

# Bilateral malar milia en plaque as primary presentation of discoid lupus erythematosus



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**Key words:** discoid lupus erythematosus; milia en plaque.

## INTRODUCTION

Milia are small benign superficial keratinous cysts, measuring 1 to 4 mm in diameter. Milia can be primary, appearing spontaneously, or secondary to trauma, skin disease, or medication.<sup>1</sup> Milia en plaque (MEP) is a rare form of this condition characterized by numerous aggregated milia on an erythematous plaque.<sup>2</sup> Discoid lupus erythematosus (DLE) represents the most common subtype of cutaneous lupus erythematosus and various presentations have been described. We report an unusual case of bilateral malar MEP as a primary presentation of DLE.

## CASE REPORT

A 31-year-old woman presented with a 4-month history of a facial rash involving both cheeks and the bridge of the nose. Her medical history was unremarkable with no personal or family history of skin disease. She reported no constitutional symptoms and denied any recent trauma or cosmetic procedures.

The patient consulted her primary care physician about 2 months after the lesions first appeared and was prescribed a mild topical steroid (hydrocortisone 1%) and given an aqueous cream but without improvement.

On physical examination, the patient had multiple yellowish cysts on an erythematous plaque and scattered open comedones in a malar distribution (Fig 1). The remainder of her body was spared, and she had no oral ulcers or alopecia.

Dermoscopy showed multiple milia cysts, follicular keratotic plugs, perifollicular whitish halos, pigmentation, and telangiectasia (Fig 2). Routine

### Abbreviations used:

DLE: discoid lupus erythematosus  
MEP: milia en plaque

laboratory investigations found a normal full blood count and negative HIV and rapid plasma regain test results. Antinuclear antibody, anti-double-stranded DNA antibody, and anti-Sm antibody screens were all normal.

Histologic examination from biopsies taken from the malar lesions found numerous keratin-containing milia within the upper dermis. The epidermis showed dilated hair follicle infundibulae with keratin plugs and focal epidermal atrophy. The papillary and reticular dermis showed a perivascular and perifollicular infiltrate comprising mainly lymphocytes (Fig 3, A). A lichenoid infiltrate of lymphocytes surrounding a fragment of follicular epithelium with vacuolar degeneration was also seen (Fig 3, B). Focal fibrosis and granulomatous inflammation was attributed to keratin from ruptured milia. An alcian blue periodic acid-Schiff stain showed mild increased dermal mucin (Fig 3, C).

A diagnosis of MEP in association with DLE was made, and the patient was started on a topical retinoid (tretinoin 0.1% cream) and topical steroid (methylprednisolone 0.1% ointment). She was counselled about sun avoidance and appropriate sunscreen use.

After 6 weeks of treatment, a partial response was noticed with a decrease in erythema and milia cysts (Fig 4).

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**Fig 1.** Multiple yellowish cysts on an erythematous plaque and scattered open comedones in a malar distribution.

## DISCUSSION

MEP is classified as primary milia deriving from the infundibulum of the vellus hair follicle and usually develops spontaneously in healthy skin with no clear etiology.<sup>1</sup>

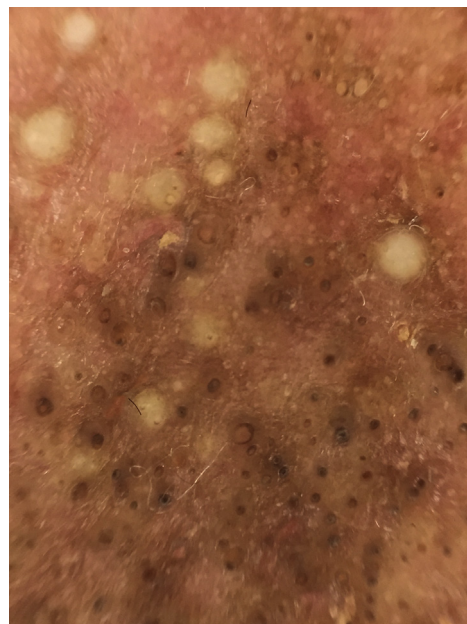
First described by Balzer and Fouquet<sup>3</sup> in 1903 and named by Hubler et al<sup>4</sup> in 1978, MEP occurs mainly in middle-aged women and has a preference for the head and neck area, especially periauricularly. It has also been described as periorbital, on the nasal bridge, and truncal.<sup>1</sup>

