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Case Report

Cutaneous sarcoidosis: a case report from North-East India

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ABSTRACT

Sarcoidosis is a multisystem granulomatous disease of unknown aetiology. Cutaneous sarcoidosis is seen in up to one-third of patients and may be the first or the only clinical sign of the disease. We report a case of cutaneous sarcoidosis with lungs and renal involvement in a 28-year-old housewife. She presented with itchy reddish-brown indurated plaques on left cheek and back for 2 years. Skin biopsy revealed non-caseating epithelioid granulomas. Contrast enhanced computed tomography thorax showed focal consolidation and multiple enlarged mediastinal lymph nodes and serum angiotensin-converting enzyme level was raised. Since cutaneous manifestations of sarcoidosis are extremely variable, its early recognition can provide a clue to the diagnosis and its systemic involvement.

Keywords: Cutaneous sarcoidosis, Granuloma, Non-caseating, Renal calculi

INTRODUCTION

Sarcoidosis is a multisystem granulomatous disease of unknown aetiology involving the lungs, mediastinal and peripheral lymph nodes, eyes and skin. Cutaneous sarcoidosis is seen in around 25-35% of patients and may be the first clinical sign of the disease.[1] It is more prevalent in developed countries, ranging from 10 to 40 per 100000 in the US and Europe.[2] In India, though the exact prevalence is not known, Delhi and Kolkata hospitals have reported 61 and 150 cases per 1,00,000 outdoor patients respectively.[3]

CASE REPORT

A 28-year-old housewife with bilateral renal calculi was referred from urology for itchy erythematous plaques on left cheek and back for 2 years. The skin lesions had been treated elsewhere with anti-tubercular and anti-leprosy medicines without any improvement. She also had history of dyspnoea on exertion and general weakness. Cutaneous examination revealed well-defined annular reddish-brown indurated scaly plaques with intact sensations over left cheek and back (Figure 1 A, B and C). Lymphadenopathy was absent and systemic examination findings were normal.

On repeat skin biopsy, non-caseating epithelioid granulomas with occasional Langhans giant cells were seen in the dermis and there was no evidence of acid fast bacilli (Figure 2). Routine blood investigations showed anaemia. Contrast enhanced computed tomography thorax revealed focal consolidation and multiple enlarged mediastinal lymph nodes. Restrictive pattern was seen in pulmonary function test. Serum angiotensin-converting enzyme (ACE) level was raised but serum and urinary calcium levels were normal. Mantoux test, slit skin smear and fungal culture were negative. Electrocardiogram and ophthalmological examinations were normal. Sarcoidosis was diagnosed based on the clinical, histological, radiological and laboratory findings.

Topical and intralesional steroid along with oral prednisolone 40 mg daily (tapering dose) and hydoxychloroquine 200 mg twice daily were started.

Good clinical response was noticed at regular follow ups (Figure 3 and 4).

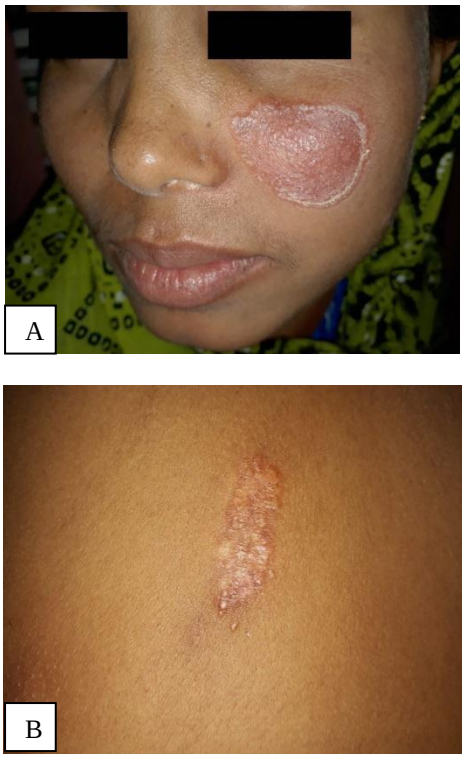

Figure 1: (A): Well-defined annular reddish-brown indurated scaly plaques on left cheek, (B): upper back and (C): lower back.

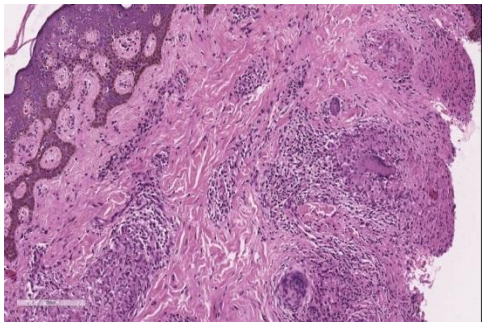

Figure 2: Non-caseating epithelioid granulomas with occasional Langhans giant cells in the dermis (H and E, 40X).

