

## CASE REPORT

### Cutaneous Sarcoidosis - the master mimicker : A report of two cases

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#### ABSTRACT

Sarcoidosis is a multisystem granulomatous disease, with an inadequately defined etiology affecting several organ systems. Cutaneous lesions provide important visible clues and an easily accessible source of tissue for histologic examination to reach early diagnosis and management. Along with a brief literature review, we report two cases presenting with rare forms of psoriasiform and annular sarcoidosis, apprising clinicians that cutaneous sarcoidosis called “the master mimicker” due to lesional polymorphism, could be mistaken for common dermatosis like psoriasis or granuloma annulare.

KEY WORDS: Cutaneous sarcoidosis, master mimicker, psoriasiform, annular

#### INTRODUCTION

Sarcoidosis is an idiopathic multisystem granulomatous disease that can involve almost any organ system, the most common being lungs, eyes, lymph nodes and skin. In Greek “Sarco” means “flesh”, “eidos” means “like” and “osis” means “condition”, hence Sarcoidosis translates into flesh-like condition. The first unequivocal case in the English literature was described by Boeck in 1899, although Besnier in 1889 and Hutinchson in 1898 had proposed the term “lupus pernio” and “Mortimer’s malady” after his famous patient, respectively.<sup>1</sup> A third of patients present with only skin lesions, without any systemic involvement. The vast variety of cutaneous morphologies of sarcoidosis, pose a challenge to diagnose it correctly. The lesions of sarcoidosis mimic various common dermatologic conditions, causing great confusion in diagnosing “the master mimicker - cutaneous sarcoidosis”. Besides a brief literature review, we present two cases, with rare presentations of cutaneous sarcoidosis without systemic involvement, mimicking common dermatologic conditions.

#### CASE 1

A 45-year-old Kuwaiti female presented with multiple reddish and mildly scaly skin lesions over both legs, arms, chest and upper back, increasing in number and size over one year duration. They were associated with mild itch, but not with exacerbation by trauma, medication, stress or infection. Cutaneous examination revealed multiple, well defined, discrete, erythematous, scaly papules and plaques over both upper and lower limbs (Fig. 1, 2), predominantly on the extensor surfaces, bilaterally over breasts (Fig. 3, 4) and upper back (Fig. 5), extending on to the back of neck. At places papules coalesced to form plaques with many of them showing fine, silvery, semiadherent scaling, suggestive of psoriasis, but Auspitz sign was negative. She had been repeatedly diagnosed as psoriasis in the past, for which she had received various topical steroids, keratolytic agents and emollients without much improvement. There was no history of any other cutaneous lesion, joint pains, mucosal, nail or systemic involvement. General physical and systemic examination was normal. Past and family history was not contribu-

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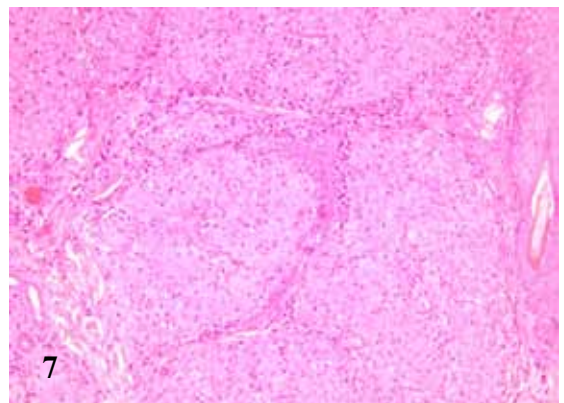
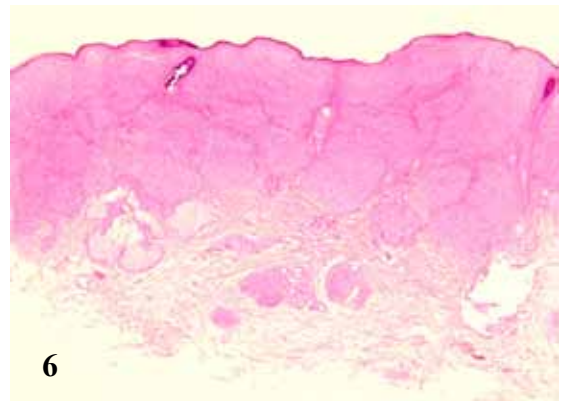


