

# Sudden Onset of Photodistributed Milia - an Unusual Presentation of Cutaneous Connective Tissue Disease?

Etta Hanlon MS<sup>1</sup>, Casey E. Engel MD<sup>2</sup>, Kiley F. Fitzgerald MD<sup>2,3</sup>, Zachary E. Holcomb MD<sup>2</sup>

<sup>1</sup>Virginia Tech Carilion School of Medicine (VTC SOM) <sup>2</sup>Division of Dermatology, Department of Internal Medicine, Carilion Clinic, <sup>3</sup>Dominion Pathology Associates

## Background

- Discoid lupus erythematosus (DLE) is a subtype of cutaneous lupus erythematosus.
- A rare presentation of DLE includes facial milia cysts and inflammatory plaques<sup>1</sup>.
- Milia en plaque (MEP) is characterized by aggregated milia on an erythematous base, commonly appearing on the face, neck, and retroauricular area<sup>2,3</sup>.
- A previous study suggested that milia may form due to damage to adnexal structures in the setting of existing DLE<sup>2</sup>.

## Case Presentation

- We describe a 69-year-old female who presented with a 5-month history of facial rash. She reports developing widespread milia cysts in a malar distribution across the cheeks and forehead, which then spread to her neck and postauricular area.
- Physical exam was notable for hundreds of small uniform white milium on the face and neck. (Figure 1A, 1B).
- The patient had no other cutaneous or systemic evidence of systemic lupus or other autoimmune disease.
- Pathology revealed a dilated superficial epidermoid cyst with surrounding lymphocytic inflammation and a vacuolar interface dermatitis around the cyst, as well as increased dermal mucin (Figures 2, 3, 4 & 5).
- Immunofluorescence showed fibrinogen deposition at the dermoepidermal junction. PAS staining was negative for fungal forms.

