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Case Report

Sarcoidosis – the great imitator: report of two cases

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Abstract Sarcoidosis is a multisystem granulomatous disease and has aptly been referred to as the great imitator in view of the myriad of lesions it presents with and of varying morphology. We report two such cases where cutaneous lesions of sarcoid was not the initial presentation and patients were being evaluated for other systemic symptoms..

Key words
Sarcoidosis, granulomatous disease.

Introduction

Sarcoidosis is a systemic non-caseating granulomatous disorder of unknown origin that primarily affects the lungs but often involves the skin. In Greek, Sarcoidosis means flesh-like condition. "Sarco" meaning "flesh", "eidos" meaning "like" and "osis" meaning "condition".[1] Besnier in 1889 reported what probably was the first patient with sarcoidosis and proposed the term "lupus pernio". Hutchinson described more cases and called it "Mortimer's malady" after his famous patient. The first unequivocal case of sarcoidosis in the English Literature was reported by Boeck in 1899.When the lesions were examined diagnosis was struck by close resemblance to sarcoma – thus the name "benign sarcoid".[2]

Case reports

Case 1 A 66-year-old female, admitted for evaluation of hepatosplenomegaly, for a duration of 1 year, presented with scaly lesions over the body. On cutaneous examination, erythematous scaly annular plaques were present over the left lower back with similar lesions over both legs and nose (**Figures 1** and **2**). Her cervical lymph nodes were enlarged.

All routine hematological investigations were normal except for raised ESR. Chest X ray showed subtle non-homogenous opacities in the right lower zone. Mantoux test was negative. Ultrasonogram of abdomen showed massive hepatomegaly and moderate splenomegaly. Lymph node biopsy from cervical lymph nodes showed non-caseating granulomas composed of epithelioid cells, scattered Langhan's cells and lymphocytes. Hamazaki Wesernberg bodies, Schaumann bodies and Ca^{+2} oxalate crystals were seen in the cytoplasm of giant cells suggesting a diagnosis of non-caseating granulomatous lymphadenitis in the absence of AFB. Skin biopsy was consistent with the diagnosis of a non-caseating granulomatous lesion (**Figures 3** and **4**).

Case 2 A 59-year-old male with a past history of psoriasis on methotrexate 10mg/week was being investigated in the Medicine OPD for loss of weight and appetite. He was referred to us with

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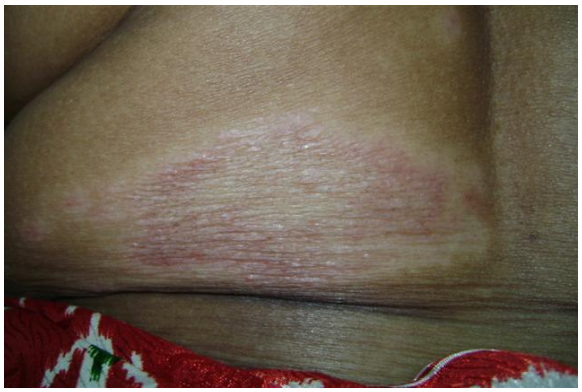

Figure 1 Erythematous scaly annular plaque on the back.

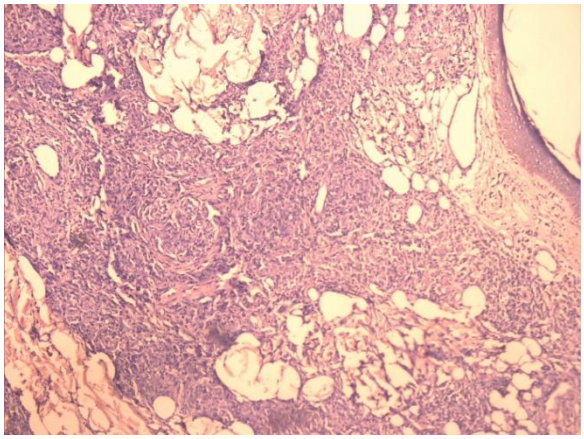

Figure 3 Photomicrograph showing granulomatous infiltrate in the dermis.