**Fig. 1, 2** Erythematous papules and plaques with scaling on extensor surface of arm and legs, resembling psoriasis.



**Fig. 3, 4, 5** Erythematous papules, some covered with fine silvery scales, over upper back and bilateral breasts.

tory. Skin biopsy revealed classical noncaseating epitheloid cell granulomas of sarcoid type (Fig. 6, 7), with fungal and acid fast bacillus staining negative and reticulin stain for sarcoidosis being positive.



**Fig. 6, 7** Low and high power photomicrograph showing classical noncaseating epitheloid cell granulomas of sarcoid type.

Routine hematologic and urine investigations were all normal. Serum Angiotensin converting enzyme (ACE) titer was raised to 105 U/L. Complete blood count was normal except for raised ESR. Whereas serum calcium, urinary calcium, liver function tests (LFT), renal function tests (RFT), ultrasound (USG) abdomen as well as chest X-ray were normal. Diagnosis of psoriasiform cutaneous sarcoidosis without systemic involvement was made and the patient was started on hydroxychloroquine 200mg twice daily. The lesions started responding within a month, with complete resolution in 3 months, without any recurrence for 6 months post-treatment.

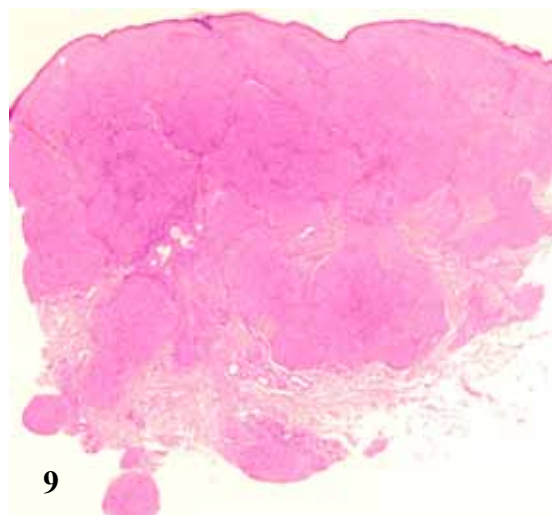
## CASE 2

A 39-year-old Pakistani male presented with a small, single, asymptomatic lesion at the root of the nose for 5 months. On examination, there was single 1×1 cm well defined annular, nontender plaque with smooth, raised, beaded, erythematous, shiny border and central atrophy, associated with a very faint telangiectasia (Fig. 8). He had no history of trauma, drug intake or systemic complaints. Sensations within the plaque were normal. Punch biopsy was performed keeping granuloma annulare as the provisional diagnosis, as well as to rule out basal cell carcinoma. But the histopathological examination revealed epithelioid cell, non-caseating, naked, sarcoidal granulomas mainly in the dermis (Fig. 9) with reticulin stain showing intact network of reticulin fibers. Routine blood and urine tests along with serum calcium, ACE levels (56 U/l), urinary calcium, chest X-ray and USG abdomen were all within normal limits. The patient was started on hydroxychloroquine 200mg orally bid for the final diagnosis of annular sarcoidosis. The lesion started resolving within a

month of initiating treatment, although the patient was lost for further follow-up.