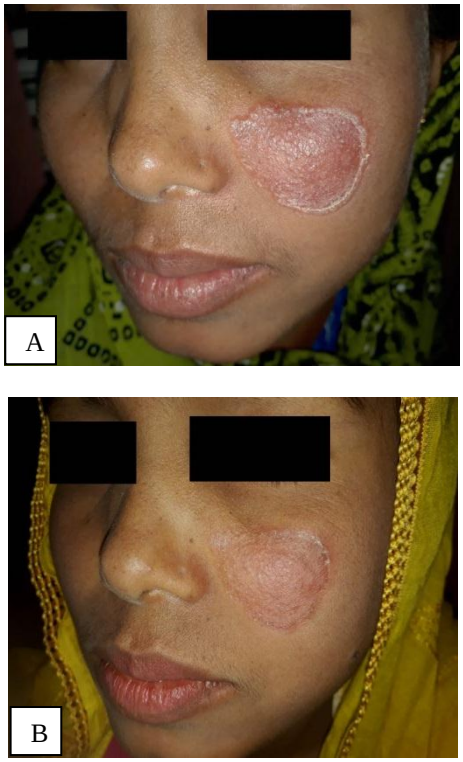

Figure 3: (A) Lesion on left cheek before treatment and (B) after 1 month of treatment.

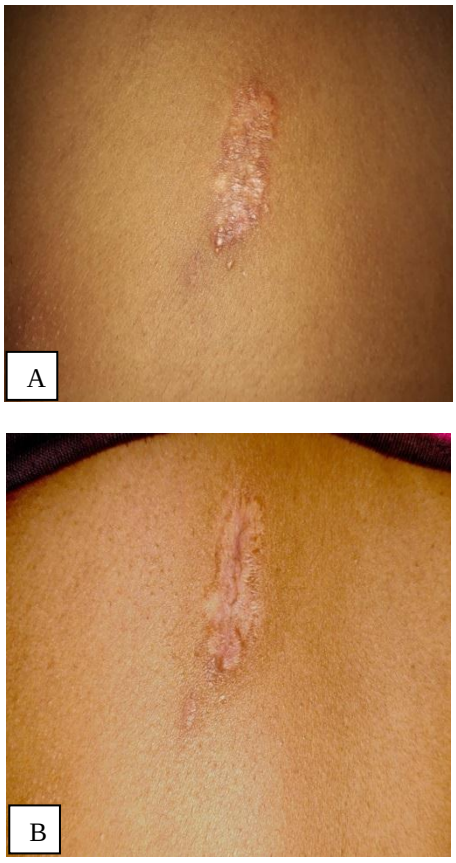

Figure 4: (A) Lesion on upper back before treatment and (B) after 1 month of treatment.

DISCUSSION

Sarcoidosis can present with specific and non-specific cutaneous lesions. Specific lesions include maculo-papules, plaques, lupus pernio, scar-sarcoidosis, subcutaneous sarcoidosis and they display sarcoid granulomas in histopathology. Typical plaque lesions are infiltrated red-brown to violaceous round or oval plaques, favouring the nose, periocular and perioral regions of the face, neck, upper trunk, and extremities. Our case presented with the features of a typical plaque on left upper cheek and back but covered with fine scales. Erythema nodosum is the most common non-specific lesion which is seen in acute sarcoidosis.

Serum ACE level is elevated in around 60% of patients; although not diagnostic it may be useful for monitoring disease progression.[4] Serum and urinary calcium may be elevated.[1] Negative tuberculin sensitivity is a well-known feature of sarcoidosis especially in tuberculosis endemic areas.[5] In our case, ACE level was raised with normal serum and urinary calcium levels.

Most common systemic involvements are lung disease and hilar and/or paratracheal lymphadenopathy which occur in around 90% of patients.[4] Renal calculi have been reported in about 10% of patients with chronic sarcoidosis and the cause may be due to abnormal calcium metabolism leading to hypercalcaemia and hypercalciuria.[6] Our case had lungs involvement with renal calculi.

Sarcoidosis is a diagnosis of exclusion which is based on compatible clinical, histological, radiological picture with negative cultures for mycobacteria and fungus, and exclusion of other granulomatous diseases. Our case was misdiagnosed as lupus vulgaris and leprosy with unsatisfactory treatment response for almost 2 years.

Corticosteroid was the mainstay of therapy. Topical or intralesional corticosteroids are indicated for localized and mild disease limited to the skin and systemic corticosteroids remain the treatment of choice for rapidly progressive, generalized, or highly disfiguring skin disease and systemic involvement.[7] Alternative treatment options include hydroxychloroquine, chloroquine, minocycline, methotrexate and thalidomide.

CONCLUSION

Cutaneous manifestations of sarcoidosis are extremely variable and it is considered one of the 'great imitators' in dermatology.[8] Therefore, a proper clinical history accompanied by histological findings is necessary for its early diagnosis, treatment and timely management of systemic complications.

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