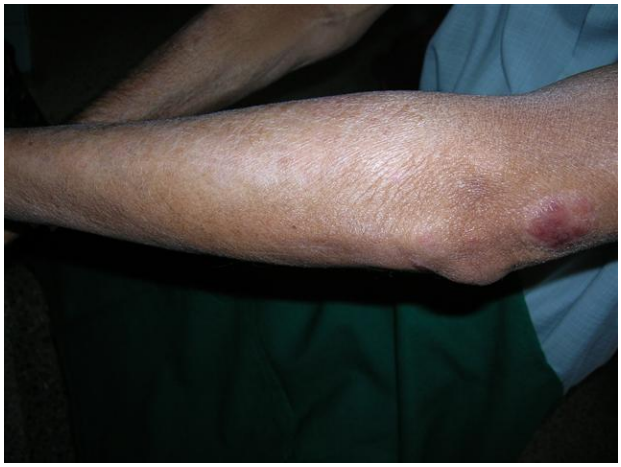

Figure 5 Hypopigmented macules and infiltrated plaques on the elbow.

skin lesions comprising of hypopigmented macules and infiltrated plaques present around the elbows (**Figure 5**). Complete blood picture

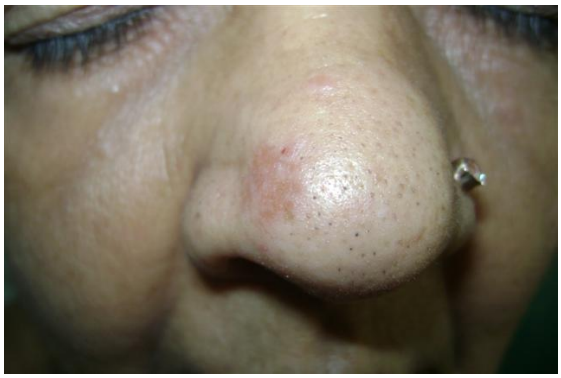

Figure 2 Erythematous annular plaque on nose.

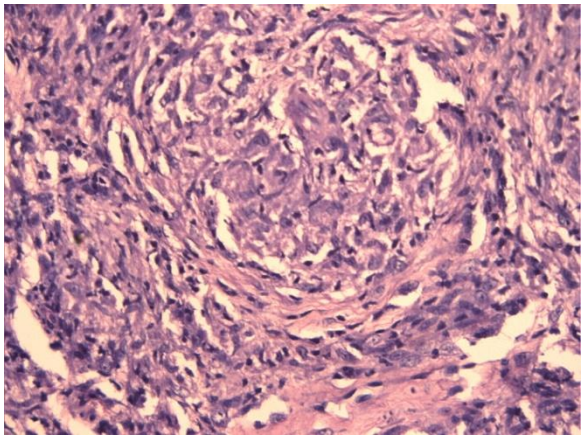

Figure 4 Photomicrograph showing the close up of the granuloma (20X).

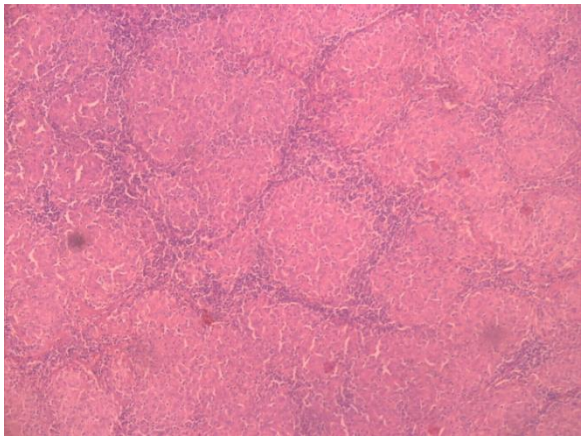

Figure 6 Photomicrograph showing non-caseating lymph adenitis (10X).

was normal except for raised ESR. 24 hours urinary Ca^{+2} levels and Serum ACE levels were elevated. Chest X-ray showed bilateral hilar prominence and non homogenous opacities in both lung fields. Mantoux test was negative. USG showed massive splenomegaly, moderate

hepatomegaly and lymphadenopathy. Lymph node biopsy (**Figure 6**) as well as skin biopsy revealed non-caseating granulomas and was negative for acid fast bacillus. Reticulin stain for sarcoid was positive.

