Case Report

Annular cutaneous sarcoidosis with systemic involvement

Ajay Kumar Mishra[1], Anu Anna George[2], Dincy Peter[2]

[1]Departments of Internal Medicine, [2]Dermatology, Christian Medical College, Vellore, Tamil Nadu, India

Abstract

Sarcoidosis is a granulomatous disease involving multiple systems. Cutaneous involvement is present in 25% of patients. A 42-year-old woman presented with itchy skin lesions on her face for 5 years duration. She was found to have annular and discoid plaques with prominent overlying telangiectasia. A biopsy from the plaque was suggestive of sarcoidosis. On further evaluation, she was found to have both pulmonary and ocular involvements. Annular sarcoidosis is a rare variant of cutaneous sarcoidosis. We report this case to highlight this rare variant of sarcoidosis and discuss the various cutaneous manifestations of sarcoidosis.

Keywords: Annular, cutaneous, sarcoidosis, systemic

Introduction

Sarcoidosis is a granulomatous disease involving multiple systems in the body. Cutaneous involvement can occur up to 25% of patients. Annular sarcoidosis is a rare variant of cutaneous sarcoidosis. Early recognition of this form of sarcoidosis is important as it is commonly associated with systemic involvement.

Case Report

A 42-year-old woman from Assam presented with itchy lesions on the face for 5 years. The lesions initially started on the right cheek and gradually spread to involve the rest of the face. Her general and systemic examinations were within normal limits. On cutaneous examination, she was found to have dull erythematous coin shaped and annular scaly plaques varying in size from 1 cm × 1 cm to 4 cm × 5 cm scattered on the forehead, infraorbital area, cheeks, and chin [Figure 1]. On dermoscopy, a structureless area with multiple large wavy vessel loops was seen [Figure 2].

Address for correspondence: Dr. Ajay Kumar Mishra, 56-B4, Old PG Quarters, Christian Medical College and Hospital, Vellore - 632 004, Tamil Nadu, India.
E-mail: ajaybalasore@gmail.com

Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/2249-4863.222012

The clinical differentials considered were sarcoidosis, granulomatous rosacea, subacute lupus erythematosus, lupus vulgaris, and fungal infections. Her blood investigations including a serum angiotensin-converting enzyme and antinuclear antibody were within normal limits. Mantoux test was negative. Skin biopsy done from one of the plaques showed epidermal hyperkeratosis with follicular plugging. The superficial dermis showed multiple confluent granulomas composed of epithelioid histiocytes and Langhans type of multinucleated giant cells with few very lymphocytes. On reticulin staining, there are preserved reticulin fibers in and around these granulomas. Special stains for acid-fast bacilli and fungal microorganisms were negative. Based on the above, a diagnosis of cutaneous annular sarcoidosis was made. A spirometry done showed a mild restrictive pattern. A computed tomography scan showed multiple scattered calcified hilar nodes and subpleural nodules consistent with pulmonary involvement of sarcoidosis [Figure 3]. Ophthalmology examination showed the presence of a chronic dacryocystitis. An ENT evaluation to look for any sinus pathology was noncontributory.

She was started on tablet hydroxychloroquine and topical tacrolimus and is on follow-up.

How to cite this article: Mishra AK, George AA, Peter D. Annular cutaneous sarcoidosis with systemic involvement. J Family Med Prim Care 2017;6:660-2.

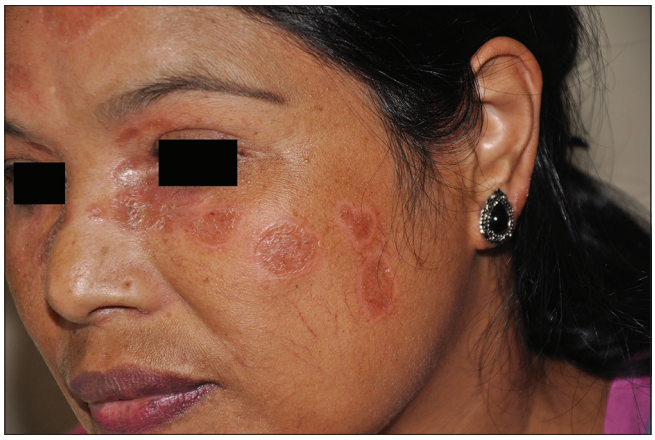

Figure 1: Dull erythematosus discoid and annular scaly plaques varying in size from 1 cm × 1 cm to 4 cm × 5 cm scattered on the forehead, infraorbital area, cheeks, and chin with prominent telangiectasia