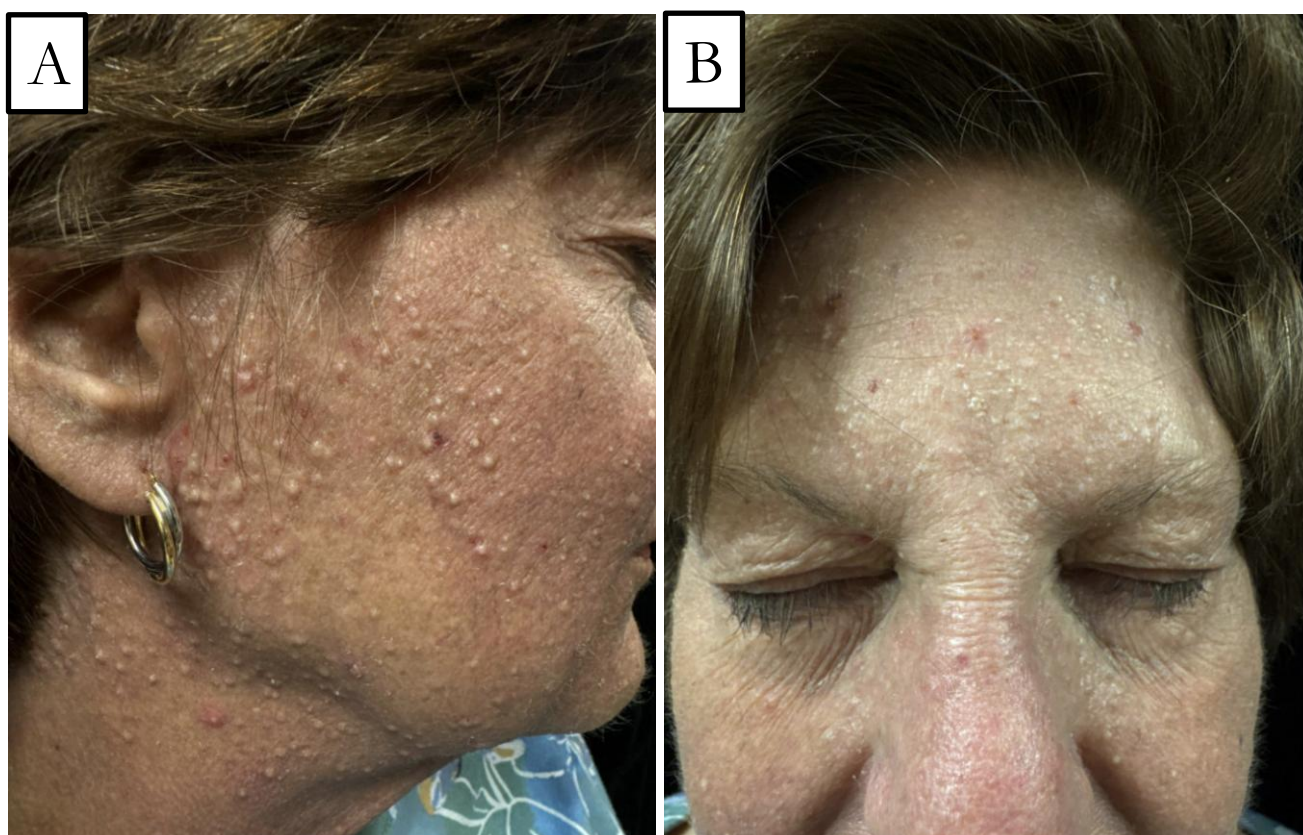


Fig. 1A, 1B

Clinical photograph of widespread milia cysts in a malar distribution across the cheeks, forehead and postauricular area.

## Histologic Findings

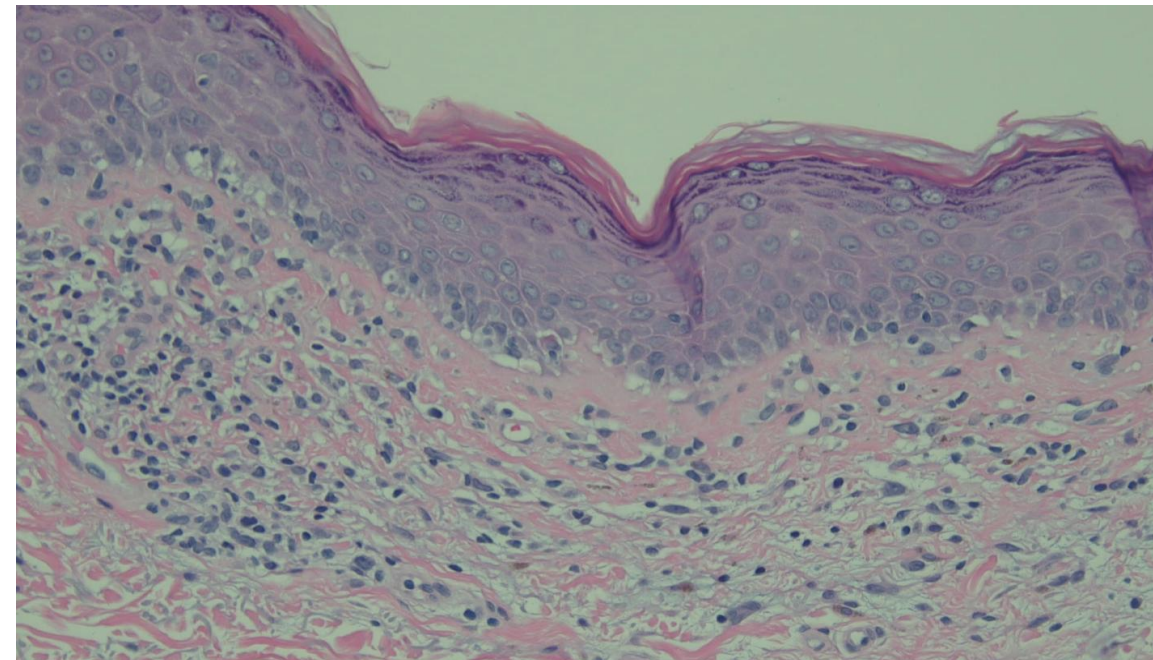


Fig. 2 H&E x40 shows vacuolar interface dermatitis along the dermoepidermal junction

