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LETTER TO THE EDITOR

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Striking appearance of Mortimer's malady

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To the Editor,

Cutaneous sarcoidosis is one of the great imitators in medicine, with clinical presentation varying from specific skin lesions of papules, plaques, nodules, lupus pernio, scar or tattoo infiltration among others to nonspecific lesions like erythema nodosum [1, 2]. One-third of patients with sarcoidosis or Mortimer's Malady may have cutaneous lesions and these may be the first clinical sign [3]. Sarcoidosis may get misdiagnosed, thereby delaying management and increasing risk of complications. We report the case of a patient being treated with histoid leprosy for more than a year and later diagnosed as sarcoidosis, treated with methotrexate and prednisolone with improvement in 4 weeks of therapy.

Case report

The patient is a 45-year-old male, farmer by occupation, and hailing from Madhya Pradesh, India; his condition started 2 years ago with insidious onset of multiple, red, raised cutaneous skin lesion and accompanied with constitutional features like fatigability, anorexia, intermittent low-grade fever, and symmetrical polyarthralgia in the small joints of hands and feet. The patient had a history of implantable cardioverter defibrillator placement for ventricular arrhythmia. He had initially been diagnosed by another physician with histoid leprosy based on the clinical presentation and high endemicity of leprosy in that area and received 18 blister packs of WHO-MDT (World Health Organization-multi-drug therapy), without any significant improvement.

General and systemic examination was unremarkable, with absence of organomegaly or lymphadenopathy. Cutaneous examination revealed multiple, dome-shaped, shiny, infiltrated erythematous papules and plaques, some with central delling, over face involving eyebrows, ear helices and alae nasi, and all over the chest and back (Fig. 1a, b).

Chest X-ray revealed enlarged right and left hilar and right paratracheal lymph nodes. Pulmonary function testing revealed restrictive airway disease. Viral markers for human immunodeficiency virus, hepatitis B/C, quantiFERON-TB Gold and slit skin smear for acid fast bacilli were negative.

Important investigations for diagnosis revealed non-caseating epithelioid granulomas with occasional multinucleated histiocytes in the dermis on fematoxylin and eosin staining from cutaneous biopsy. Reticulin stain was positive (Fig. 2a, b). Fite-Faraco staining and tissue CB-NAAT (cartridge-based nucleic acid amplification test) for mycobacteria were negative.

Based on the above findings, the patient was diagnosed as a case of sarcoidosis and put on methotrexate at the dose of 15 mg/week and prednisolone at 40 mg/day. After 4 weeks, the patient came back with almost complete resolution of skin lesions and improvement in general condition (Fig. 3a, b). Prednisolone was tapered slowly after 4 weeks and stopped after 3 months, while methotrexate was continued.

Discussion

Histoid leprosy and sarcoidosis are close clinical differentials in skin color. Both may present with skin-colored to erythematous papules, plaques, and nodules and feature non-caseating granulomas on

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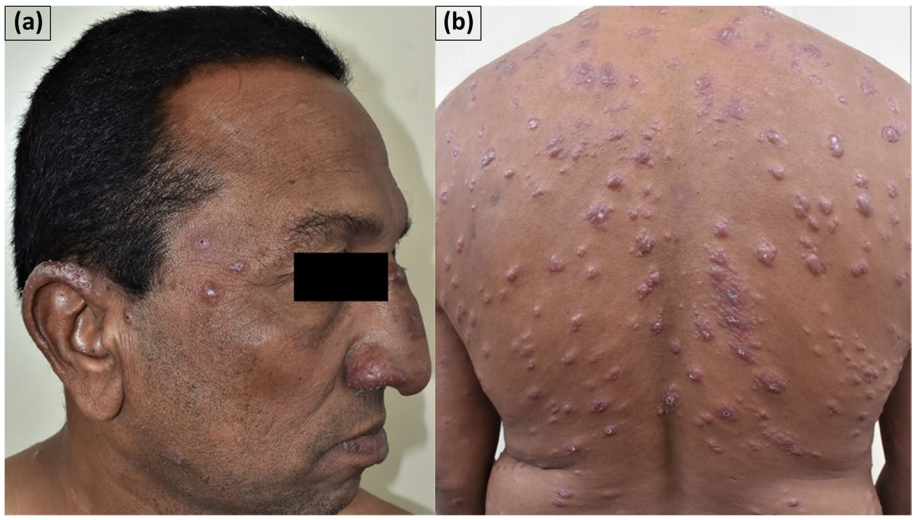

Fig. 1 **a** Multiple erythematous papules and plaques involving temporal region, earlobes, root of nose and alae nasi. **b** Shiny erythematous papules and plaques with central umbilication over few lesions on the trunk.

