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SARCOIDOSIS – a study of two cases

Dermatology

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ABSTRACT

This article is about two cases of cutaneous sarcoidosis with variable manifestations, where one case had protean manifestations in the form of pulmonary sarcoid while the other had only cutaneous manifestations.

KEYWORDS

Sarcoidosis, protean manifestations, cutaneous

INTRODUCTION:

Sarcoidosis is a great imitator, mimicking a wide variety of conditions1. It's a granulomatous condition which commonly affects the lungs and rarely the skin, eyes, liver, lymph nodes, cardiovascular and central nervous system. Here, we report two cases of sarcoidosis with different presentations.

Case report 1: A 40-year-old male patient presented to the skin OPD with complaints of multiple skin coloured raised lesions on the face, neck and axillae for 4 years. Patient was apparently normal 4 years back after which he noticed dark, flat lesions on the legs and over the next few years he developed the lesions mentioned above. He gave a positive history of intermittent cough for the past 3 years. He reported a right sided facial palsy two years back. On examination, multiple skin coloured as well as pigmented papules were present over the face, neck and axillae. Chest X-ray showed few fibrotic strands in the left middle and upper zone. HRCT lungs showed bilateral hilar lymphadenopathy. Based on pulmonary function test which was suggestive of mild restriction and the pulmonologist's opinion, the patient was diagnosed with pulmonary sarcoid stage 2. Serum angiotensin converting enzyme levels were increased while calcium levels were within normal limits. On histopathology, the epidermis was atrophic with well circumscribed collections of epithelioid cells and scanty lymphocytes.

Case Report 2: A 45-year-old female patient presented to the skin OPD with complaints of a red, raised lesion over the nose for the past 3 years. Initially, the lesion was small and present over the right side of the nose and progressively increased in size to involve the entire nose. History of itching was present. There were no pulmonary or any other systemic complaints. On dermatological examination, an erythematous, scaly, verrucous plaque was seen over the nose. There was no systemic involvement. Serum angiotensin converting enzyme and calcium levels were normal. Suspecting a diagnosis of lupus vulgaris, a biopsy was taken which revealed a picture of sarcoidosis. She was started on steroids but showed no signs of improvement. She was empirically started on anti-tuberculosis therapy. As there was no improvement on completion of 3 months of therapy, but progressive enlargement instead, a repeat biopsy was taken which showed normal epidermis with non-caseating granulomas in the dermis amidst collections of epithelioid cells, few giant cells and lymphocytes.

Based on the above said findings, we made a diagnosis of cutaneous sarcoidosis with protean manifestations for case 1 and a diagnosis of cutaneous sarcoidosis without systemic involvement for case [2].

Discussion: Sarcoidosis is an inflammatory disease affecting multiple organs commonly the lungs, skin and eyes2. Cutaneous manifestations of the disease are classified into specific and non-specific lesions3. Specific lesions will show non-caseating granulomas on biopsy and include lupus pernio, papules, plaques, subcutaneous (Darier-Roussy sarcoid), angiolupoid, verrucous, psoriasiform, lichenoid, ichthyosiform, maculopapular, annular, nodular, scar and ulcerative forms. Non-specific lesions will not show any granuloma formation and includes erythema nodosum, a major part of the disease forming the Lofgren's syndrome.

Systemic manifestations are more common, with lung involvement ranging from hilar lymphadenopathy to pulmonary fibrosis. Ocular involvement4 may be in the form of granulomatous uveitis, conjunctivitis and scleral plaques. Other system involvement includes osseous manifestations, heart block, cranial nerve palsies, diabetes insipidus and hypogonadism[5].

Histopathology will reveal non-caseating granulomas with epithelioid cells but without the mantle of lymphocytes – the naked granulomas. Some cases may also show giant cells, Schaumann bodies and Asteroid bodies.

Other diagnostic tests include chest X-ray, pulmonary function test, HRCT lungs, Gallium scan, elevated serum angiotension converting enzyme and serum calcium levels, and Kviem's test[2].

Sarcoidosis is a diagnosis of exclusion and has to be differentiated from a number of other dermatological diseases.

Sarcoidosis is extremely recalcitrant and steroids remain the mainstay of treatment. Steroid sparing agents like methotrexate, antimalarials, cyclosporine, cyclophosphamide, azathioprine, thalidomide and infliximab2 can also be used.

Conclusion

These two cases are a remainder of the diverse nature of the disease. When a diagnosis of cutaneous sarcoidosis is made, it must be borne in mind that it's rare and an acute condition prone to develop systemic manifestations in the course of the disease.

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Conflict of interest: The authors declare that they have no conflict of interest.

LEGENDS TO FIGURES

Figure 1: Case 1- Clinical image showing multiple papules over the neck and histopathology showing epidermal atrophy with collections of dermal epithelioid cells

Figure 2: Case 2- Clinical image showing an erythematous, scaly plaque over the nose and histopathology showing collections of epithelioid cells in the dermis with few giant cells.

Figure 1

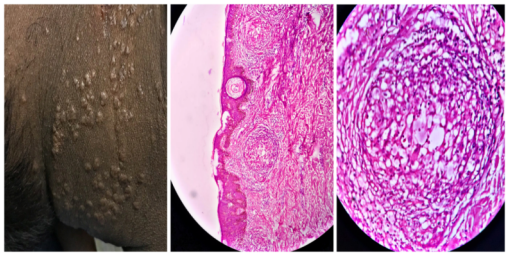
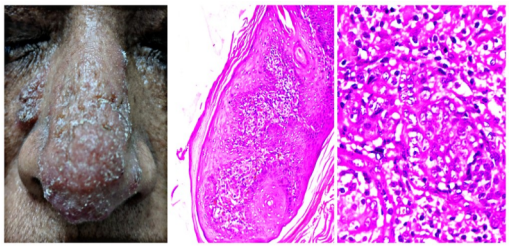


Figure 2



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