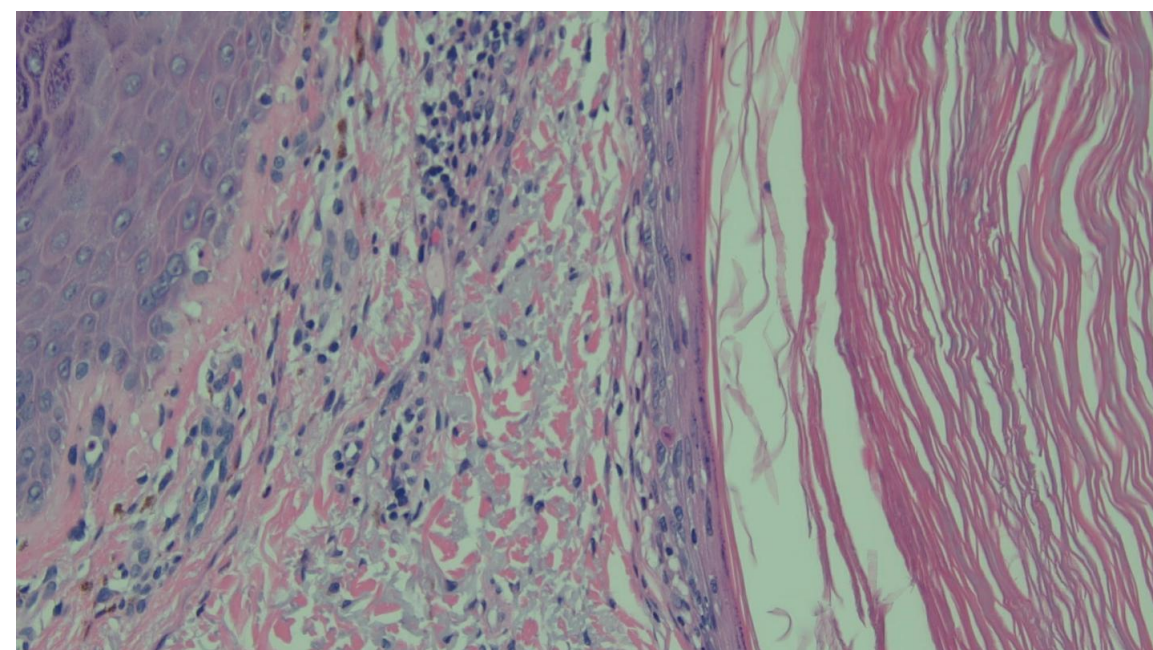


Fig. 3 Higher power photo of vacuolar interface dermatitis at the dermoepidermal junction and along milia with dyskeratotic keratinocytes (H&E 200X)

