Purpuric lesions in an elderly woman

A diagnosis of asthma 3 years earlier shed some light on the lesions that had recently erupted on this patient's trunk and extremities.

Rami Abadi, MD; Habib Dakik, MD; Ossama Abbas, MD
Department of Dermatology (Drs. Abadi and Abbas); Department of Internal Medicine (Dr. Dakik), American University of Beirut Medical Center, Lebanon

ossamaabbas2003@yahoo.com

DEPARTMENT EDITOR
Richard P. Usatine, MD
University of Texas Health Science Center at San Antonio

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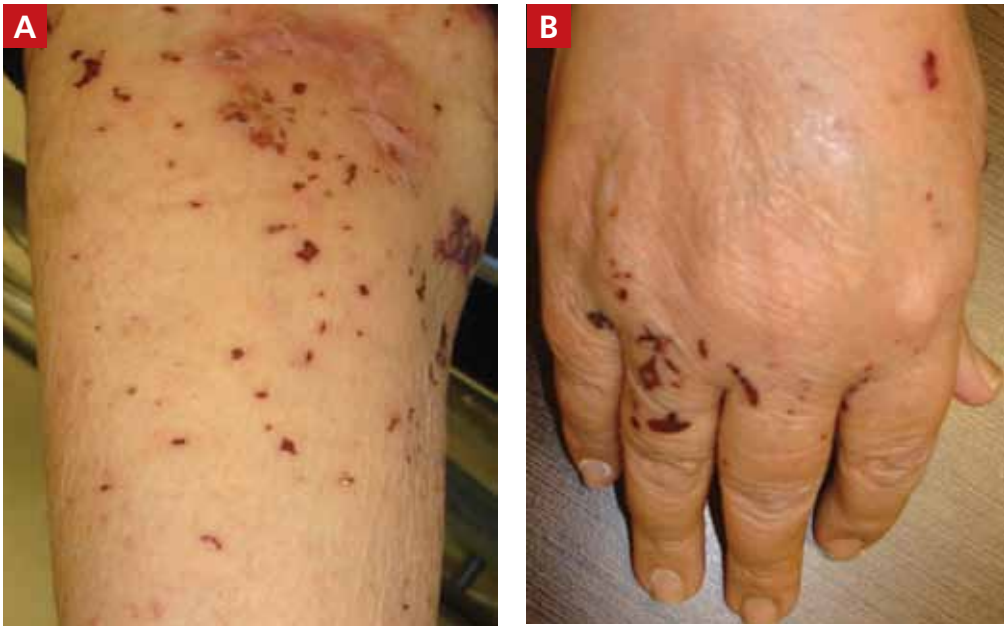
A 68-YEAR-OLD WOMAN presented with a 5-day history of extensive pruritic purpuric skin lesions of varying sizes on her trunk and extremities (**FIGURE 1A AND 1B**). In addition, the patient had a few nonblanching, erythematous macules on her extremities.

Three years earlier, the patient had been given a diagnosis of asthma and since then had been maintained on fluticasone propionate oral inhaler. Recently, she'd experienced unintentional weight loss and malaise.

The patient had no neurological complaints and her family history was negative for a similar condition. We performed a punch biopsy.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE 1
Purpuric macules and papules on the right leg (A) and hand (B)



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Diagnosis: Churg-Strauss syndrome

Churg-Strauss syndrome (CSS)—also known as allergic granulomatosis and angiitis—is a rare multisystemic vasculitis of small- to medium-sized vessels characterized by asthma, chronic rhinosinusitis, and prominent peripheral blood eosinophilia.[1,2] Mean diagnosis age is 50 years with no gender predilection.[2] Any organ system can be affected, although the lungs are most commonly involved, followed by the skin.[1,2]

