

# Dermatomyositis associated with an adrenal adenoma



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**Key words:** adrenal adenoma; adrenal tumor; dermatomyositis.

## INTRODUCTION

Dermatomyositis (DM) is an autoimmune disease characterized by skin inflammation and muscle weakness, but skin inflammation may occur without clinically detectable muscle involvement in the amyotrophic variant of the disease (also called dermatomyositis sine myositis). There is a well-established link between DM and internal malignancies such as cancers of the lungs, ovaries, pancreas, and breast.<sup>1</sup> We describe a rare case of DM associated with an adrenal tumor.

## CASE REPORT

A 71-year-old Puerto Rican woman presented with a 2-year history of a waxing and waning pruritic, burning rash that started on her chest and subsequently spread to her back, arms, and face. She did not experience muscle pain or weakness. Her past medical history included hypertension, gastroesophageal reflux disease, and asthma. She was taking omeprazole, verapamil, fexofenadine, and occasional ibuprofen. She had no known family history of malignancies or autoimmune conditions.

Her skin examination demonstrated violaceous erythema of the forehead and upper eyelids. She had diffuse pink-red patches and plaques on the dorsal surfaces of both her arms and hands, including over the metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints. There was a 14 × 9-cm pink-brown patch on her upper chest (Fig 1, A). She also had several pink-red patches with white scale on her upper back (Fig 1, B).

Laboratory findings, including creatine kinase, anti-nuclear antibody, anti-Smith antibody, aldolase, anti-Ro (SS-A) antibody, anti-La(SS-B) antibody, and anti-Scl70 antibody, were within normal limits. A

### Abbreviations used:

DM: dermatomyositis  
IIM: idiopathic inflammatory myopathy

skin biopsy of the dorsal surface of the right hand showed vacuolar alteration of the dermal-epidermal junction, scattered apoptotic keratinocytes, and a superficial and mid-dermal lymphocytic infiltrate consistent with dermatomyositis (Fig 2). Given her skin findings, the absence of musculoskeletal symptoms, and biopsy result, a diagnosis of amyotrophic dermatomyositis was made. Because the agreement between the clinical examination and the histopathologic findings established a high confidence in the diagnosis, further testing for myositis-specific antibodies was not done.

The patient started hydroxychloroquine 200 mg twice daily for the next year but continued to have periods of worsening erythema and pruritus. Trials of prednisone 20 mg daily and methotrexate 20 mg weekly were added to her regimen but did not produce any improvement in her symptoms. As part of screening for occult malignancy, she underwent a computed tomography scan of her chest, abdomen, and pelvis, which revealed a 4.0 × 4.1-cm left adrenal mass (Fig 3).

The remainder of her screening tests, including a Papanicolaou smear, mammogram, and transvaginal ultrasound, were unremarkable. A left adrenalectomy was performed 14 months after initial presentation. The cut surface of the adrenal gland revealed a tan-white, firm, circumscribed adrenal nodule measuring 2.8 × 2 cm (Fig 4, A). Histologic examination showed a necrotic, hypercellular nodule with

