Case Report

A DIAGNOSED CASE OF SARCOIDOSIS PRESENTING WITH DLE (DISCOID LUPUS ERYTHEMATOSUS):

Farah Sameem, Seerat Fatima, Sheikh Javeed Sultan, Sheikh Manzoor

Abstract

Sarcoidosis is a multisystem granulomatous disease involving mainly the lungs, mediastinal and peripheral lymph nodes, eyes and skin. Sarcoidosis is considered one of the 'great imitators' in dermatology due to its extremely variable cutaneous manifestation. One of the cutaneous forms of sarcoidosis includes the nodular and plaque type which can mimic many conditions including DLE (Discoid Lupus Erythematosus). The systemic involvement and progressive nature of sarcoidosis makes it important for us to differentiate sarcoidal lesions from DLE (Discoid Lupus Erythematosus), so as to avoid potential long term sequelae and provide proper treatment. Also, the concurrent existence of sarcoidosis and DLE (Discoid Lupus Erythematosus) is rarely documented in literature and needs further research and evaluation. Hence we present the case of a 37 year old female, a known case of sarcoidosis with skin lesions clinically and histologically similar to DLE (Discoid Lupus Erythematosus) so as to fill in some of the gaps in this area of study.

JK-Practitioner2021;26(1):55-57

Introduction

Sarcoidosis is an idiopathic systemic granulomatous disease, in which noncaseating granulomatous inflammation can occur in any organ. The lungs are the most affected organs, with approximately 90% of the patients presenting alterations in chest radiographs during the disease. Other manifestations include cutaneous, cardiac, ophthalmologic, hepatic and joint involvement. Sarcoidosis is, therefore, a heterogeneous disorder, both in terms of clinical presentation and severity. The skin being affected in about one fourth of the cases. Various specific forms of cutaneous sarcoidosis include maculopapular, nodular, subcutaneous, scar, and plaque sarcoidosis. In patients with plaque-type lesions, the activity of the systemic disease usually persists for more than 2 years [1-2]. They are associated with chronic forms of sarcoidosis including pulmonary fibrosis, peripheral lymphadenopathy, splenomegaly and chronic uveitis [2-3] and hence has a worse prognosis than the other cutaneous sub-types.

DLE is an inflammatory disorder of the skin, most frequently involving areas like the face and scalp, and characterized by well-demarcated erythematous, scaly plaques of variable size, that heal with atrophy, scarring and pigment changes. The disease may also affect areas away from the face and scalp and is known to have characteristic histology. DLE forms one end of a spectrum of this multi-systemic disease, with SLE at the other end of the spectrum. The risk of a patient with DLE developing overt SLE varies from 1.3% to about 6.5%. The risk is higher in patients with disseminated DLE (22%) than in DLE confined to the head and neck (1.2%) [4].

Clinically, sarcoidosis resembles an ensemble of diseases which may include discoid lupus erythematosus (DLE) or necrobiosis lipoidica among others. Diagnosis difficulties because of clinical similarities between cutaneous lesions of sarcoidosis and DLE have been reported

Author Affiliations
Farah Sameem MD , Assistant Professor ;**Seerat Fatima** MBBS, Postgraduate resident, **Sheikh Manzoor** MD ,Professor & Head : Department of Dermatology, Venereology, and Leprosy SKIMS MC-H Bemina,
Sheikh Javeed Sultan MD, Assistant Professor ,Department of Dermatology,GMC Srinagar, Kashmir,

Correspondence:
Dr.Seerat Fatima
Postgraduate Resident,Department of Dermatology, SKIMS MC-H Bemina, Srinagar, 190018
J&K, India.
rizviseeratfatima@gmail.com

Indexed:
Scopus ,INMED , EBSCO @ Google Scholar among others

Cite This Article as:
Sameem F , Fatima S , Manzoor S. A Diagnosed Case Of Sarcoidosis Presenting With Dle (Discoid Lupus Erythematosus): JKPract2021;26(1): 55-57

Full length article available at **jkpractitioner.com** one month after publication