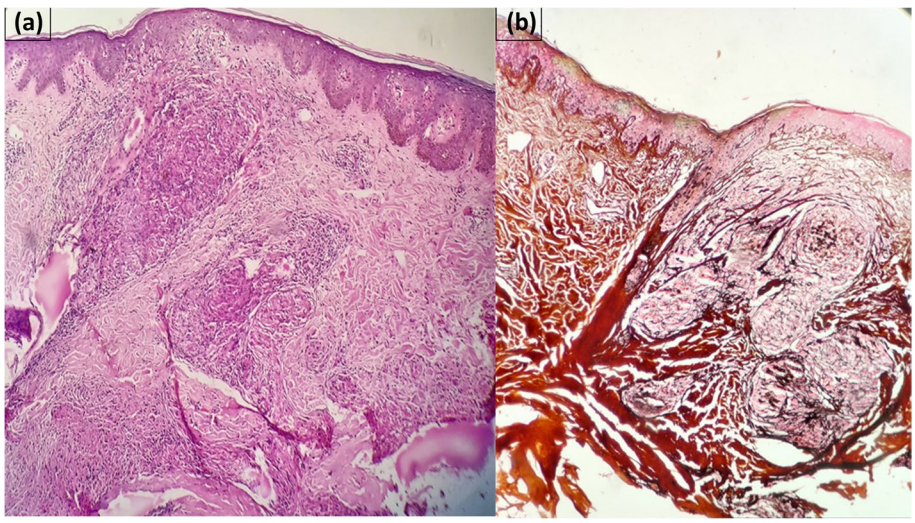

Fig. 2 **a** Hematoxylin-eosin staining, 10× magnification showing naked sarcoidal granulomas in dermis. **b** Reticulin staining, 10× magnification showing intact black fibers in granulomas denoting positivity

histopathology. The high prevalence of de novo cases of histoid leprosy in India may result in erroneous diagnoses. Histoid leprosy has an insidious onset with spontaneous appearance of skin lesions simulating cutaneous sarcoidosis, generally without any systemic involvement and there is presence of abundant lepra bacilli on slit skin smear and histopathology. In histoid leprosy, involvement of ear lobes is common while the alae of the nose are classically involved in sarcoidosis, as was seen in our case. Apart from cutaneous presentation, the presence of constitutional symptoms, cardiac involvement, hilar lymphadenopathy, and reticulin positivity of the granuloma favored the diagnosis of sarcoidosis over histoid leprosy in this case.

This case emphasizes the importance of looking beyond what strikes the eye and being open to reviewing and revising your diagnosis when faced with a clinical conundrum.

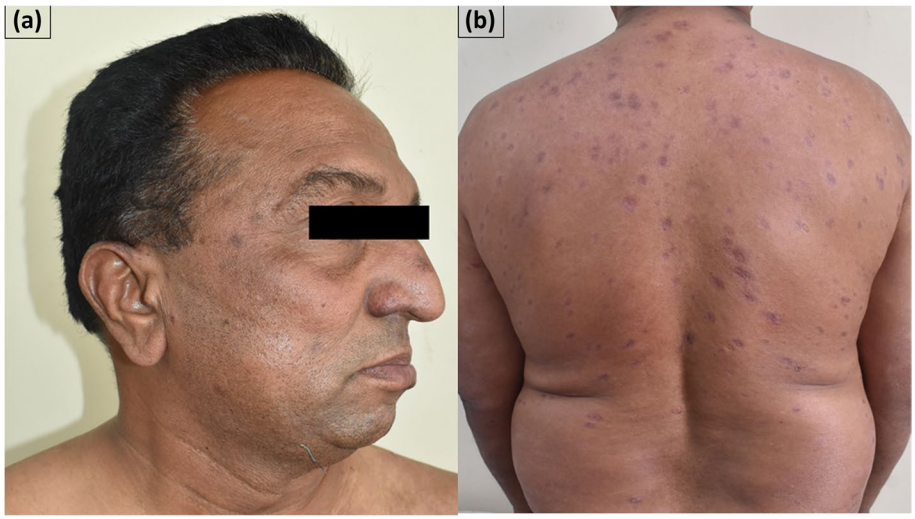

Fig. 3 **a**, **b** Complete resolution of cutaneous lesions with post-inflammatory hyperpigmentation after 4 weeks of prednisolone and methotrexate therapy

Abbreviations
WHO-MDT World Health Organization-multi-drug therapy
CB-NAAT Cartridge-based nucleic acid amplification test

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Authors' contributions
All authors contributed substantially to the conception and design of the manuscript, along with the acquisition, analysis, drafting and approval of data for the work. All authors agreed to be accountable for all aspects of the work and ensured that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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