De-novo Histoid Leprosy Masquerading as Lupus Miliaris Disseminatus Faciei: A Unique Presentation

Dear Editor,

A 23-year-old female presented to a tertiary care center in eastern India with complaints of sudden onset multiple facial lesions for 3 months, having been previously treated with acne vulgaris with oral isotretinoin without improvement. On cutaneous examination, multiple skin-colored, non-tender, discrete papules were noted over the centrofacial area of the face [Figure 1]. Clinical differential diagnoses considered were lupus miliaris disseminatus faciei (LMDF), papular sarcoidosis, trichoepithelioma, and post kala-azar dermal leishmaniasis (PKDL). Hematological and biochemical investigations including serum ACE levels were normal. Given the patient's apprehension regarding a facial biopsy, a therapeutic trial with doxycycline considering a clinical diagnosis of LMDF was initiated. On subsequent follow-up, the number of lesions increased with lesions appearing over bilateral ears raising suspicion of Hansens's disease [Figure 2]. There was no history of treatment taken for leprosy or monotherapy with dapsone for other indications. No history of leprosy or intake of multidrug therapy among family members and close contacts was elicited. Peripheral nerve examination was normal.

Slit skin smear showed a full field containing numerous acid-fast bacilli arranged singly (no globi), uniform in length, with longer than normal lepra bacilli with tapering ends. The average bacteriological index was 6+. A punch biopsy from a representative lesion revealed an atrophic epidermis with the presence of a grenz zone. Circumscribed sheets of elongated to epithelioid cells with histoid habitus and mild chronic inflammatory infiltrate were noted in the dermis [Figure 3a and b]. Wade fite stain for lepra bacilli was positive [Figure 3c and d]. The absence of caseating granuloma, naked granulomas, superficial nests of basaloid cells, and leishman-donovan bodies ruled out LMDF, sarcoidosis, trichoepithelioma, and PKDL, respectively.

Based on clinical history and examination, evidence from slit skin smear and histopathology, a final diagnosis of de novo histoid leprosy was made, and the patient was started on multidrug therapy of rifampicin, clofazimine, and dapsone for a duration of at least 2 years. Family members were

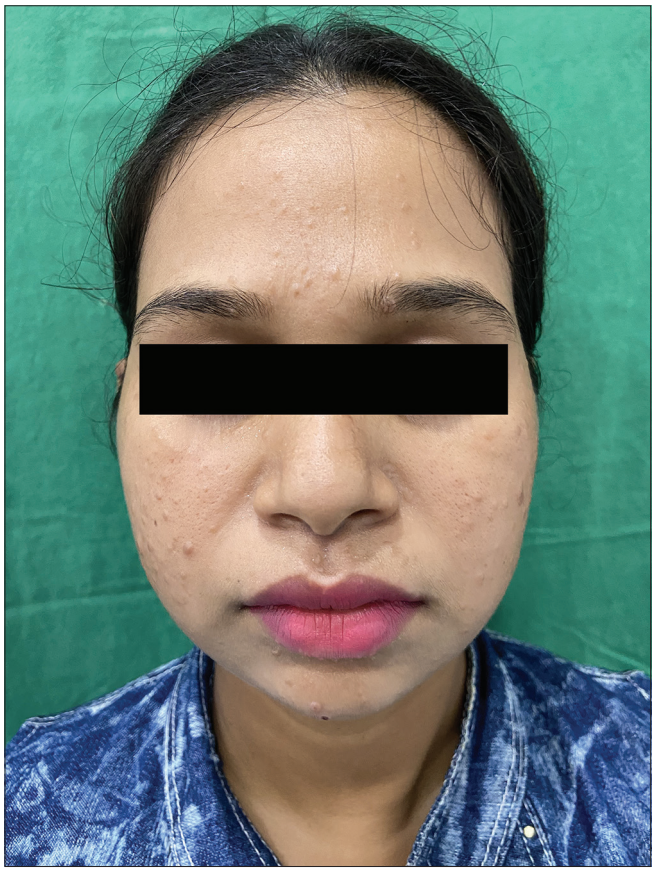

Figure 1: At presentation, multiple asymptomatic skin-colored papules over entire face

