaneous sarcoidosis is hydroxychloroquine. Methotrexate, clofazamine, intralesional glucocorticoids, thalidomide, dapsone, allopurinol, and minocycline are also used for treatment.[4, 7]

Competing interests

The authors declare that they have no competing interests.

Citation

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