Characteristic vascular finding in TIF1-γ dermatomyositis

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DESCRIPTION
A 72-year-old man presented to the connective tissue disease clinic with a several month history of intense scalp itching and redness. Allergic contact dermatitis had initially been suspected and patch testing was performed, however, topical steroids and allergen avoidance had not provided relief. On physical examination, the patient had many small hypopigmented macules on a background of erythema affecting the scalp, face and shoulders (figure 1). Dermoscopy revealed dilated tortuous capillary loops dispersed over avascular atrophic areas corresponding to clinically red and white areas, respectively (figure 2). The clinical appearance was consistent with a recently described capillary vasculopathy, termed ‘red on white’, which is associated with transcriptional intermediary factor 1-γ (TIF1-γ) autoantibodies in dermatomyositis. This finding is distinct from the more classic poikiloderma seen in many dermatomyositis patients. Our patient reported no muscle weakness. Skin biopsy demonstrated interface dermatitis with vacuolar changes, perivascular lymphocytic infiltrate and dermal mucin deposition. A comprehensive myositis panel confirmed the presence of antibodies to TIF-1γ and a diagnosis of dermatomyositis was made.

Hypopigmented and telangiectatic (‘red on white’) patches are characteristic findings in patients with TIF1-γ autoantibodies and recognising this vasculopathy can facilitate prompt diagnosis. The exact mechanism underlying this vasculopathy is not completely understood, although there is evidence of a true inflammatory small vessel vasculitis early on in the disease course and a non-inflammatory occlusive vasculopathy with capillary drop outs occurring later on. We hypothesise that the ‘red on white’ phenomenon may reflect early inflammatory vasculitis (red, dilated) and late non-inflammatory occlusive vasculopathy (white, drop outs).

The presence of anti-TIF1-γ antibodies is associated with increased risk of malignancy in dermatomyositis and should prompt the clinician to initiate age-appropriate cancer screenings. A significant

Figure 1 Hypopigmented patches on a background of erythema on the scalp, face and shoulders.

Figure 2 Dermoscopy of the scalp at initial presentation showing dilated capillary loops and dropouts.

Figure 3 Dermoscopy of the scalp after 6 months of treatment showing resolution of dilated capillary loops.
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proportion of anti-TIF1-γ positive adult patients with dermatomyositis have no detectable malignancy at the time of the disease onset, thus continued cancer surveillance and reassessment is critical in patients who relapse with dermatomyositis symptoms. This patient was started on oral methotrexate 10 mg weekly and hydroxychloroquine 200 mg two times per day with a good clinical response. A comprehensive cancer screening was performed and did not reveal evidence of underlying malignancy.

At 6 months follow-up, dermoscopy of the scalp demonstrated remarkable improvement in previously observed dilated capillary loops (figure 3).

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REFERENCES

Learning points
► Red on white patches are a characteristic finding in patients with antitranscriptional intermediary factor1-γ (anti-TIF1-γ) dermatomyositis.
► A diagnosis of anti-TIF1-γ dermatomyositis warrants age-appropriate cancer screening for underlying malignancy.
► Recognition of red on white patches can facilitate prompt diagnosis treatment, particularly in the absence of muscle weakness or other classic dermatomyositis skin findings as seen in this case.