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Case Report

Cutaneous sarcoidosis without systemic involvement: Another presentation of a 'master mimicker'

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Abstract Sarcoidosis is an idiopathic, noncaseating, granulomatous, inflammatory disease involving multiple organs. Skin involvement can be the earliest presentation of disease effecting nearly 25 to 30 percent of cases. Cutaneous form of sarcoidosis is considered as the 'great imitator', because it masquerades various other dermatological conditions causing a huge complexity in diagnosis and management of this condition. Moreover, presence of other more prevalent granulomatous infections in our region causes a further delay in diagnosis. Here we present a case report concerning coexistence of differential morphological patterns of sarcoidosis (papules, nodules, psoriasiform and annular) all in the same patient.

Key words
Cutaneous sarcoidosis; Systemic involvement; Master mimicker.

Introduction

Sarcoidosis is described as non-necrotizing granulomatous disease that affects numerous organ in the body. The most commonly effected organs are lungs, skin, hilar lymph nodes and eyes.[1] Prevalence is 10-64 per 100,000 and varies among different geographical areas and races.[2] Females have higher propensity to have sarcoidosis than male. Peak incidence is around 40 years of age.[3] In literature African-Americans and Scandinavians have reported the highest cases worldwide however cases are not quite rare in Pakistan.[4] In recent years more number of cases have been reported relating to the presence of sarcoidosis limited to the skin.[3] Multidisciplinary approach is required for the treatment. Corticosteroids and other immunosuppressant is mainstay of therapy.[1]

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Case Report

An eighty years old male, known case of hypertension, presented to us in OPD with the complaint of multiple papules, nodules and plaques all over the body over the past month. Initially they were only present over his back then gradually increased in size and number, now involving his forearms, legs, face and abdomen. They were associated with mild itching. There was no history of fever, fatigue, night sweats, weight loss, muscle pain, joint pain, cough, shortness of breath, pain or redness of eyes. No history of nasal crusting, epistaxis, muscle weakness, paresthesia or numbness. No history of oral or genital lesions. No personal or family history of tuberculosis. Systemic review was unremarkable.

On examination multiple, erythematous to violaceous, papule, plaques and nodules of varying sizes were present all over the body. Some of these plaques were psoriasiform covered with loosely adherent white scale. These were more dense over his back, forearms, shins

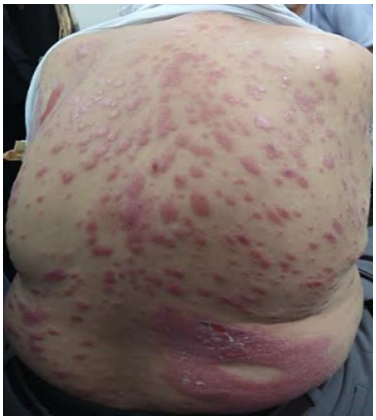


Figure 1

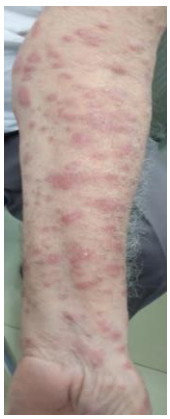


Figure 2

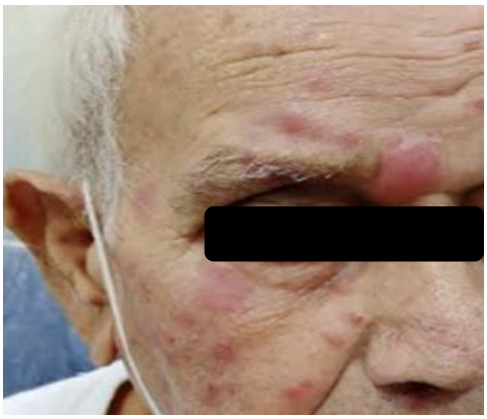
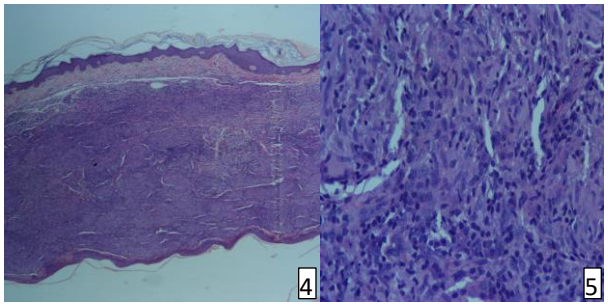


Figure 3

and face and sparse over abdomen, chest and thighs. Largest annular plaque was located over his lower back measuring 5X4 cm in size with well demarcated inner border and ill-defined outer border. It was covered with white, loosely adherent scale **(Figures 1-3)**. Palms, soles, flexures and scalp were spared. On palpation lesions were firm in consistency, no temperature difference, non tender, Diascopy and Auspitz sign were unremarkable. Sensations over the lesion were intact, surrounding nerves and peripheral nerves were not palpable. Slit lamp examination was unremarkable. Systemic examination was unremarkable.

