Urticarial Vasculitis and Dermatomyositis in a Patient With Nasopharyngeal Carcinoma

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Urticarial vasculitis has rarely been described in association with polymyositis. We report the case of a 37-year-old man with dermatomyositis and nasopharyngeal carcinoma who presented initially with urticarial vasculitis. The lesions of urticarial vasculitis were initially photodistributed, indicating photosensitivity. The patient was treated with systemic steroids, chemotherapy (cisplatin and fluorouracil), and radiation therapy. The tumor and urticarial vasculitis completely resolved, and the myositis improved.

Cutis. 2003;72:399-402.

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestations, including heliotrope rash, Gottron papules, poikiloderma, and periungual telangiectasia. Polymyositis is thought to differ only by the absence of characteristic skin changes.[1,2] Urticarial vasculitis sometimes occurs during the course of connective tissue disease. However, the association of urticarial vasculitis and polymyositis is only rarely reported.[3,4] Because so few cases are described in the literature, the significance of this unusual association is difficult to ascertain. We report a case of dermatomyositis with nasopharyngeal carcinoma in a patient who presented initially with urticarial vasculitis.

Case Report

In March 2001, a 37-year-old Taiwanese man visited our outpatient department with a 2-month history of itching as a result of multiple, round-to-polycyclic erythematous plaques on his face, neck, chest, and upper back (Figure 1). Neither fever nor additional symptoms were expressed at the time. His medical history was unremarkable, and no drug history was noted before this episode. Histopathology of the skin lesion showed perivascular mononuclear cell and neutrophilic infiltration. Results of the lupus band test were negative. Unfortunately, the patient's skin lesions were not relieved with antihistamines or topical steroids.

An enlarged neck mass gradually formed, and the patient experienced neck pain in July 2001. The patient also had lost 6 kg of body weight in 6 months. A nasopharyngoscopic biopsy of the mass was performed, and nasopharyngeal carcinoma was diagnosed. Magnetic resonance imaging showed that the tumor had invaded to the skull base, with metastasis to the bilateral neck lymph nodes. Whole-body bone scanning showed local hyperemia of the nasopharyngeal roof. No distant metastasis was found on either chest radiography or abdominal sonography. In addition, the strength of the patient's extremity muscles, particularly the proximal muscles, was reduced (muscle power grade, 3). A complete blood cell count revealed the following results: white blood cell count, 16,500/µL (neutrophils, 83% ; lymphocytes, 13%; atypical lymphocytes, 1%); hematocrit level, 40.5%; and platelet count, 492×10^3 µL. Serum chemistry results showed the following levels: aspartate aminotransferase, 543 U/L (reference range, 0–34 U/L); alanine aminotransferase, 737 U/L (reference range, 0–36 U/L); creatine kinase (CK), 14,910 U/L (reference range, 15–130 U/L) with CK-MB fraction, 6.6%, and CK-MM fraction, 93.4%; and myoglobin, 1965.2 µg/L (reference range, <80 µg/L). Alkaline-phosphate, bilirubin, creatinine, and electrolyte levels were normal. Immunoglobulin G antibodies against the Epstein-Barr virus capsid antigen were present, with a titer

Accepted for publication April 22, 2003.
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The authors report no conflict of interest.
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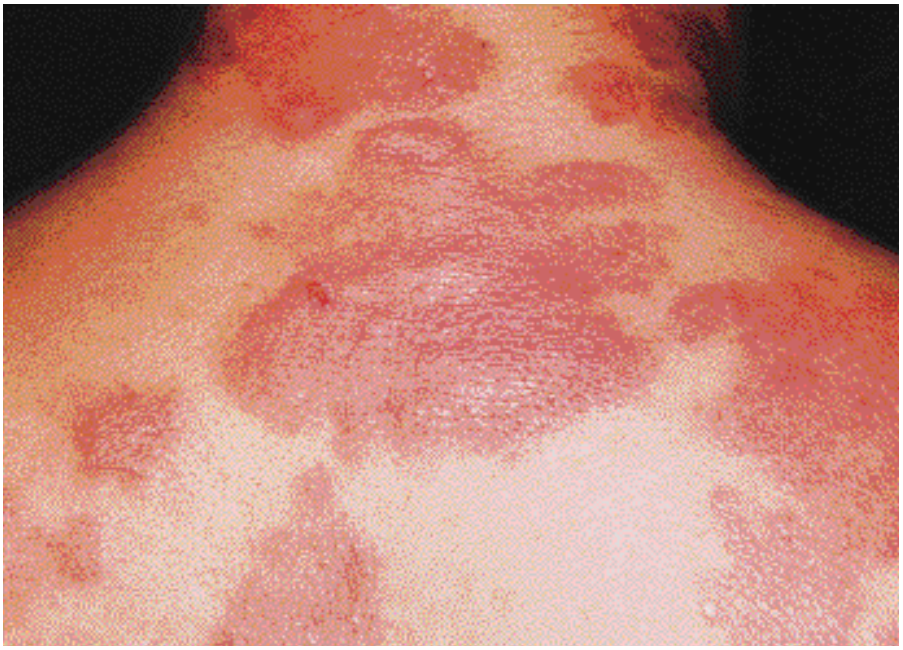