Based on criteria from the American College of Rheumatology, the diagnosis of CSS can be made if 4 of the following 6 criteria are met: (1) asthma, (2) eosinophilia >10% on a differential white blood cell (WBC) count, (3) paranasal sinus abnormalities, (4) a transient pulmonary infiltrate detected on chest x-ray, (5) mono- or polyneuropathy, and (6) a biopsy specimen showing extravascular accumulation of eosinophils.[2]

Skin biopsy specimen from our patient showed leukocytoclastic vasculitis with prominent tissue eosinophilia. Laboratory studies showed an elevated WBC count of 12,300/mcL (reference range, 4500-11,000/mcL), and eosinophilia of 40% (reference range, 1%-4%). A serologic test for perinuclear pattern antineutrophil cytoplasmic antibodies (p-ANCA) was positive. (More on this in a moment.) Radiography of the chest showed transient pulmonary infiltrates.

Based on the clinical and laboratory findings, the patient was positive for 4 of 6 criteria and given a diagnosis of CSS.

The most common skin finding is palpable purpura on the lower extremities.

What we know—and don't know—about CSS

The exact etiopathogenesis of CSS is unknown.[2-4] Although ANCAs are detected in about 40% to 60% of CSS patients, it is not yet known whether ANCAs have a pathogenic role.[2-3] Abnormalities in immunologic function also occur, including heightened Th1 and Th2 lymphocyte function, increased recruitment of eosinophils, and decreased eosinophil apoptosis. Genetic factors, including certain interleukin-10 polymorphisms and HLA classes such as HLA-DRB4, may also contribute to CSS pathogenesis.[4]

Three distinct sequential phases have been described, although these are not always clearly distinguishable.[2,5]

- **The first is the prodromal or allergic phase,** which is characterized by the onset of asthma later in life in patients with no family history of atopy. There may or may not be an associated allergic rhinitis.
- **In the eosinophilic phase,** peripheral blood eosinophilia and eosinophilic infiltration of multiple organs (especially the lungs and gastrointestinal [GI] tract) occur.
- **The vasculitis phase** is characterized by life-threatening systemic vasculitis of the small and medium vessels that is often associated with vascular and extravascular granulomatosis.

Cutaneous and extracutaneous findings

One-half to two-thirds of patients with CSS have cutaneous manifestations that typically present in the vasculitis phase.[2,5] The most common skin finding is palpable purpura on the lower extremities. Macular or papular erythematous eruption, urticaria, subcutaneous skin-colored or erythematous nodules, livedo reticularis, and erythema multiforme–like eruption may also be seen.[2,5,6] Skin biopsies will show numerous eosinophils with either leukocytoclastic vasculitis or extravascular necrotizing granuloma.[5]

Extracutaneous manifestations of CSS include renal, cardiac, GI tract, and nervous system involvement.[2,7]

To identify patients with a poor prognosis, the 5-factor score (FFS) can be used. This score assigns 1 point each to GI tract involvement, renal insufficiency, proteinuria, central nervous system involvement, and cardiomyopathy.[7] CSS patients with an FFS ≥2 have a considerably greater risk of mortality.[7]

Treatment involves corticosteroids

Systemic corticosteroids (prednisone, 1 mg/kg/day) are the primary treatment for patients with CSS; most patients improve dramatically with therapy.[2] Adjunctive therapy with immunosuppressive agents such as cyclophosphamide, methotrexate (10-

15 mg per week), chlorambucil, or azathioprine may be needed if a patient does not respond adequately to steroids alone.[2]

Prednisone for our patient

We started our patient on prednisone 1 mg/kg/d. Her skin lesions resolved and subsequent laboratory tests, including eosinophil counts, normalized. Prednisone therapy was gradually tapered over several months to attain the lowest dose required for control of symptoms—in this case, 5 mg/d. **JFP**

CORRESPONDENCE
Ossama Abbas, MD, Associate Professor, Department of Dermatology, American University of Beirut Medical Center, PO Box 11-0236, Riad El Solh, Beirut 1107 2020, Beirut, Lebanon; ossamaabbas2003@yahoo.com

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