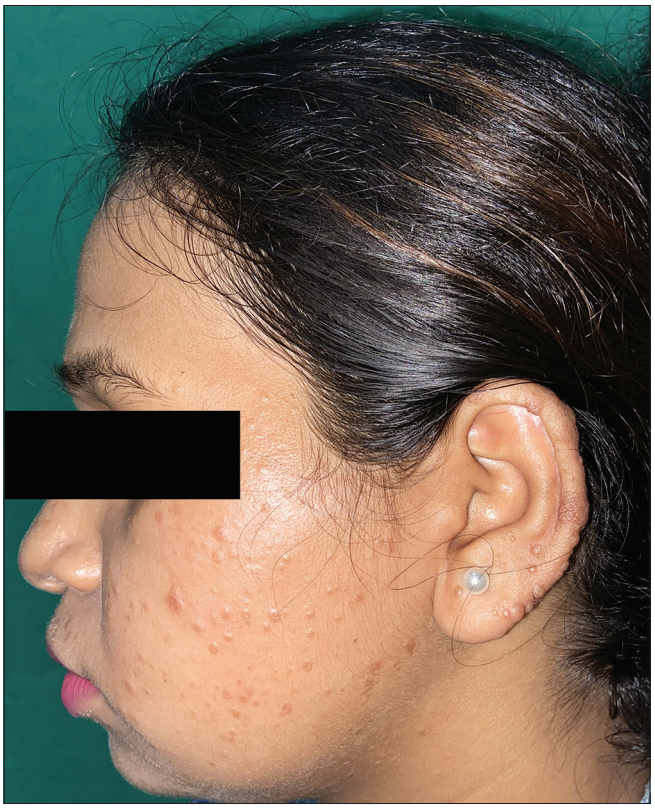

Figure 2: On subsequent follow-up, new lesions appeared over bilateral ear lobes

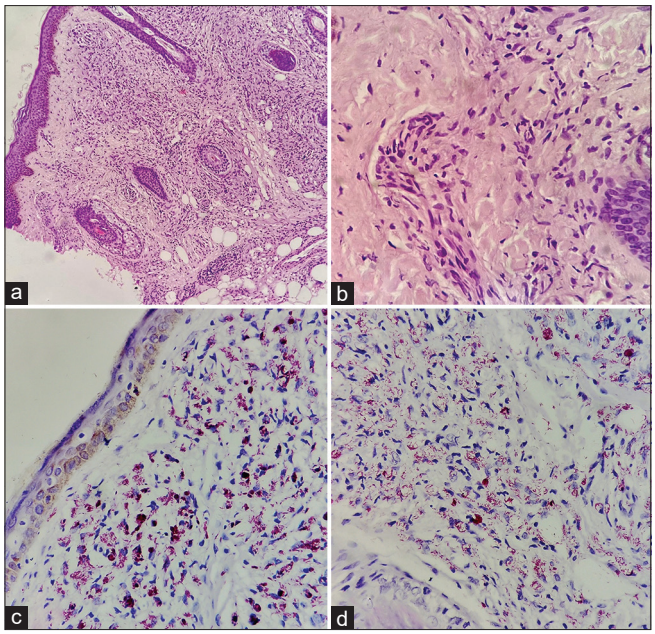

Figure 3: (a) Photomicrograph shows atrophic epidermis, presence of grenz zone and inflammatory pathology in the dermis [H and E stain, 10 ×]. (b) Sheets of elongated to epitheloid cells along with mild chronic inflammatory infiltrate present in the dermis [H and E stain, 40 ×]. (c) Wade fite stain along epidermis shows atrophic epidermis, grenz zone along with numerous histoid bacilli (100x). (d) Wade fite stain shows numerous histoid bacilli in the dermis (100x)

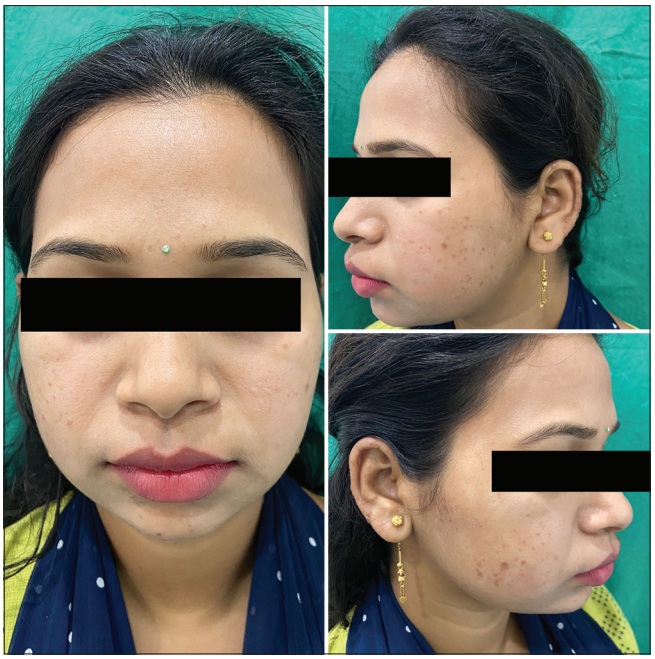

Figure 4: At follow-up visit after 6 months, the size of older lesions had decreased with no appearance of new lesions

screened clinically and showed no evidence of Hansen's disease. Follow-up at 6th month revealed a decrease in the size of older lesions with no new lesions [Figure 4].

The term "histoid" comes from the histopathological appearance of nodules containing spindle-shaped cells resembling a dermatofibroma. It is regarded as an uncommon variant of lepromatous leprosy, however, some authors consider it to be a distinct entity. Indian data reports incidence in the range of 2.79%–3.60% with an average age of presentation being 21–40 years with a male predominance.[1] The history of dapsone monotherapy prevalent before multidrug therapy is the most common implicated factor.[2] Denovo histoid leprosy represents a rarer subtype of histoid leprosy wherein no cause can be ascertained with incidence rates being 12.5% of all histoid leprosy cases.[3] Clinically, it presents as multiple nontender, firm, discrete, smooth, dome-shaped, skin-colored papules and nodules over the face, abdomen, back, buttocks, and extremities. Erythema nodosum-like lesions and figurate lesions have also been described.[4,5] Diagnosis is made via classical histopathological findings and Wade fite staining for lepra bacilli. Immunohistochemistry using factor XIIIa and S100 helps differentiate it from dermatofibroma and neurofibroma, respectively.[6]

LMDF is a rare inflammatory dermatoses of unknown etiology characterized by centrofacial asymptomatic papular eruptions typically around the eyelids that heals with scarring. Attributable histopathological findings are dermal epitheloid granuloma with caseous necrosis.[7] Our patient presented with a similar distribution prompting us to consider LMDF first and give a therapeutic trial with doxycycline. However, failure of response to therapy and subsequent appearance of lesions over earlobes prompted consideration for leprosy, which was later confirmed on slit-skin smear and biopsy.

A differential diagnosis of histoid leprosy should be considered in such scenarios, especially in countries like India where leprosy still represents a public health challenge. In conclusion, LMDF-like presentation is rare and must be added to various cutaneous morphological presentations of histoid leprosy.

Declaration of patient

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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