

# Paraneoplastic Dermatomyositis Presenting With Interesting Cutaneous Findings

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## PRACTICE POINTS

- Dermatomyositis has myriad cutaneous features including the shawl sign, the heliotrope sign, and Gottron papules.
- Less commonly, patients can present with the Holster sign (poikiloderma of the lateral thighs).
- Even less commonly, as in this report, patients can present with a psoriasiform papular eruption on the knees or with flagellate erythema on the back.



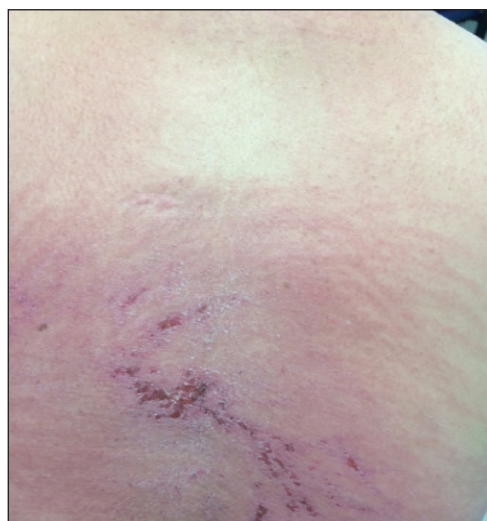
**FIGURE 1.** Confluent erythematous papules noted predominantly on the left knee.

To the Editor:

We report an interesting clinical case of dermatomyositis (DM) that presented with an associated malignancy (small cell lung cancer). This patient also had an unusual clinical finding of predominantly unilateral, confluent, erythematous papules on the knee, a cutaneous sign that is seldom described in the DM literature. This case serves to reinforce the classic findings and associations of DM, in addition to the uncommon manifestation of predominantly unilateral papules on the knee.

A 68-year-old woman presented with several cutaneous manifestations including the classic findings of photodistributed erythema on the arms and face, a heliotrope rash, Gottron papules, and confluent pink papules on the left knee (Figure 1). The patient also had one of the more rare manifestations of DM, flagellate erythema on the back (Figure 2). She had a history of breast cancer and was found to have metastatic small cell lung cancer at the time of the DM diagnosis.

A punch biopsy from an area of flagellate erythema on the back revealed an interface dermatitis with a superficial, perivascular, lymphocyte-predominant



**FIGURE 2.** Flagellate erythema on the back.

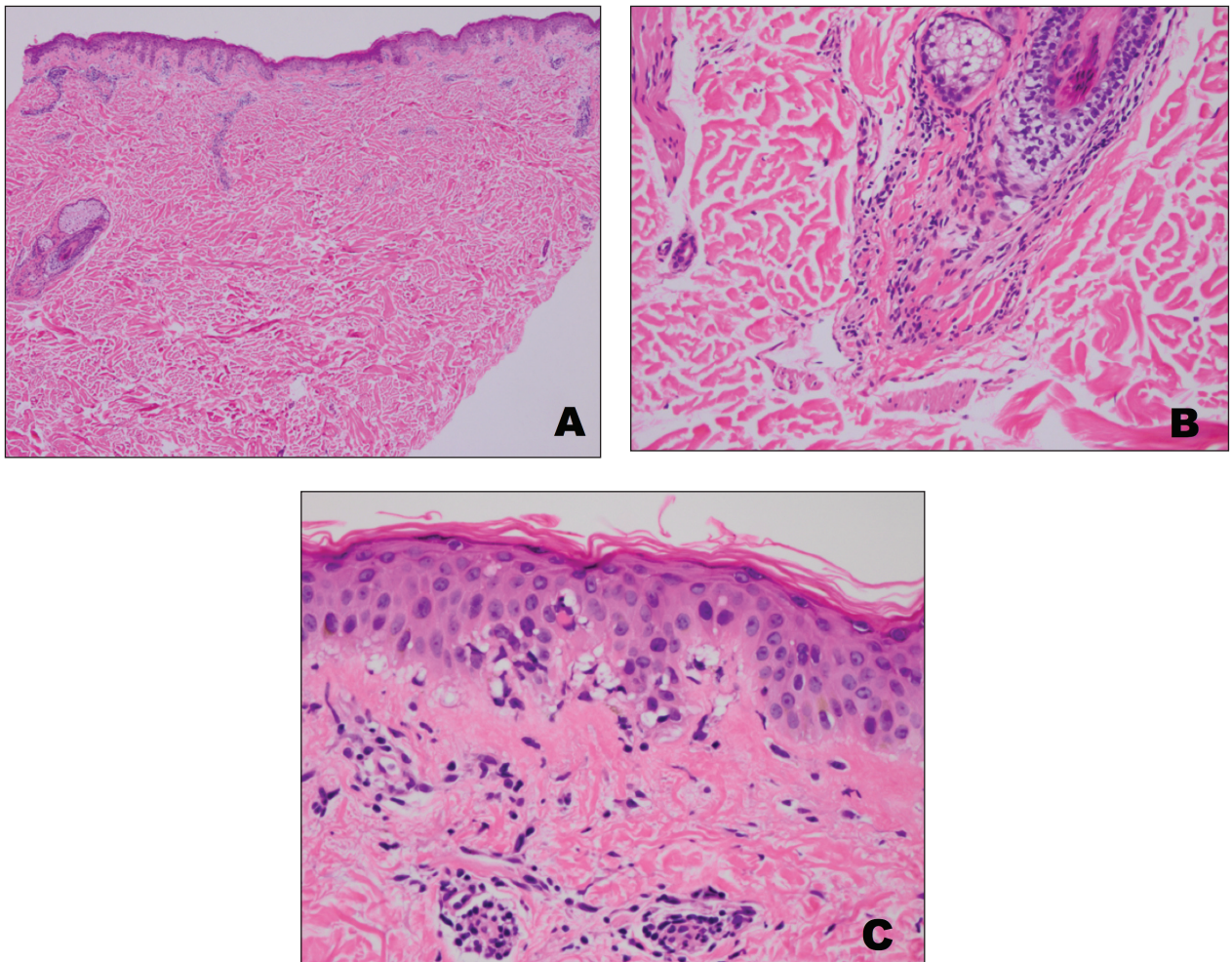
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inflammatory infiltrate (Figure 3). Alcian blue and colloidal iron stains revealed a marked increase in papillary dermal mucin. With the characteristic changes on skin biopsy and the classic skin findings present in our patient, we felt confident diagnosing her with DM. At the time of diagnosis, the patient also was found to have metastatic small cell lung cancer, suggesting a true paraneoplastic relationship.