Figure 1. Multiple, round-to-polycyclic erythematous plaques on the neck and upper back.

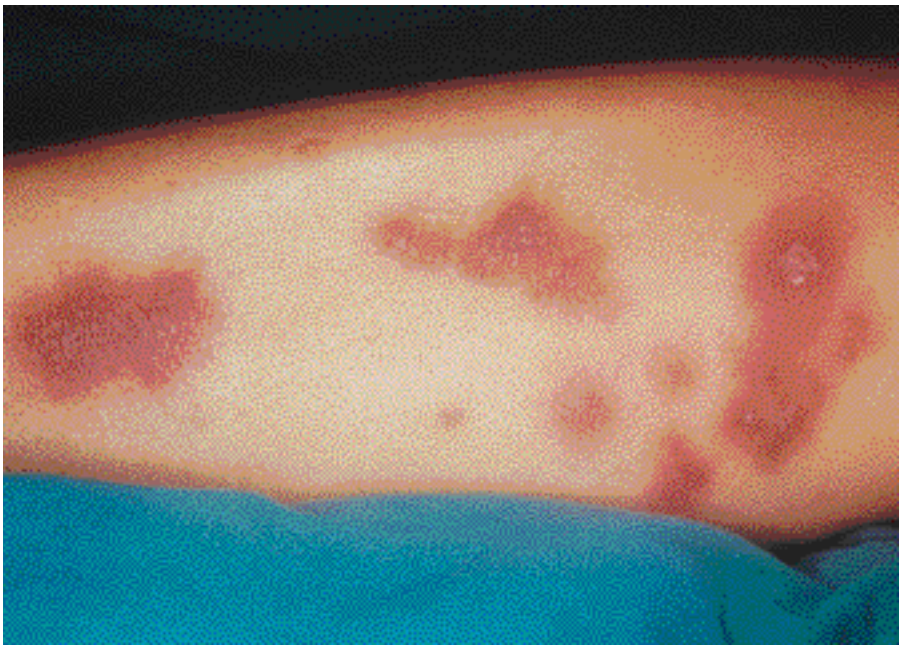

Figure 2. Multiple erythematous-to-purpuric plaques on the arm.

of more than 1:1280. Results of antinuclear antibody tests and hepatitis B and C serologic tests were negative. Histopathology of the muscle biopsy sample showed myopathic changes consistent with polymyositis.

Dermatologic consultation was requested because of the progression of the skin lesions. Findings from the physical examination showed multiple erythematous-to-purpuric plaques, as well as hyperpigmented patches on the face, neck, chest, upper back, and extremities (Figure 2). The skin lesions were itchy and stinging. Individual lesions lasted for several days and then resolved with a hyperpigmented hue; however, new lesions kept developing. The heliotrope sign and Gottron papules were absent. Repeat biopsy of the skin was performed and showed perivascular neutrophilic infiltration, nuclear dust, and mild red blood cell extravasation (Figure 3).

The patient was diagnosed with polymyositis associated with urticarial vasculitis and received prednisolone 1 mg/kg per day. Nasopharyngeal

