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Letter to the Editor

Sarcoidosis – The master mimicker

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Sir,

Sarcoidosis, also known as Besnier-Boeck-Schaumann disease/Mortimer's Malady, is an idiopathic multisystem granulomatous disease, of unknown etiology, that mainly involves lungs, mediastinal and peripheral lymph nodes, eyes and skin, and totally spares the adrenal glands. The disease usually begins at 20–40 years of age, with a second peak of incidence around the age of 60. Nearly two-thirds of the cases are females. Cutaneous lesions of sarcoidosis may be specific, showing histopathologically characteristic "naked granulomas," or non-specific (mainly erythema nodosum). As the lesions assume a vast array of morphologies, cutaneous sarcoidosis is considered as one of the "great imitators" in dermatology. Here, we report the case of a 34-year-old female who presented with multiple morphological patterns of sarcoidosis at the same time.[1,2]

A 34-year-old female without any comorbidities presented with 1 year history of scaly erythematous plaques on the scalp extending toward forehead, that was treated as scalp psoriasis from outside. At presentation, the scalp lesions had already resolved. On examination there were erythematous well-defined plaques with slightly elevated borders and minimal central atrophy over forehead, ears and nose with deformed alae nasi. Multiple hypopigmented to yellowish white xanthelasmata like papules were seen over the upper back, neck. and arms [Figure 1a-c]. Sensations were preserved over the lesions and there was no peripheral nerve thickening. Systemic examination was within normal limits and there was no lymphadenopathy. The differential diagnoses considered were subacute cutaneous lupus erythematosus, sarcoidosis, Jessner's lymphocytic infiltrate, lymphocytoma cutis, Hansen's disease, and eruptive xanthoma (specifically for the upper back lesions). Complete hemogram was normal. Slit and scrape skin smear for *Mycobacterium Leprae* and Mantoux test were negative.

Two biopsy specimens were taken from the face and the upper back respectively, from lesions showing different morphologies. Both of which histopathologically demonstrated well-defined dermal granulomas composed of epitheloid cells without a prominent lymphocytic mantle (naked granulomas) and Langhans giant cells with asteroid bodies. Reticulin staining further showed accentuation of fibers around granulomas extending to the subcutis [Figure 2a-c].

Further investigations showed a normal serum calcium and an elevated serum angiotensin converting enzyme (78.9 U/L). Chest X-ray showed prominent hilar shadows and a subsequent contrast-enhanced computed tomography (CECT) of chest demonstrated nodular opacities bilaterally, subpleural scarring and bilateral hilar, and mediastinal lymphadenopathy.

With the above findings, patient was diagnosed as a case of sarcoidosis. She was started on hydroxychloroquine 200 mg twice a day (considering the photosensitive distribution of the

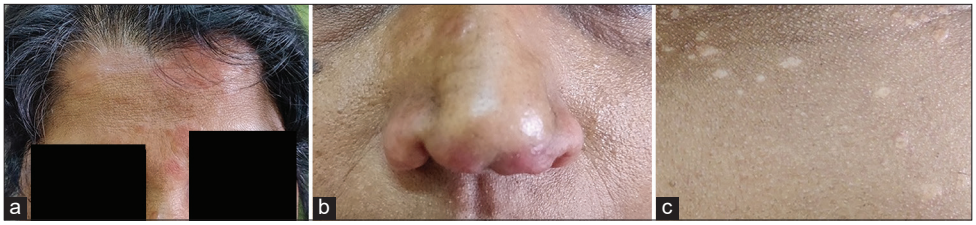

Figure 1: (a) Facial erythematous plaques with minimal central atrophy. (b) Deformed alae nasi. (c) Xanthelasmata like lesions over the upper back.

