The pulseless patient: Profound vasculopathy as the presenting feature of fulminant dermatomyositis and response to therapy

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The idiopathic inflammatory myopathies are uncommon childhood conditions that present with proximal muscle weakness and characteristic cutaneous manifestations. Juvenile dermatomyositis (JDM) is the most common subtype, comprising 80% to 85% of cases.[1] The overall prognosis of JDM has improved considerably over the last few decades because of more aggressive use of corticosteroids and other immunosuppressant medications.[2] JDM is known to cause myriad symptoms that can vary from patient to patient, including calcinosis cutis, ulcerations, and, most significantly, cutaneous and systemic vasculopathy. The latter is the chief cause of mortality in JDM. We present a complex case of a JDM-like presentation of dermatomyositis (DM) in a young adult that was complicated by widespread vascular occlusive disease.

Abbreviations used:

DM: dermatomyositis
JDM: juvenile dermatomyositis

CASE REPORT

A 21-year-old young adult with no significant medical history presented to the hospital with a 3-week history of severe proximal muscle weakness, abdominal pain, profound dysphagia, myalgia, and arthralgias. He had been started on prednisone, 60 mg/d, 10 days before admission, but his symptoms continued to worsen.

He was found to have nonpruritic poikilodermatous patches with fine scale over his eyelids (Heliotrope rash; Fig 1, *A*), scalp, upper shoulders, and extremities in addition to his metacarpophalangeal joint, proximal interphalangeal, and DIP joints with a papular component (Gottron papules). Cuticular overgrowth and hemorrhage were noted. Violaceous erythema with fine scale was additionally noted over his elbows (Fig 1, *B*) and knees (Gottron sign). Faint flagellate erythema was noted over his bilateral upper extremities (Fig 1, *C*). Moreover, he was noted to have striking retiform purpura over the bilateral lower extremities and absent palpable dorsalis pedis, tibialis posterior, and radial pulses on examination (Fig 1, *D*).

Arterial Doppler studies found toe brachial pressures of zero bilaterally. Magnetic resonance imaging of the lower extremity found diffuse proximal muscle edema, consistent with DM. Given his persistent abdominal pain, abdominal computed tomography was ordered, which found colonic and small bowel wall thickening consistent with vasculitis. Testicular ultrasound likewise found evidence of vasculitis.

Given concern for fulminant DM with vasculopathic features more commonly seen in the juvenile variant, the patient was started on a 6-day course of pulse-dose steroids (methylprednisolone 1000 mg/d) as well as a heparin drip given the severity of the vasculopathy with return of pulses within hours of starting therapy. He additionally received a single dose of intravenous immunoglobulin.

Initial laboratory evaluation found positive antinuclear antibodies but negative ds-DNA, Smith, SSA, SSB, Scl-70, RNP, Jo-1, Mi-2, PL-7, PL-12, p155/140,

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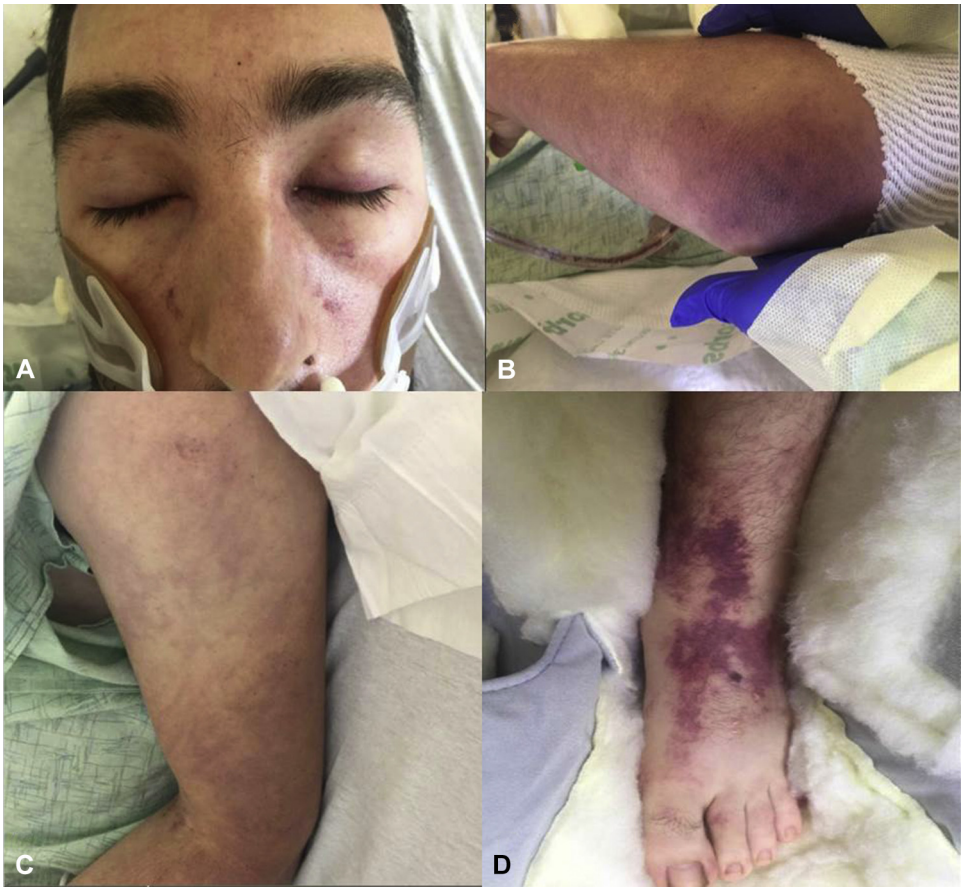

