Cutaneous sarcoidosis without systemic involvement: A modest face of the 'great imitator'.

Ghazala Butt[1*], Aimen Maqsood[2], M. Arsalan Ibrahim[2], Muhammad Umer Mukhtar[2], Muhammad Uzair[3], Uzma Malik[4]

[1]Department of Dermatology, King Edward Medical University, Pakistan

[2]King Edward Medical University, Pakistan

[3]Department of Dermatology, Khyber Medical College, Pakistan

[4]Department of Medicine, King Edward Medical University, Pakistan

Abstract

Sarcoidosis is a systemic inflammatory disease characterized by non-caseating granulomas of multiple organs in the body resulting in a myriad of symptoms. Amidst the unpredictable, multisystem presentation of this disease, cutaneous symptoms that develop early in the course of the disease, acquire a particular significance as a source of timely and early diagnosis. Here we describe a case of a fifty-five-year-old lady with sarcoidosis that manifested as an isolated cutaneous disorder. This article not only reports this occurrence that has been rarely reported in literature but also reviews the cutaneous manifestations of sarcoidosis relevant to the dermatologist.

Keywords: Sarcoidosis, Granulomas, Erythema nodosum, Lupus pernio.

Accepted on July 31, 2020

Introduction

Sarcoidosis is a systemic inflammatory disease characterized by non-caseating granulomas. The disease targets multiple organs such as lungs, skin, heart, eye, and liver resulting in a myriad of pulmonary, cutaneous, cardiac and renal symptoms that can be severely debilitating. Sarcoidosis is a rare disease with a prevalence of 10 per 100000 persons affected every year. Thus, the disease has an unpredictable, multisystem presentation. The cutaneous symptoms that develop in 25% (about one-third) of the patients are apparent early in the course of the disease and hence can serve as the source of timely diagnosis and intervention. The etiology of sarcoidosis is unknown and although its development has been associated with various environmental, genetic, immune and microbial factors [1,2]. Here, we describe a case of a fifty-five-year-old lady with sarcoidosis that manifested as an isolated cutaneous disorder.

Case Report

A fifty-five-year-old lady presented to the Dermatology department of Mayo Hospital with complaints of erythematous, indurated plaques on the face. The patient was in a usual state of health two years back when she started developing erythematous nodules on the face that gradually progressed and lead to the erythematous, indurated plaque formation around the mouth, over the skin of the mandible (Figure 1). Lips were also swelled. There was no history of facial trauma, topical medication or surgery. Nail, scalp, oral mucosae were also examined and found to be unremarkable.

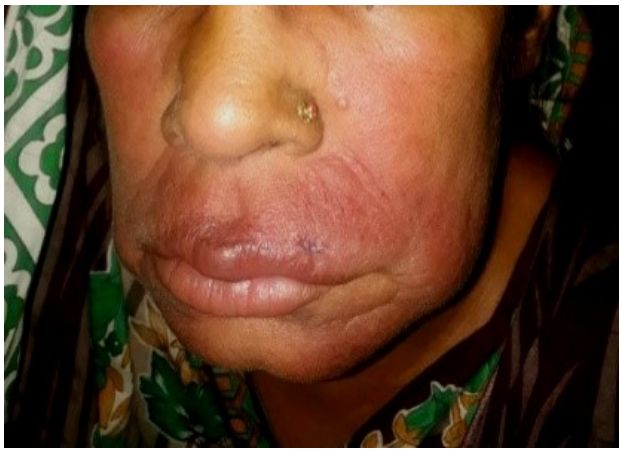

Figure 1. *Erythematous, indurated plaques around the mouth; lip swelling.*

Diascopy showed an "apple jelly" color. Sample for histopathology was taken by punch biopsy. Histopathology showed naked, sarcoidal noncaseating granulomas characteristic of sarcoidosis (Figure 2). Angiotensin-converting enzyme, C- reactive protein, and erythrocyte sedimentation rate were elevated. Suspecting sarcoidosis, extensive laboratory tests, systemic and radiological examinations were

performed. The chest X-ray and HRCT showed old, fibrotic changes in the lungs. The cause of these pulmonary changes was not sarcoidosis but pulmonary tuberculosis that the patient had eight years back. No systemic involvement except the skin was found. Other diseases causing non-caseating granulomas were carefully excluded. Thus, on the basis of histopathology (gold standard diagnostic criteria) and the elevated ACE (potential diagnostic test), a diagnosis of sarcoidosis was made.