MEP has been reported in association with DLE.<sup>5-8</sup> It can also be associated with pseudoxanthoma elasticum, lichen planus, trauma, drugs such as cyclosporine, or renal transplantation.<sup>1</sup>

The patient reported no pre-existing skin lesions and also denied any precipitating factors that could lead to secondary milia like blistering skin conditions, dermabrasion, contact dermatitis, trauma and radiotherapy.<sup>1</sup>

The patient was using a topical steroid, but that was not considered a trigger for secondary milia, as she was only using it for about a month and only after the lesions developed.

Her clinical presentation was suggestive of DLE based on the morphology of erythematous plaques with open comedones in a malar distribution. The clinical suspicion was reinforced by the dermoscopy features present on examination. Perifollicular



**Fig 2.** Dermoscopy found multiple milia cysts, follicular keratotic plugs, perifollicular whitish halos, pigmentation, and telangiectasia.

whitish halos, follicular keratotic plugs, and telangiectasia are the most common dermoscopic criteria for DLE.<sup>9</sup>

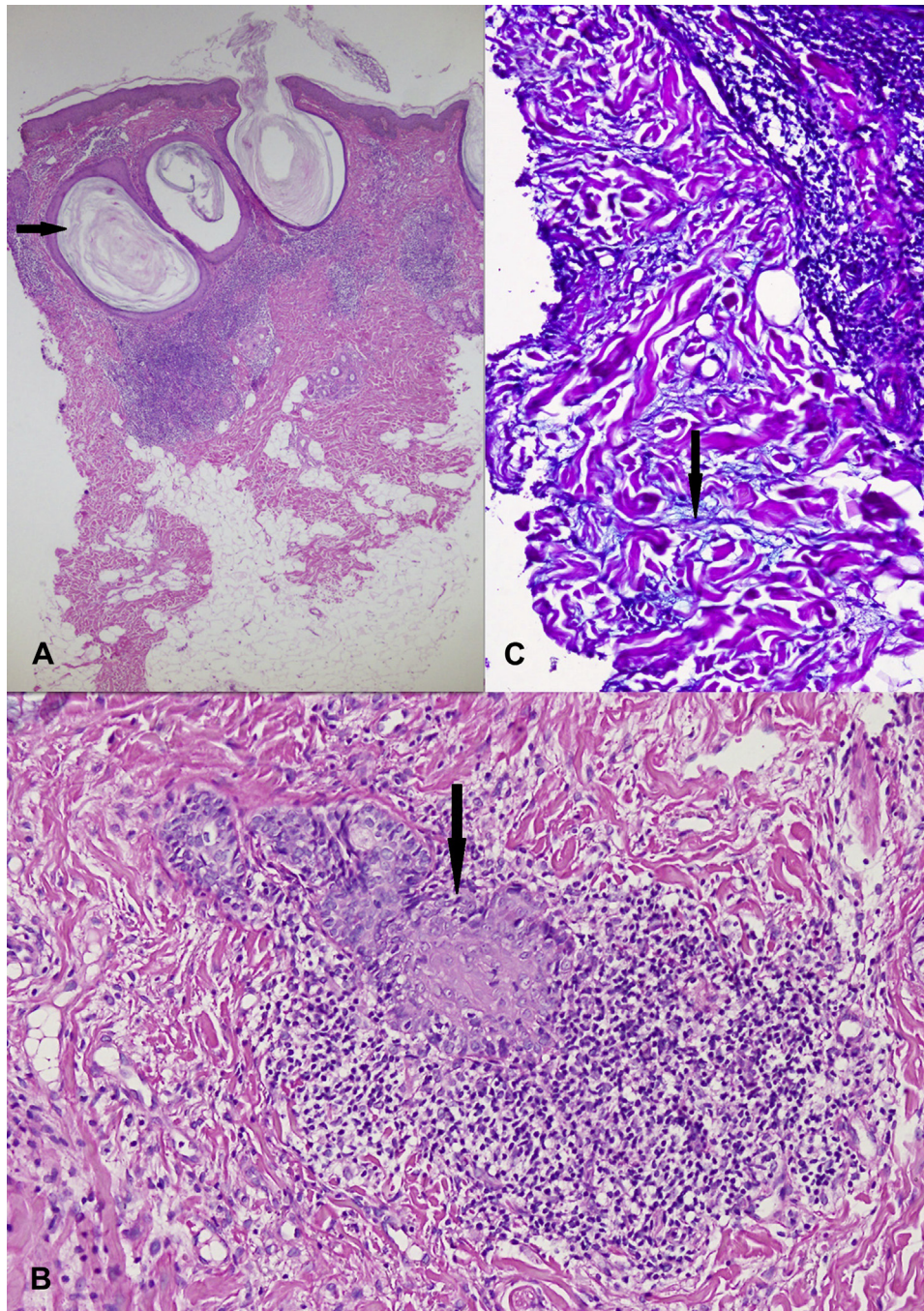
The multiple milia cysts, however, are not typical for DLE and we considered secondary causes for milia formation as part of our differential diagnosis.