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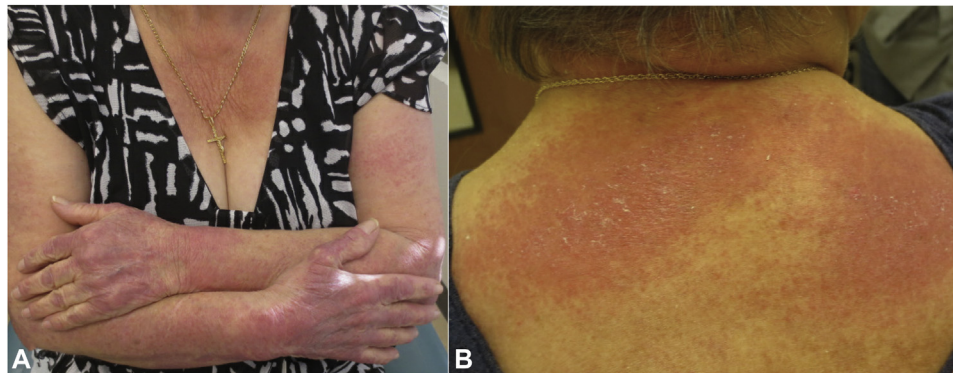
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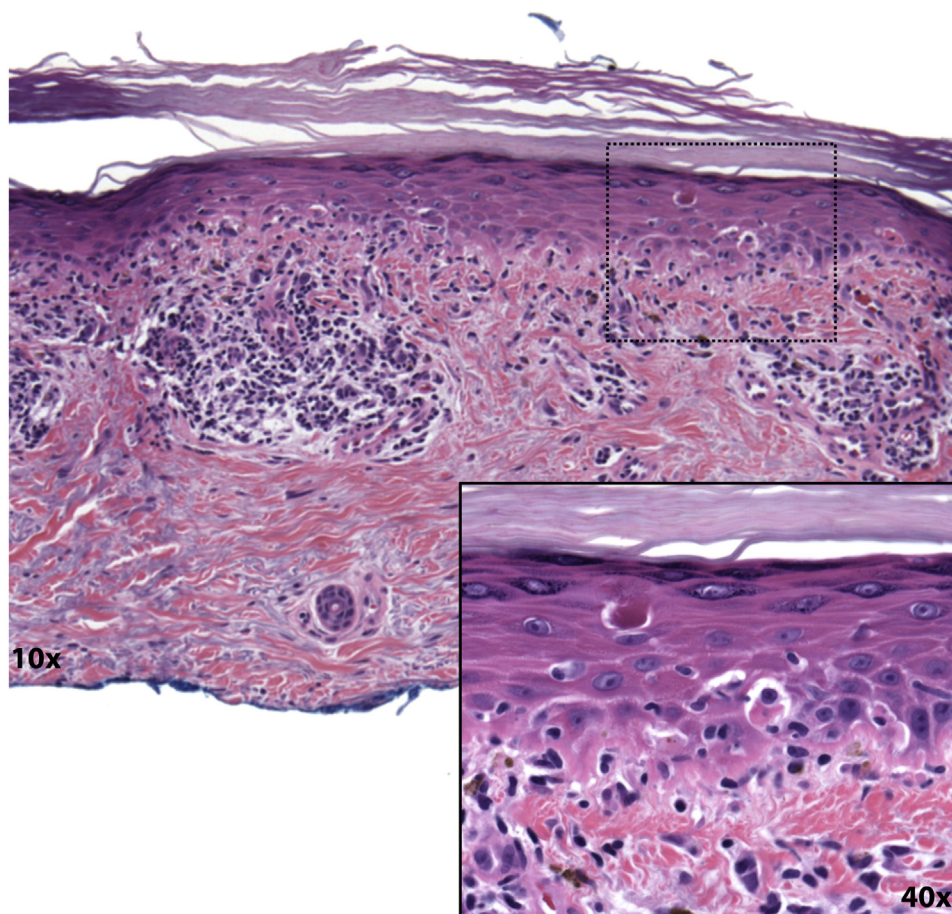
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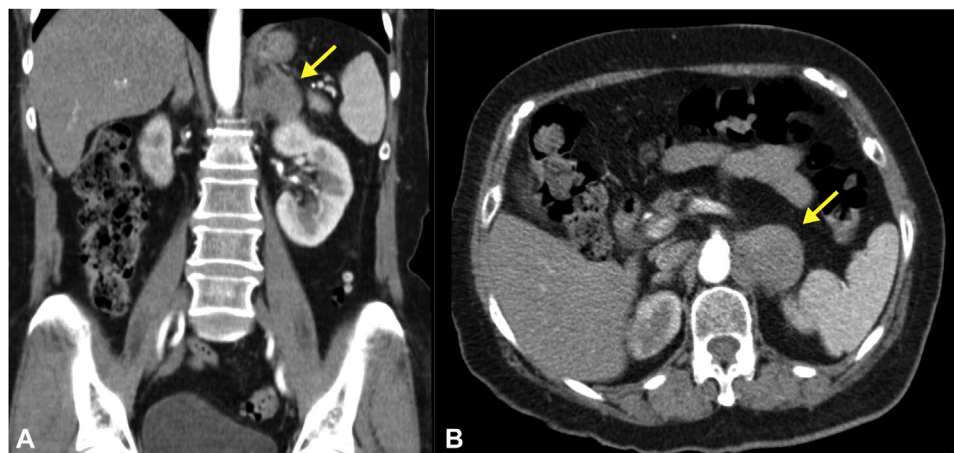
**Fig 1.** Clinical examination findings. **A**, Large patch on the upper chest and diffuse patches and plaques on the dorsal surfaces of the arms and hands. **B**, Patches with white scale on the upper back.