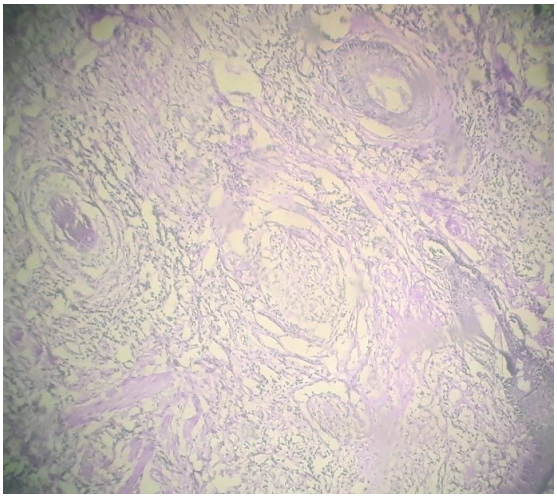

Figure 2. *Multiple naked granulomas characteristic of sarcoidosis. 100X H&E. The patient was started on steroid therapy. Cutaneous erythema, indurations and lip swelling resolved in three weeks.*

Discussion

Sarcoidosis is an unpredictable chronic, inflammatory, granulomatous disease disease causing symptoms according to the specific organ that the disease afflicts [3]. General symptoms include low-grade fever, malaise, weight loss, whereas specific symptoms indicating specific organ involvement include respiratory problems, pulmonary fibrosis (pulmonary involvement), cirrhosis, hepatic nodules (liver involvement), uveitis, conjunctivitis (eye involvement), heart blocks, cardiac failure (heart involvement), encephalopathy, meningitis, cranial nerve paralysis, diabetes insipidus (neuronal involvement), polyarthralgia and osteolytic lesions (musculoskeletal involvement). Thus, it can be concluded that the disease is multifarious and severely debilitating [4].

Skin is the second most commonly involved organ, second only to the lungs. Skin lesions that occur in about one-third of the patients are usually an early and apparent manifestation of the disease. The risk of development of systemic involvement in patients with cutaneous limited disease is unknown [5]. Sarcoidosis is known as the “Great Imitator”, a term signifying the fact that the cautaneous findings mimick many skin conditions. The classic cutaneous manifestation of sarcoidosis is non-caseating granulomas in the skin. There are several forms of cutaneous sarcoidosis on the basis of features of the cutaneous lesions: Papular sarcoidosis (the most common form) is characterized by papules on head, neck and the nasolabial folds and is associated with good prognosis. The maculopapular form is associated with an increased risk of systemic involvement. Plaque sarcoidosis is another common form. Lupus pernio, a special form is characterized by indurated, raised purple lesions on the face. Scar sarcoidosis results from granulomatous inflammation of tattoos, surgical or trauma scars.

Erythema nodosum is a common nonspecific lesion characterized by erythematous, tender, subcutaneous nodules on the shins [6].

Various other forms of cutaneous sarcoidosis also exist: subcutaneous (Darier-Roussy sarcoidosis), angiolupoid, hypopigmented, ulcerative, psoriasiform, verrucous, ichthyosiform or erythrodermic, mucosal (buccal and genital) and nail forms of sarcoidosis. All these are fairly self-explanatory as far as the character of the lesion is concerned and thus undeserving of individual descriptions [3,7]. There are still more indistinct forms and these have been reported rarely in literature.

Corticosteroids administered (topical or intralesional) remain the treatment of choice for cutaneous sarcoidosis. Severe, systemic sarcoidosis or cutaneous sarcoidosis refractory to topical medication may require systemic corticosteroids. Second-line therapy includes antimalarials and cytotoxic agents like methotrexate, azathioprine, leflunomide. Biologic agents like infliximab constitute third-line therapy. Several alternative therapies are also available. Reactivation of sarcoidosis on surgical attempt to remove an inactive sarcoid scar is particularly noteworthy [8].

Thus, in this multisystem, unpredictable disease, the early appearing cutaneous manifestations acquire a special significance. Thus, a well-informed diagnosis based on these cutaneous lesions by the dermatologist will not only lead to timely systemic investigations but may also prompt interventions that may be lifesaving.

References

1. Noe MH, Rosenbach M. Cutaneous sarcoidosis. Curr Opin Pulm Med. 2017;23:482-6.
2. Zhao S, Wang Q, Cheng B et al. Rare scar sarcoidosis: A case report. Exp Therap Med. 2017;4:1535-1537.
3. Karadağ AS, Parish LC. Sarcoidosis: A great imitator. Clinics in Dermatology. 2019 ;37:240-54.
4. Bargagli E, Prasse A. Sarcoidosis: A review for the internist. Intern Emerg Med. 2018;13:325-31.
5. Mañá J, Marcoval J. Skin manifestations of sarcoidosis. La Presse Médicale. 2012 ;41:355-74.
6. Yavuz GÖ, Yavuz İH, Aktar R, et al. A case of sarcoidosis diagnosed via skin involvement. Turkderm. 2019;53:78
7. John AM, Lee BW, Schwartz RA. Cutaneous sarcoidosis. Dermatol Cryosurg Cryotherap. 2016;40: 583-588.
8. Heath CR, David J, Taylor SC. Sarcoidosis: Are there differences in your skin of color patients? J Am Acad Dermatol. 2012;66:121-e1.

***Correspondence to**

Ghazala Butt

King Edward Medical University

Mayo Hospital

Pakistan

E-mail: ghazalakashmiri@gmail.com

Tel: 923334306199