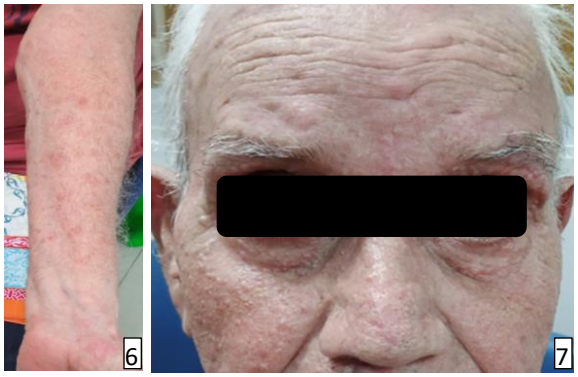
Our differentials were lepromatous/ borderline leprosy, secondary syphilis, sarcoidosis, cutaneous tuberculosis, cutaneous lymphoma, progressive nodular histiocytosis.

Hematological and biochemical investigations were normal. ESR was 8 mm/hour. Chest X-ray was unremarkable. VDRL and TPHA were also within normal limit. Corrected calcium was 9.1 mg/dl and ACE level was raised 140 U/l. Mantoux test was negative. Slit skin smear for Mycobacterium Leprae was negative. Skin biopsy collection of epithelioid histiocytes along with multinucleated giant cells forming granuloma. ZN stain and PAS stain were negative. No necrosis was seen **(Figures 4,5)**.



Figures 4, 5 No necrosis.

Our final diagnosis was cutaneous sarcoidosis. Methylprednisolone aceponate 0.1% cream once daily over the face and Fluocinolone acetonide 0.025% ointment once daily over the body were given along with oral prednisolone 30 mg/day for a month which gradually tapered to 5mg/ day. Fortnightly follow-up with regular measurement of blood pressure, blood sugar and electrolytes and complete blood count were advised. Significant improvement was observed after 1 month of treatment **(Figures 6,7)**.



Figures 6, 7 Significant improvement**.**

Discussion

First case of sarcoidosis was recorded by physician DR Jonathan Hutchinson.[1] It is a chronic granulomatous disease affecting multiple organs. The disease has female predominance affecting about two-thirds women, with the peak incidence recorded around 40 years of age. In United States, African American females have reported higher number of cases but the prevalence is not racial.[1,2] Although the disease is not uncommon in Pakistan there is limited data in the literature. Cases are infrequently reported because of the more common and well recognized granulomatous diseases in this part of the world, like tuberculosis, leprosy and fungal infections.[3]

Even though sarcoidosis is an idiopathic disease, multiple factors have been linked to its etiopathogenesis. Genetic (HLA genes), infective organisms (Mycobacterium and Propionibacterium), environmental and industrial toxins has been studied to have a role in disease pathogenesis.[1]

Most commonly involved organs include lungs, kidney, heart, eyes, skin and lymph nodes. The skin is the second most commonly involved organ after lungs accounting for 25 to 30% of cases. Lung and heart involvement being the leading cause of mortality. Skin involvement can be the first sign of sarcoidosis presenting to clinician and may indicate a need for prompt action to see the presence of underlying systemic disease.[4]

Cutaneous sarcoidosis exhibits two types of lesions, specific and non-specific. Histopathology of specific lesions have sarcoidal granulomas, which are absent in non-specific ones.[2,5,6] Specific lesions include maculopapular and plaque sarcoidosis, lupus pernio, subcutaneous sarcoidosis and scar sarcoidosis. While erythema nodosum is the most important nonspecific skin lesion.[6] Other rare types such as angiolupoid, hypopigmented, icthyosiform, ulcerative, annular and psoriasiform, have also been reported.[3] Cutaneous lesions most commonly involve face, neck, upper back and trunk.[3]

Diagnosis is made on clinical, hematological, biochemical (renal and liver function tests, serum and urine calcium level, ACE level) radiological (Chest X ray, CT scan chest), histopathological features, pulmonary function tests, Broncho alveolar lavage and ECG findings. Presence of specific skin lesions along with Broncho alveolar lavage lymphocytosis, raised CD4:CD8 count and high serum ACE levels are markers of progressive disease.[7]

In general, the aim of the sarcoidosis treatment is to alleviate symptoms, improve quality of life and to prevent organ damage.[8] Treatment is based on severity of disease. For localized and mild cutaneous sarcoidosis topical or intralesional corticosteroids are used as the first-line therapy and generally recommended for disfiguring, cosmetically distressing, and symptomatic disease. Systemic therapy is reserved for cases that do not respond to first-line treatment including oral steroids, antimalarials, methotrexate, tetracycline, thalidomide, and TNF alpha inhibitors, such as infliximab. Surgical intervention by removal of the granulomas from the skin are reserved only for mutilating disease and for cases refractory to drug therapy.[4]

Conclusion

This case highlights the varying morphological presentation of sarcoidosis in an individual. Sarcoidosis which was once considered an uncommon disease is not a rarity now. Real challenge is faced by countries in our region

were other granulomatous conditions are more prevalent such as tuberculosis, leprosy and fungal infections. Cutaneous manifestation can be the first and solitary presentation of sarcoidosis so it should always be kept in consideration in order to save patients from organ damage, unnecessary antibiotics/ antifungal treatment and financial burden.

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Author's contribution

SAA: Diagnosis and management of case, critical review, final approval of the version to be published.

AZ: Diagnosis of case, manuscript writing, final approval of the version to be published.

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