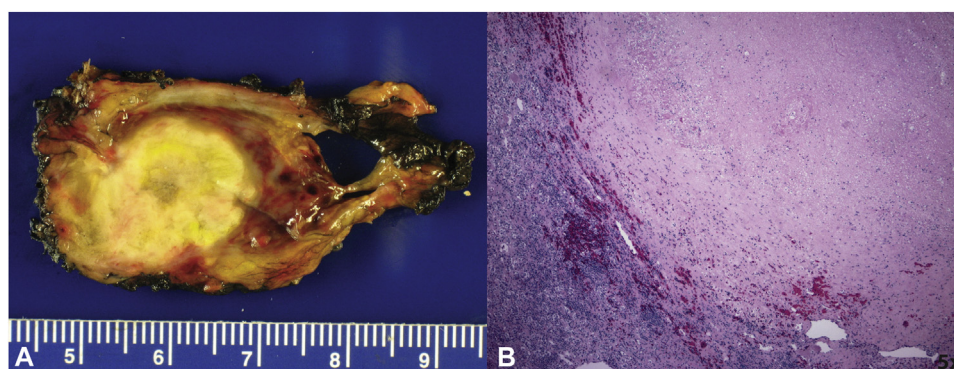
**Fig 2.** Skin histopathologic findings. Biopsy of the dorsal surface of the right hand showing interface dermatitis consistent with dermatomyositis. (Hematoxylin-eosin stain.)

surrounding chronically inflamed, atrophic adrenal cortex (Fig 4, B). Keratin immunomarkers and the adrenal parenchymal markers synaptophysin, inhibin, and calretinin were not expressed in the nodule; however, CD45 and the B cell marker CD20 decorated ghosted, necrotic cells. An infarct adrenal

cortical adenoma was strongly favored in the differential diagnosis. In the year after the resection, symptom flares became less frequent, and in the subsequent 4 years, her skin has been completely clear. After surgery she only took hydroxychloroquine 200 mg daily for her DM.



**Fig 3.** Radiographic imaging. (A) Coronal and (B) axial computed tomography scan images showing a left adrenal mass (yellow arrows).



**Fig 4.** Gross pathology and histology. (A) Gross specimen and (B) low-power view of hematoxylin-eosin stain of resected adrenal mass showing an infarct nodule within the adrenal cortex consistent with an infarcted cortical adrenal adenoma.

## DISCUSSION

Our patient, who had classic cutaneous and histologic findings of DM, initially experienced difficulty with pharmacologic treatment but had significant improvement following resection of an infarct adrenal nodule consistent with an adrenal cortical adenoma. The association of DM with an adrenal tumor is rare; the most similar cases in the literature describe an association between adrenal cancers and polymyositis, which belongs to the same category of idiopathic inflammatory myopathy (IIM) as DM.<sup>2</sup> Although the exact pathophysiologic link between IIMs and malignancies is not fully understood, the prevailing hypothesis is that IIMs can occur as paraneoplastic processes. It is also worth noting that our patient had amyotrophic DM, which represents the minority of DM cases (approximately 10% to 20%).<sup>3</sup>

This case reinforces the importance of a full screening for occult malignancy following a diagnosis of DM. One large retrospective analysis

suggests this screening procedure should include not only age-appropriate tests (mammogram, colonoscopy) but also computed tomography scans of the chest, abdomen, and pelvis and laboratory studies including prostate-specific antigen, urinalysis, complete blood count, and creatine kinase.<sup>4</sup> There appears to be no clear evidence to suggest how frequently this screening should be repeated. As with other paraneoplastic syndromes, although certain cancers are commonly the underlying trigger, any type of neoplastic growth can cause the syndrome. Furthermore, this case illustrates the therapeutic impact in DM of tumor treatment even in benign cases. Although treatment of an underlying malignancy does not always improve DM, in many cases resection results in significant improvement or resolution of symptoms.<sup>5</sup>

## Conflicts of interest

None disclosed.



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