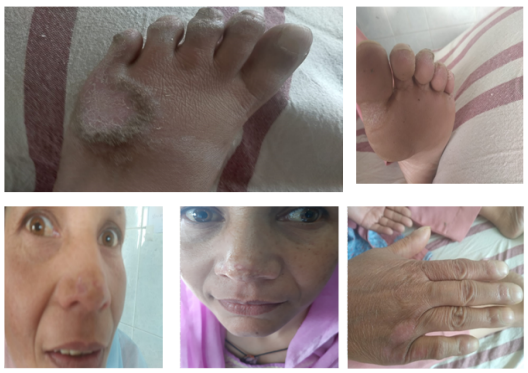
Key words: sarcoidosis , discoid lupus, erythematosus, co-exixtent dermatosis

since 1966, and the histopathological examination is necessary for establishing this diagnosis. The association of sarcoidosis with DLE is poorly documented in literature. Hence, we are reporting a case of sarcoidosis presenting with DLE.

CASE REPORT:

A 37-year-old female presented to the consultant Outpatient Department of Dermatology SKIMS MCH Bemina with the development of cutaneous lesions in a period of past 8-10 months. The patient was a labeled c/o sarcoidosis with CXR (chest x ray) and HRCT (high resolution computed tomography) chest showing Stage ii pulmonary involvement and was on treatment for the same (systemic corticosteroids). Other investigations included normal 1. EBUS (endo-bronchial ultrasound), 2.FNAC (fine needle aspiration cytology) -> lymphocytes with histiocytic collection – no granulomas).

The patient presented with the development of well demarcated, erythematous, scaly, annular to discoid plaques on the dorsum of her hands ,feet, fingertips, toes and some on the nose(mainly on photo-exposed areas)with slightly adherent scales and central atrophy and hypo pigmentation.



FIGURES 1: WELL-DEMARCATED ERYTHEMATOUS ANNULAR PLAQUE WITH SCALY SURFACE ON DORSUM OF THE FOOT, HANDS, NOSE AND TOES HEALING WITH SCARRING (CENTRAL ATROPHY AND HYPO PIGMENTATION)

The patient was investigated on lines of sarcoidal and DLE like lesions.

A skin biopsy was performed and HPE (histopathology) favoring DLE (Discoid Lupus Erythematosus) showed the following features: -

1. Lymphohistiocytic infiltrate the DEJ with prominent capillaries and unremarkable endothelial cells
2. Focal basal cell degeneration and colloid bodies
3. No granulomas were seen
   ANA, ENA (dsDNA), ACE levels came out to be negative.

Taking the clinical picture with histological confirmation into consideration a diagnosis of sarcoidosis with DLE (Discoid Lupus Erythematosus) was made and the patient was managed in lines of the same.

DISCUSSION:

DLE (Discoid Lupus Erythematosus) presents as erythematous plaques, which tend to regress spontaneously, causing scars and mainly affecting the face, scalp, ear and neck and in disseminated disease may extend to involve the trunk and extremities. In the present case, the synchronous occurrence of clinical features of both sarcoidosis and connective tissue disorder(DLE) suggests that a probable coexistence of LED and sarcoidosis cannot be ruled out, since both diseases can occur simultaneously or mimic one another[5].Previous genetic studies have established that variants in class I and II locus of human leukocyte antigens (HLAs) play roles in the susceptibility of developing sarcoidosis, as well as other autoimmune diseases, including lupus[6]. Additionally, evidence suggests that, given the similarities in the immunophenotyping profiles of patients with DLE and SLE, the pathogenesis of these two disorders also present resemblance between themselves[7].

The first clinical association between SLE and sarcoidosis was described in 1945, in which two patients with SLE presented non-caseous granulomas in the lungs, lymph nodes and blood vessels at the autopsy, thus suggesting the relationship between both diseases[8]. Since then, there have been reports of many cases that propose this simultaneous occurrence. The histopathological findings of sarcoidosis are independent of the involved organ or the clinical presentation of the lesion. The epidermis is usually not involved, while the dermis manifests a superficial and deep infiltrate of granulomatous formations, composed of epithelioid cells and surrounded by sparsely arranged lymphocytes (also known as the naked granulomas). Also, histopathological findings alone do not accurately differentiation between a sarcoid reaction and true sarcoidosis.