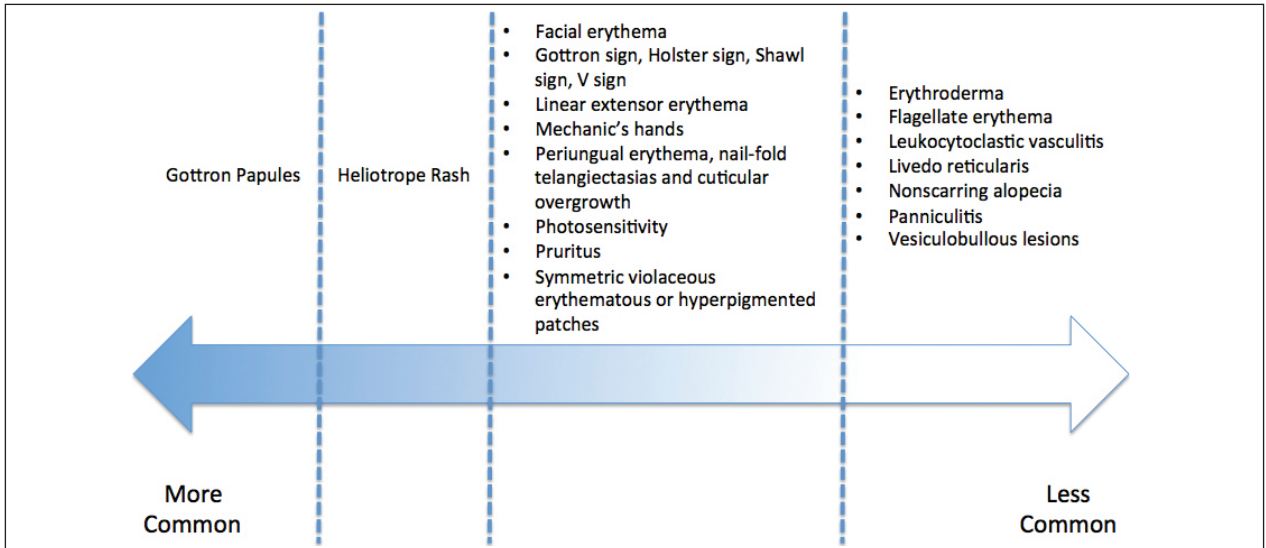
In 1975, Bohan and Peter<sup>1</sup> noted that the presence of cutaneous involvement was considered necessary for definite identification of DM. Since then, numerous dermatologic features have been identified (Figure 4),<sup>2</sup> including the characteristic findings of a heliotrope rash and Gottron papules.<sup>3</sup> The DM literature describes knee involvement consisting of confluent pink papules<sup>1,2,4</sup> similar to the rash in our patient. However, a symmetric pattern of involvement typically has been described, either the macular erythematous patches of Gottron sign<sup>2</sup> or the

follicular eruption and pustules of the knees described by Lister et al.<sup>4</sup> Our case represents a unique dermatologic manifestation of DM that requires further research to identify its incidence given its unilaterality.

The association of DM and amyopathic DM with internal malignancy is well known. Bohan and Peter<sup>1</sup> noted an overall figure ranging from 15% to 34% with an increased frequency in patients with skin and muscle involvement.<sup>1</sup> Hill et al<sup>5</sup> examined this link in a population-based study that identified corresponding malignancies. Specifically, they noted cancers to arise most frequently in the airway (eg, lung, trachea, bronchus), ovaries, breasts, colorectal region, and stomach.<sup>5</sup> There also has been work performed to identify if certain dermatologic findings may be associated with a higher risk of malignancy.<sup>6,7</sup> A meta-analysis by Wang et al<sup>6</sup> showed that Gottron sign did not have an association with cancer, but findings of cutaneous necrosis did have an association. It is unknown



**FIGURE 3.** Histopathologic findings. A, Low-power view demonstrated an interface dermatitis with a mild superficial and mid-dermal perivascular and periadnexal inflammatory infiltrate (H&E, original magnification ×40). B, Higher-power view highlighted the periadnexal inflammation (H&E, original magnification ×200). C, An interface dermatitis with scattered necrotic keratinocytes was evident on higher magnification (H&E, original magnification ×200).



**FIGURE 4.** Select dermatologic findings in dermatomyositis.<sup>2</sup>

if the specific cutaneous findings in our patient, including the predominantly unilateral papules on the knee, may have been a clue to the underlying malignancy.

In summary, we believe that our patient presented with the classic manifestations of DM in addition to the curious cutaneous sign of predominantly unilateral, confluent, erythematous papules on the knee, a clinical finding that may aid in the diagnosis of DM and also may alert the clinician to a possible underlying malignancy.

#### REFERENCES

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