**Fig. 8** Annular plaque with smooth, raised, beaded, erythematous, shiny border and central atrophy.



**Fig. 9** Epithelioid cell, non-caseating, naked, sarcoidal granulomas mainly in the dermis.

## DISCUSSION

Sarcoidosis occurs worldwide, with the highest prevalence in Scandinavian region. Sarcoidosis is the result of an immune dysfunction due to persistent but inadequately defined etiological antigen that is poorly cleared by the immune system.<sup>1</sup> Cutaneous involvement is known to occur in 20-35% of patients of sarcoidosis, with 10% having only exclusive involvement of the skin.<sup>2</sup> Fifteen percent of patient had skin involvement in an ear-

lier study on 20 patients from Kuwait.<sup>3</sup> Cutaneous sarcoidosis can manifest as specific lesions, which contain granulomas and non-specific lesions, which are the result of reactive processes. Specific lesions follow a more chronic course, often with systemic involvement. They typically appear as maculopapular eruptions, subcutaneous nodules, lupus pernio, infiltrated plaques and infiltrations of old scars or tattoos. Whereas nonspecific manifestations include erythema nodosum, erythema multiforme, prurigo and calcifications. Erythema nodosum, a non-specific reactive process, is the hallmark of an acute and benign disease associated with good prognosis.

Our first case of psoriasiform sarcoidosis masquerading as psoriasis, is an accepted but rare morphological presentation of sarcoidosis. Klauder gave the first clinical description of psoriasiform sarcoidosis and Elgant stated it to be peculiar to Negroes with no report of such lesions in European literature.<sup>4</sup> Psoriasiform eruptions are found in 0.9% of cases of sarcoidosis<sup>5</sup> and it occurs when the granulomatous pathology affects the epidermis leading to a clinical resemblance to psoriatic plaques with scaling.<sup>6</sup> A similar case report by Al-Hoquail,<sup>7</sup> is the only other case of psoriasiform sarcoidosis reported from the Arabian Gulf region. Other atypical forms of cutaneous sarcoidosis described in literature are ulcerative sarcoidosis, hypopigmented sarcoidosis, ichthyosiform sarcoidosis, morpheaform sarcoidosis, lichenoid lesions, cicatricial alopecia, lupus erythematosus-like lesions, verrucous and papillomatous lesions.<sup>8</sup>

Our second case, having an annular plaque at the root of the nose turned out to be annular sarcoidosis masquerading as granuloma annulare. Apart from granuloma annulare, Hansen's disease and

annular basal cell carcinoma are the other differential diagnosis usually considered for annular plaques with indurated borders appearing on the face. Reddish brown sarcoidal papules can coalesce to form annular plaques mimicking granuloma annulare.

Our second patient had normal ACE levels. Bunting et al<sup>9</sup> reported that increased serum ACE level is not specific for sarcoidosis, with a sensitivity and specificity of 77% and 93% respectively, although it has been used as an important laboratory test since 1975. ACE levels in serum are derived from epitheloid cells of the granulomas, reflecting the granuloma load in the patient. Hence, the ACE level was normal in our second patient, who had a single small annular lesion. ACE levels are neither diagnostic nor predictive of systemic involvement, although they are useful to monitor the clinical course of sarcoidosis. They are elevated in 60% of patients and are useful to determine the activity of disease.<sup>1</sup> There is no single confirmatory test for sarcoidosis. Patients are diagnosed as having sarcoidosis based on histological evidence of noncaseating granuloma with exclusion of other potential causes such as infections along with a compatible clinical or radiological picture, when present.<sup>4</sup> Typical sarcoidal granuloma are discrete, distributed uniformly in the dermis and surrounded by sparse lymphocytic cuffing (naked tubercles), with fine reticulin fibres in and around the tubercles.<sup>1</sup>

A previous study has revealed that approximately 30% of patients who initially had only cutaneous lesions developed systemic involvement months to years later.<sup>2</sup> Therefore, every patient presenting with cutaneous sarcoidosis alone requires an initial work-up, followed by periodic screening, for the possible development of systemic manifesta-

tions at a later date. Cutaneous involvement in sarcoidosis, is most often present at the onset and may even be the presenting complaint. Awareness of these varied and at times rare presentations of cutaneous sarcoidosis is very essential, as cutaneous lesions provide important visible clues and are an easily accessible source of tissue for histologic examination, to reach early diagnosis and management.

To conclude, cutaneous sarcoidosis is a great mimicker due to lesional polymorphism and could be mistaken for common dermatoses like psoriasis or granuloma annulare, as reported in our cases, inferring that it should be kept in mind even while dealing with such common dermatological diseases.

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