Discussion

Sarcoidosis has rightly been called "the great imitator" as it presents with a myriad of lesions of varying morphology. Cutaneous involvement occurs in 10-35% of patients of sarcoidosis, with only 2% having exclusive involvement of the skin.[3] Cutaneous sarcoidosis can manifest with specific and non-specific lesions. Specific lesions contain granulomas whereas non-specific lesions are reactive processes.[4] Specific lesions are maculopapular eruptions, subcutaneous nodules, lupus pernio, infiltrated plaques, and infiltration of old scars. The specific lesions are associated with more severe systemic involvement and a more chronic course whereas erythema nodosum, the only nonspecific manifestation of sarcoidosis is the hallmark of an acute and benign disease.

Maculopapular eruptions are the most common type of granulomatous cutaneous involvement in Sarcoidosis. Maculopapules are only slightly infiltrated, with little epidermal change, usually red brown to purple and less than 1 cm in diameter. They are most commonly seen on the face, particularly on the eyelids, around the orbits, and in the nasolabial folds.

Many atypical skin lesions have been described in sarcoidosis such as extensive ulcerative lesions, psoriasiform plaques, hyperpigmentation in black patients, verrucous and papillomatous lesions, ichthyosiform lesions, pustular folliculitis, papules in light exposed areas, lichenoid eruptions, erythrodermic eruption, cicatrical alopecia, lupus erythematosus-like lesions, mutilating lesions, erythema and plaques involving palms and soles, pruritus caused by the granulomatous skin eruption and a rare form of cutaneous sarcoidosis with diffuse skin plaques, associated with uveitis and arthritis seen in children younger than 4 years. All these granulomatous skin lesions can be accompanied by features of systemic sarcoidosis, but their prognostic significance is difficult to assess.

Specific cutaneous lesions may be the first complaint of patient or may be a finding in physical examination in the initial assessment of systemic disease. Atypical cutaneous lesions like the ones reported here may masquerade as or simulate other skin conditions such as psoriasis, lichen planus, erythroderma, ichthyosis etc.

Olive and Katarin noted in a chart review of 329 patients that specific skin lesions were most likely to have lymphadenopathy and hepatosplenomegaly than in sarcoidosis patients without skin lesions.[5]

In our first case, in addition to the infiltrated plaques, psoriasiform lesions were seen, which are an accepted but rare morphological manifestation of sarcoidosis. Elgant attributed the first clinical description of psoriasiform lesion to Klauder (1925) and states that this type of sarcoidosis is peculiar to Negro, no report of such lesions being found in European literature.[6] Longcope and Freiman in a review of 160 cases, referred to only one case demonstrating psoriasiform appearance.[6] Our second case was suffering from psoriasis who later presented with infiltrated plaques which on histology were proved to be that of sarcoidosis.

To conclude, recognition of cutaneous sarcoid lesions is very important because they provide an important visible clue to the diagnosis and are

an accessible source of tissue for histopathological examination. The diagnosis of sarcoidosis should be based on the comprehensive clinical examination, chest X ray findings, histological demonstration of non-caseating granulomas involving one or more tissues with negative stains or culture for mycobacteria and fungi and exclusion of other granulomatous diseases. In our cases, clinical examination coupled with CXR findings, raised ACE levels and histopathology picture were sufficient to make a diagnosis of sarcoidosis.

References

1. English JC, Patel PJ, Greer KE. Sarcoidosis. *J Am Acad Dermatol* 2001; **44**: 725-43.
2. Francisco A. Kerdel, Samuel L. Moschella SL. Sarcoidosis. *J Am Acad Dermatol* 1984; **11**: 1-19.
3. Mana J, Marcoval J, Graells J *et al*. Cutaneous involvement in sarcoidosis. *Arch Dermatol* 1997; **133**: 882-8.
4. Burov EA, Kantor GR, Isaac M. Morpheaform sarcoidosis: report of three cases. *J Am Acad Dermatol* 1998; **39**: 345-8.
5. Olive KE, Kataria YP. Cutaneous manifestations of sarcoidosis: relationships to other organ system involvement, abnormal laboratory measurements and disease course. *Arch Intern Med* 1985; **145**: 1811-44.
6. Burgoyne JS, Wood MG. Psoriasiform sarcoidosis. *Arch Dermatol* 1972; **106**: 896-8.