Fig 1. Cutaneous manifestations of JDM. **A**, Faint violaceous erythema over bilateral eyelids (heliotrope rash). **B**, Violaceous erythema with fine scale over elbow (Gottron sign). **C**, Faint flagellate erythema over upper extremities. **D**, Retiform purpura over bilateral lower extremities.

EJ, Ku, U2, SRP, OJ, myeloperoxidase, proteinase 3, and rheumatoid factor antibodies. Hypercoagulable panel was negative (including factor V Leiden, protein C/S, cryoglobulins, antiphospholipids, B2-microglobulin, and anticardiolipin antibody). Creatine kinase was 44,957 initially and downtrended with immunosuppressive therapy. Biopsy of the flagellate erythema of the left upper extremity found rare necrotic keratinocytes suggestive of subtle interface dermatitis, compatible with DM (Fig 2, *A*), and biopsy of the retiform purpura on the left lower extremity found vascular occlusion with fibrin thrombi, compatible with DM-associated vasculopathy (Fig 2, *B*).

A diagnosis of DM with profound vasculopathy was made. The hospital course was complicated by aspiration in the setting of his severe progressive dysphagia and acquired aspiration pneumonia, requiring temporary intubation as well as several days of empiric antibiotics. As his condition improved, he was transitioned back to prednisone, 60 mg/d, and was started on mycophenolate mofetil, which was given at 1500 mg twice daily upon discharge with gradual prednisone taper (by 5 mg/wk).

The patient was stable on this regimen during the prednisone taper but complained of floaters in his right eye when the prednisone dose reached a total of only 5 mg/d, which was approximately 70 days after initial presentation. He was referred to the ophthalmology department, and evaluation found right branch retinal artery occlusion with retinal vasculitis (Fig 3, *A-D*). His prednisone was increased to 60 mg/d, and the patient was transitioned from mycophenolate mofetil to methotrexate, 20 mg/wk, in addition to hydroxychloroquine, 200 mg twice daily, alternating with 200 mg once daily. His visual impairment improved rapidly in 7 to 10 days, and the vasculitis completely resolved after 2 months. Currently, he

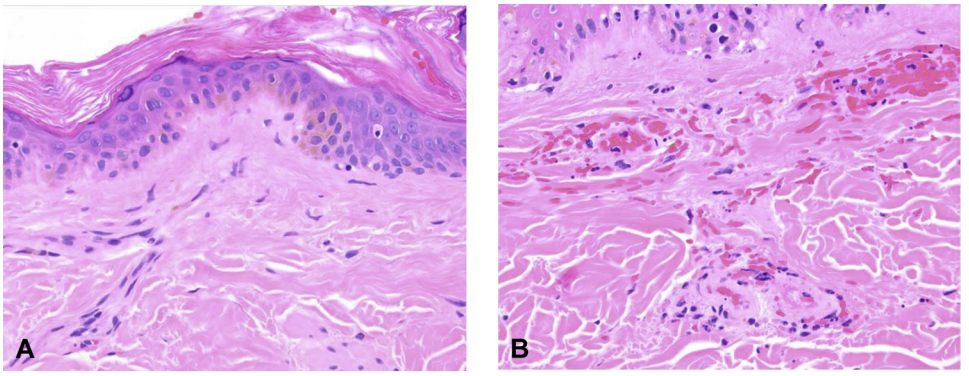

Fig 2. Histopathologic findings in JDM. **A**, Punch biopsy of flagellate erythema on left upper arm shows rare necrotic keratinocytes, suggestive of subtle interface dermatitis, compatible with DM. **B**, Punch biopsy of the right shin shows vascular occlusive disease with fibrin thrombi, compatible with JDM-associated vasculopathy. (Original magnifications: ×40.)