Being a diagnosis of exclusion, there are no well-established criteria for a diagnosis of sarcoidosis. Despite the various clinical manifestations inherent to this disease, histological confirmation is required in most of the cases with some exclusion for diagnosis. Heinle and Chang[9] proposed, in 2014, major (presence of non-caseous granulomas and absence of acid-alcohol resistant bacillus on biopsy) and minor (erythema nodosum, hypercalciuria, anemia, pancytopenia, cardiac arrhythmia, hilar adenopathy

on chest radiography, uveitis, spondyloarthritis, elevated immunoglobulins and liver enzymes and bronchoalveolar lavage findings) criteria, in order to guide clinical evidence to a more accurate diagnosis [10]. Also, ACE levels are seen to be raised in many of the patients.

As a result of its clinical heterogeneity, there are no well-established protocols for treatment of the disease, and corticotherapy is mostly indicated for cases of severe ocular, pulmonary involvement in stages 2 and 3, neurological, renal, cardiac and cutaneous manifestations, as well as splenomegaly.

DLE and several other diseases that resemble cutaneous sarcoidosis are relatively benign. Because of its characteristic clinical appearance and infrequent association with systemic lupus erythematosus, therapy for DLE is often administered without histological confirmation of lesions[4].This precludes identification of its close clinical simulation, cutaneous sarcoidosis. Hence a skin biopsy must be performed to confirm the diagnosis of DLE and to exclude sarcoidal skin lesions. Because of the systemic and progressive nature of sarcoidosis, it is critical that this distinction is made and that the patient be treated accordingly[4].

Hence in our case the appearance of new skin lesions was fully evaluated, even though she was a known c/o sarcoidosis to rule out other clinical differentials (DLE in our case) and also cutaneous annular plaque type sarcoidosis which holds a comparatively bad prognosis to other cutaneous forms of sarcoidosis and may have called for a more aggressive treatment regimen. Such simultaneous occurrence of sarcoidosis and Discoid Lupus Erythematosus (DLE) also demands a need for increasing research into the concurrence and association of the two conditions and the effect it may or may not have on the course of either of the two diseases.

REFERENCES:

1. 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.
2. Veien NK, Stahl D, Brodthagen H. Cutaneous sarcoidosis in caucasians. J Am Acad Dermatol 1987;16:534–40.
3. Olive KE, Kataria YP. Cutaneous manifestations of sarcoidosis: relationship to other organ system involvement, abnormal laboratory measurements, and disease course. Arch Intern Med 1985; 145:1811–14.
4. Mark Goodfi eld1, Jan Dutz2 and Collette mccourt2.Lupus Erythematosus.In: Griffiths C, Barker J, Blieker T, Chalmers R, Creamer D, editors. Rook's Textbook of Dermatology. 9th ed. Wiley Blackwell;2
5. Henderson CL, Lafleur L, Sontheimer RD. Sarcoidal alopecia as a mimic of discoid lupus erythematosus. J Am Acad Dermatol.2008; 59:143-5.
6. Sweiss NJ, Patterson K, Sawaqed R, Jabbar U, Korsten P, Hogarth K, et al. Rheumatologic manifestations of sarcoidosis. Semin Respir Crit Care Med. 2010; 31:463-73.
7. Wouters CH, Diegenant C, Ceuppens JL, Degreef H, Stevens EA. The circulating lymphocyte profiles in patients with discoid lupus erythematosus and systemic lupus erythematosus suggest a pathogenetic relationship. Br J Dermatol. 2004; 150:693-700.
8. Teilum G. Miliary epithelioid-cell granulomas in lupus erythematosus disseminatus. Acta Pathol Microbiol Scand.1945; 22:73-9.
9. Heinle R, Chang C. Diagnostic criteria for sarcoidosis. Autoimmun Rev. 2014; 13:383-7.
10. Suresh L, Radfar L. Oral sarcoidosis: a review of literature. Oral Dis. 2005; 11:138-45.