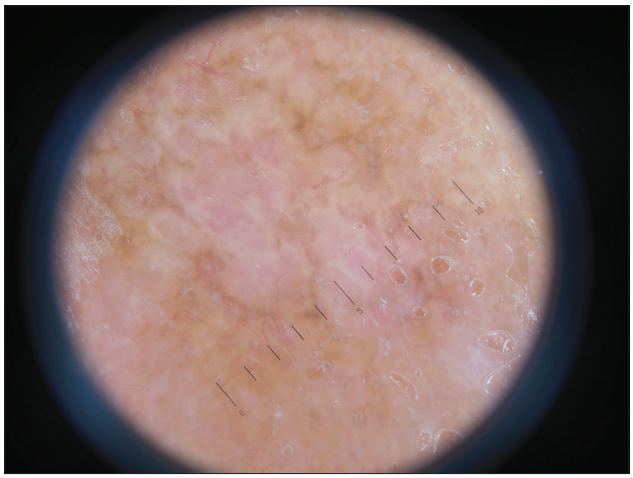

Figure 2: Dermoscopic photograph of the plaque on the face showing a structureless area with well-defined, large, wavy, vessel loops

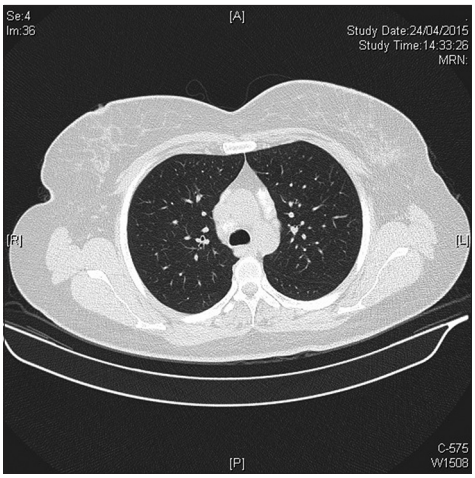

Figure 3: Computed tomography scan showing multiple scattered calcified hilar nodes and subpleural nodules consistent with pulmonary involvement of sarcoidosis

Discussion

Sarcoidosis is a noncaseating granulomatous disease involving multiple systems in the body.[1] Cutaneous involvement is seen in up to 25% of patients with sarcoidosis and is present with or without systemic involvement.[1] Cutaneous sarcoidosis occurs in 20%–35% of patients with systemic sarcoidosis.[2] Patients with cutaneous sarcoidosis may not have any systemic involvement in about 25% of the patients.[3] About 25% of the skin lesions of sarcoidosis have been found to occur on the face. The extent of the cutaneous lesions does not correlate with systemic skin involvement. Cutaneous involvement in sarcoidosis can be divided either into specific and nonspecific based on the presence of granulomas in the former or into acute erythema nodosum and chronic granulomatous form.[1] Skin lesions in sarcoidosis can be of varying morphologies, and hence, it has been known as one of the "great imitators."[4] The classical lesions described include erythema nodosum, maculopapular and erythematous forms, papules, nodules, annular lesions, angiolupoid lesions, lupus pernio, and subcutaneous sarcoid. Annular lesions are well-recognized forms of cutaneous sarcoidosis, amounting for around 8% of all skin lesions according to a recent study from India.[5] Along with the skin involvement, our patient also had systemic involvement in the form of chronic dacryocystitis as well as hilar lymphadenopathy and subpleural nodules. Cutaneous involvement not only facilitates early diagnosis owing to the presence of noncaseating granulomas but also can predict the presence of systemic involvement in case of the presence of lupus pernio, subcutaneous nodules, plaques, and angiolupoid sarcoidosis.[6] In conclusion, patients with cutaneous sarcoidosis may not have any systemic involvement in about 25% of the patients.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Newman LS, Rose CS, Maier LA. Sarcoidosis. N Engl J Med 1997;336:1224-34.
2. Kerdel FA, Moschella SL. Sarcoidosis. An updated review. J Am Acad Dermatol 1984;11:1-19.
3. Veien NK, Stahl D, Brodthagen H. Cutaneous sarcoidosis in Caucasians. J Am Acad Dermatol 1987;16(3 Pt 1):534-40.

4. Katta R. Cutaneous sarcoidosis: A dermatologic masquerader. Am Fam Physician 2002;65:1581-4.
5. Mahajan VK, Sharma NL, Sharma RC, Sharma VC. Cutaneous sarcoidosis: Clinical profile of 23 Indian patients. Indian J Dermatol Venereol Leprol 2007;73:16-21.
6. Marcoval J, Mañá J, Rubio M. Specific cutaneous lesions in patients with systemic sarcoidosis: Relationship to severity and chronicity of disease. Clin Exp Dermatol 2011;36:739-44.