Histologically, MEP is characterized by keratin-filled epidermal cysts and a mononuclear cell infiltrate.<sup>10</sup> Our patient's histology was consistent with that of multiple milia cysts and features of DLE, namely, epidermal atrophy, follicular plugging, a papillary and reticular perivascular and periadnexal lymphocytic infiltrate, vacuolar degeneration, and increased mucin in the dermis.

Two case reports in the English-language literature describe MEP as the primary presenting lesions of lupus erythematosus.<sup>7,8</sup> We believe this case to be unusual, demonstrating MEP as a primary presentation of DLE in a bilateral malar distribution.

The pathogenesis of MEP and DLE are not well delineated, but Boehm et al<sup>6</sup> attributed the formation of milia to damage of the adnexal structures in the setting of existing DLE lesions.

In a case in which MEP developed de novo as manifestation of DLE, Kouba et al<sup>7</sup> attributed milium formation to alteration of the infundibular portion of the follicle, resulting in dysfunctional keratinisation and outlet obstruction. Our case supports this hypothesis.



**Fig 3.** **A**, Histology showed a superficial and deep perivascular and periadnexal infiltrate consisting of lymphocytes. Numerous keratin-containing milia within the upper dermis (*arrow*). The epidermis showed dilated hair follicle infundibulae with keratin plugs and focal epidermal atrophy. **B**, A lichenoid infiltrate of lymphocytes surrounding a fragment of follicular epithelium with vacuolar degeneration (*arrow*). **C**, Mild increased dermal mucin (*arrow*). (**A** and **B**, Hematoxylin-eosin stain; **C**, alcian blue periodic acid-Schiff stain; original magnifications: **A**,  $\times 4$ ; **B** and **C**,  $\times 10$ .)

The patient showed a partial response to a moderate potency topical steroid (methylprednisolone 0.1% ointment) and a topical retinoid (tretinoin 0.1% cream), with decreased erythema and milia,

supporting its use for MEP in DLE. The patient will be monitored for progression of her chronic cutaneous lupus erythematosus and for development of systemic lupus erythematosus. She will be seen at



**Fig 4.** After 6 weeks of treatment with topical tretinoin 0.1% cream and topical methylprednisolone 0.1% ointment. A decrease in the number of milia cysts and erythema can be seen.

4-month intervals, and we will consider oral chloroquine in the future.

Our case supports the hypothesis by Kouba et al<sup>7</sup> that MEP is a specific manifestation of DLE and that the diagnosis of lupus erythematosus be considered if lesions arise de novo on sun-exposed skin.<sup>7</sup>

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