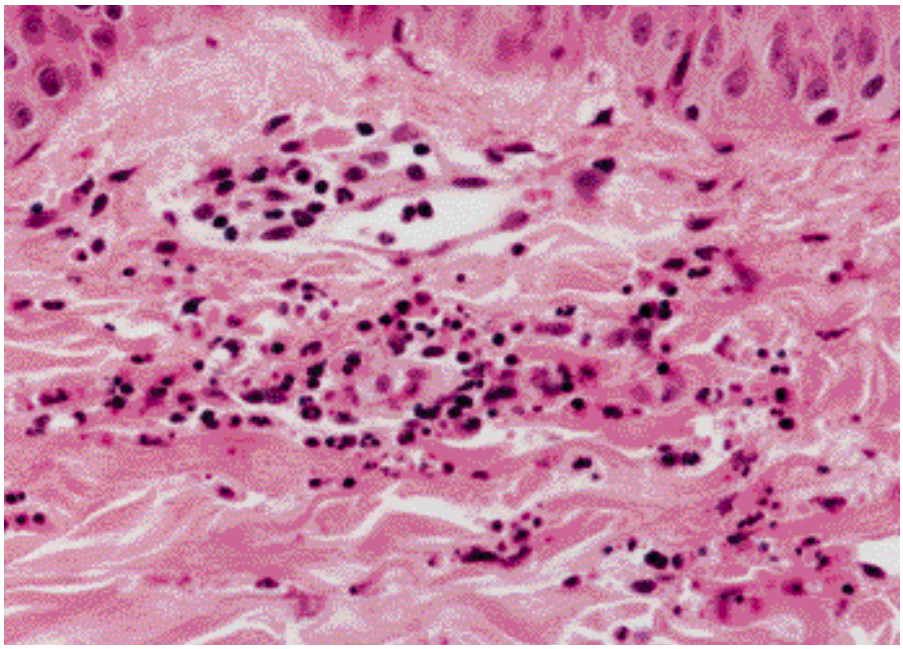

Figure 3. Perivascular neutrophilic infiltration, nuclear dust, and mild red blood cell extravasation (H&E, original magnification ×400).

carcinoma was treated with combination radiation therapy and chemotherapy (cisplatin and fluorouracil). The patient's skin lesions gradually faded and completely resolved after 2 months. He returned to our department as an outpatient after another 2 months because of radiation dermatitis. The heliotrope sign and Gottron papules were noted at that time, and dermatomyositis was diagnosed. No further treatment except prednisolone and chemotherapy was prescribed. Levels of various muscle enzymes returned to the reference range, and muscle strength was partially restored 6 months after the treatment was initiated. The neck mass also completely disappeared at about the same time.

Comment

The term *urticarial vasculitis* was introduced to describe a syndrome characterized by persistent urticarial wheals, with histologic features of either a leukocytoclastic or a lymphocytic vasculitis. A single urticarial vasculitis lesion typically persists for longer than 24 hours and resolves with residual hyperpigmentation, which indicates red blood cell extravasation. Causes or associated factors of urticarial vasculitis include idiopathic reasons, hypocomplementemic urticarial vasculitis syndrome, systemic lupus erythematosus, Sjögren syndrome, neoplasms, drugs, and various viral infections.[5] In our patient, Sweet syndrome had to be included in the differential diagnosis because of the similar clinical presentation and its scanty vascular damage in microscopic findings. However, patients with Sweet syndrome often have fever and appear ill. The skin lesions are generally painful and present in the same stages of development, as opposed to itchy and stinging, and show various stages of evolution and resolution, as in cases of urticarial vasculitis.[6] After summing up these clinical differential diagnoses, urticarial vasculitis was diagnosed in our patient.

Callen and Dubin[3] first reported urticarial vasculitis associated with polymyositis in 1978 in a patient who developed urticarial lesions with an exacerbation of muscular symptoms. Results of a skin biopsy showed leukocytoclastic vasculitis. Kao and Zeitz[4] reported a similar case in 1995. Biopsies of the skin and muscle were performed, and results showed lymphocytic vasculitis. No crossover with other connective tissue diseases, infectious agents, or therapeutic agents can explain the cause of the skin lesions in the reported cases. In addition, the parallel course of the urticarial vasculitis and the muscular involvement supports a common causative relation.[3,4] Interestingly, in our case, urticarial vasculitis was part of the initial presentation of dermatomyositis, and the skin lesions were initially photodistributed. These findings have not been mentioned by previous authors. Photosensitivity is an important cutaneous feature of dermatomyositis.[7] Also, UV radiation may induce urticarial vasculitis.[8] These factors may explain the aforementioned special presentation in our case.

The frequency of an underlying malignancy in patients of dermatomyositis is 2.4 to 12.6 times higher than that of the general population.[9,10] The malignancy can precede, occur concurrently with, or follow the diagnosis of dermatomyositis. The types of neoplasms found in association with dermatomyositis roughly parallel those observed in the general population. Carcinomas of the lung, stomach, ovary, and breast commonly are seen in dermatomyositis.[11] In Taiwan, nasopharyngeal carcinoma is the most common associated malignancy.[12] Although vasculitis occurs somewhat frequently in juvenile dermatomyositis, it is rare in adult-onset disease.[13] The presence of cutaneous vasculitis in adult-onset dermatomyositis has a significantly higher risk of malignancy compared with the risk in the absence of vasculitis. Thus, it has predictive value in determining the presence of concomitant neoplastic diseases.[13,14]

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