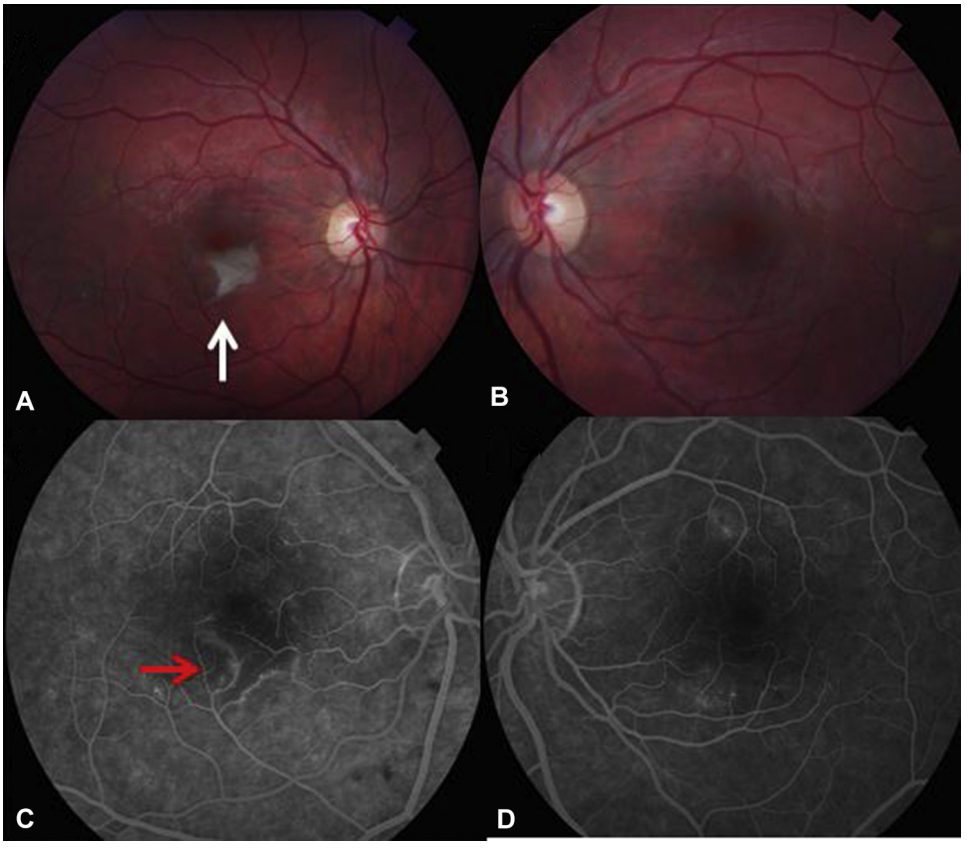

Fig 3. Retinal vasculitis and branch retinal artery occlusion in JDM and response to therapy. Color fundus photographs on initial presentation. **A**, Right eye. Small, wedge-shaped area of retinal whitening and edema (*white arrow*) noted inferior to the fovea consistent with branch retinal artery occlusion. **B**, Left eye with normal findings. Fluorescein angiography on initial presentation. **C**, Right eye with angiographic leakage of retinal arteries perfusing the inferior macula consistent with retinal vasculitis (*red arrow*). **D**, Left eye with mild patches of angiographic leakage from subtle retinal vasculitis despite lack of symptoms.

is on a slow taper of prednisone, methotrexate, 20 mg/wk, and hydroxychloroquine, 200 mg twice daily, alternating with 200 mg daily, with no complications.

DISCUSSION

Adult-onset DM and JDM are considered part of same disease spectrum. Cutaneous and systemic vasculitis, including vasculopathy of the gastrointestinal

tract, is more commonly seen in JDM than adult-onset DM. Our 21-year-old young adult patient presented with features more commonly seen in JDM. His vascular occlusive disease was widespread, with involvement of the gastrointestinal tract and other organs.

Interestingly, our patient's condition continued to deteriorate despite being on oral prednisone, 60 mg/d for 10 days prior to admission. It is likely that the oral prednisone was not absorbed because of gastrointestinal vasculitis, thus transitioning him to intravenous high-dose steroids was a pivotal moment in his treatment. It is of paramount importance to intensely treat DM patients with vasculopathy at diagnosis to avoid vasculopathy-related morbidity and mortality.[3] Gastrointestinal vasculopathy is the most common cause of mortality in JDM. The mortality rate for JDM used to be around 50%, but is currently 3% to 5.5%[3,4] because of more aggressive treatment.

Retinal vasculitis is a rare complication of DM. The few published case reports describe retinal hemorrhages and cotton wool spots.[5-7] This is a single published case report of severe vision loss from central retinal vein occlusion associated with DM.[8] To our knowledge, macular branch retinal artery occlusion associated with DM has not been described. All improved with treatment. With any case of DM with vasculopathy, prompt recognition and treatment is key to avoiding devastating effects of profound vasculopathy.

Of note, we were unable to identify a positive autoantibody in our patient. This was not surprising because 40% to 50% of patients with JDM will not have an (identified) positive antibody.[9]

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