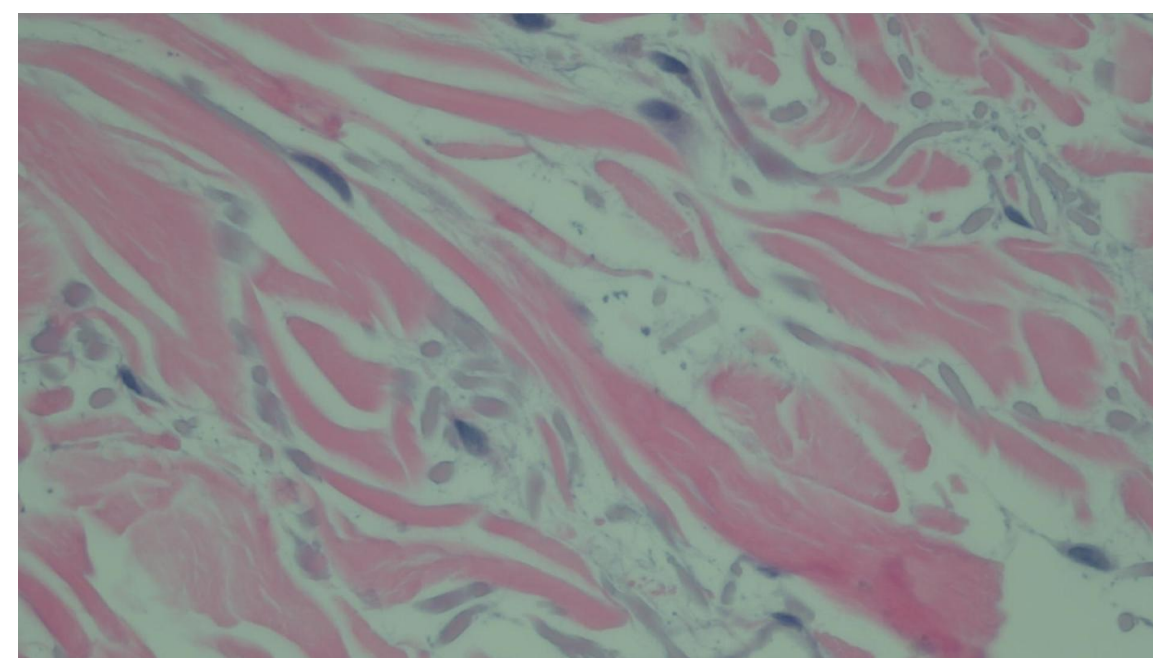


Fig. 4 H&E x600 shows increased dermal mucin

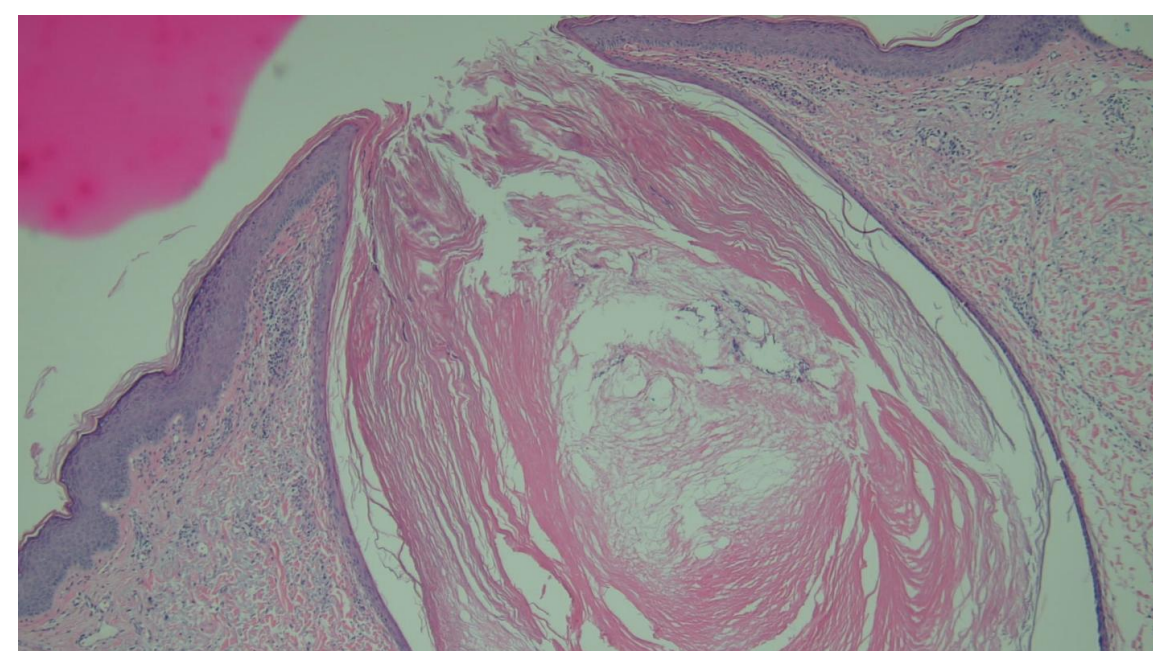


Fig. 5 H&E x40 shows milia and interface changes

## Clinical Course

- A 69-year old female presented with a 5-month history of discoid lupus on the face. She reported developing widespread milia cysts in a malar distribution across the cheeks and forehead, which then spread to her neck and behind her ears.
- Patchy inflammatory plaques of dilated cystic follicles and follicular cysts with surrounding inflammation were present, mostly chronic in nature.
- Biopsies revealed dilated keratin follicular cysts with surrounding lymphocytic inflammation. Immunofluorescence showed fibrinogen positive deposition at the dermoepidermal junction (Figures 2 and 3).
- The remainder of her body showed no ulcers, alopecia, or other signs of systemic lupus.
- Patient was advised to continue hydroxychloroquine 200mg BID and use of topical tretinoin 1%.

## Discussion

- Discoid lupus erythematosus (DLE) is a subtype of cutaneous lupus erythematosus which has a variety of presentations. Rare presentations of DLE have been described, including DLE with widespread facial milia cysts and inflammatory plaques, mimicking acneiform eruptions<sup>1</sup>.
- A similar entity of milia en plaque (MEP) is characterized by aggregated milia on an erythematous base, commonly appearing on the face, neck, and retroauricular area<sup>2,3</sup>.
- The underlying mechanism of MEP and the milia-variant of DLE remain unclear, however Boehm et al. suggested that milia may form due to damage to adnexal structures in the setting of existing DLE<sup>2</sup>.
- Only one similar presentation of DLE has been reported previously, this case broadens our knowledge of a rare manifestation of this cutaneous connective tissue disease.

## References

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