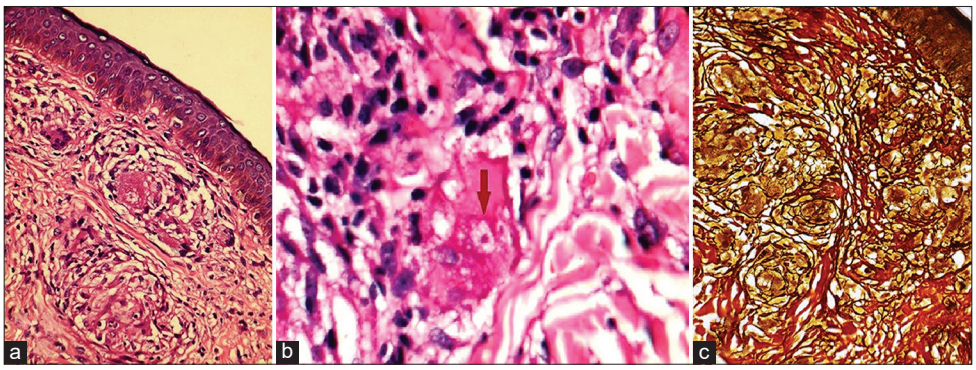

Figure 2: (a) Naked granulomas showing epitheloid cells and langhans giant cells (H & E, x100). (b) Asteroid body within Langhans giant cell (H & E, x400). (c) Reticulin staining showing accentuation of fibres around graulomas (Silver stain, x100).

lesions) and tapering doses of prednisolone (initiated at 40mg for pulmonary involvement). She showed significant improvement on subsequent follow-up.

Sarcoidosis commonly affects the lungs, mediastinal and peripheral lymph nodes, liver, eyes, bones, and skin. Hutchinson recorded the first case in 1865 and called it Mortimer's Malady after his famous patient Mrs. Mortimer. The first unequivocal case of Sarcoidosis in English literature was reported by Boeck in 1899 and the first case in India was reported by Rajam *et al*. in 1957.[1-3]

Histopathologically, it is characterized by naked granulomas (aggregates of epitheloid cells with a sparse lymphocytic component), a few Langhans giant cells and inclusion bodies namely Schaumann, asteroid, and residual bodies. IFN-γ, TNF-α, IL-2, and IL-12 play an important role in 'the formation and perpetuation of granuloma.[1]

Specific forms of cutaneous sarcoidosis display typical sarcoid/naked granulomas. The specific forms of cutaneous sarcoidosis include maculopapular, nodular, plaque, subcutaneous and scar sarcoidosis. Specific forms of sacrcoidosis also include lupus pernio, and certain less common forms (e.g, psoriasiform, lichenoid, hypopigmented, erythrodermic, verrucous, and morphea-like). Non-specific forms include erythema nodosum (most common) and rarely erythema multiforme, calcinosis cutis, and prurigo. Our patient showed plaque lesions over the face and xanthelasmata like maculopapular lesions over the upper back and arms.[1,4]

Pulmonary involvement occurs in 90% of the cases of sarcoidosis.[1] Hence the patient was advised chest radiography and CECT, though she did not manifest any symptoms and signs that suggested an internal organ involvement.

Treatment options include steroids (topical, intralesional, and systemic), antimalarials (chloroquine and hydroxychloroquine), tetracycline, methotrexate, and TNF-α antagonists. There are anecdotal reports of successful treatment with azathioprine, mycophenolate mofetil, leflunomide, and cyclophosphamide.[1,5]

Sarcoidosis, with its variable presentations, is a great imitator of other dermatological diseases. Our patient manifested coexistence of different morphological patterns of cutaneous sarcoidosis. To the best of our knowledge, this is the the second of its kind in literature, till date. This case also highlights the need for imaging techniques like x-ray and CECT while evaluating patients with sarcoidosis, even in the absence of clinically apparent systemic symptoms.

Thorough clinical examination and relevant investigations are important for the prompt diagnosis of sarcoidosis and for the timely detection of systemic involvement.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

Dr Bifi Joy, Dr Thyvalappil Anoop and Dr Rajiv Sridharan are on the editorial board of the Journal.

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