Dermacase

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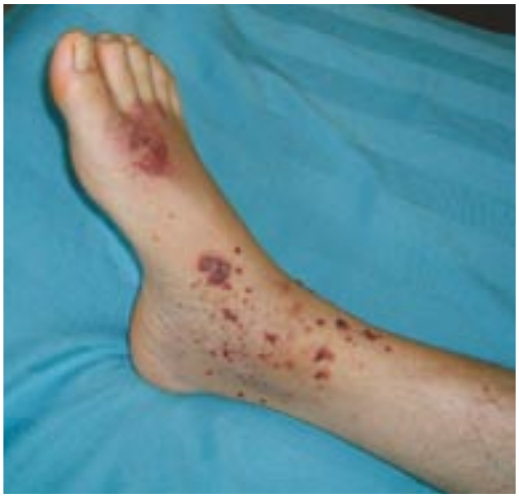

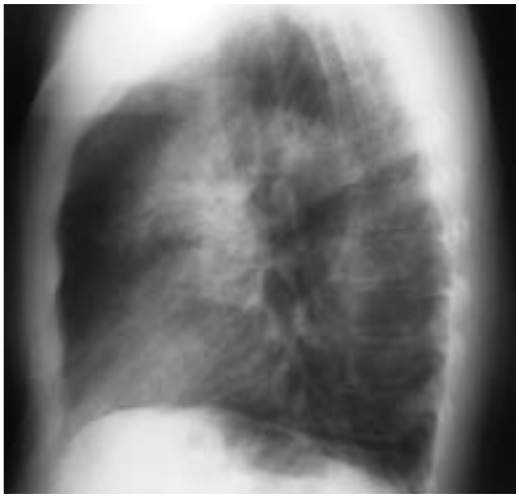

CAN YOU IDENTIFY THIS CONDITION?

A 56-year-old asthmatic man presents with a 2-month history of progressive hemorrhagic bullous rash, joint pain and swelling, decreased sensation in both ankles, and a history of one episode of hemoptysis (about 5 mL) on day of presentation. His current medications include fluticasone propionate and a fluticasone salmeterol combination for asthma control. He says he has no allergies.

Physical examination is unremarkable except for elevated temperature (38°C), crackles at the lung bases, reduced sensation up to the ankles in both feet, and palpable purpura with acral hemorrhagic blisters on his hands, feet, and sacrum. Test results were negative, except for elevated white blood cell count (18.6, with 40% eosinophils), erythrocyte sedimentation rate (40), and respiratory frequency (95). Chest x-ray examination showed a dense air-space consolidation, predominantly in his right and left upper lung anterior segments and right lower lung superior segment; no pleural effusions; and heart and mediastinum within normal limits.

The most likely diagnosis is:

1. Polyarteritis nodosa
2. Wegener granulomatosis
3. Rheumatoid arthritis
4. Churg-Strauss syndrome

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Answer to Dermacase

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4. Churg-Strauss syndrome

Churg-Strauss syndrome (CSS), or allergic granulomatous angiitis, is a rare disorder characterized by asthma, transient pulmonary infiltrates, hypereosinophilia, and systemic vasculitis.[1] Characteristic skin changes are present in 70% of patients and can lead to an early and potentially life-saving diagnosis.[2]

Churg-Strauss syndrome affects medium and small vessels and has been classified into a group of small vessel vascular conditions that includes Wegener granulomatosis (WG) and microscopic polyangiitis (MP). This group of vascular conditions is associated with neutrophil cytoplasmic antigen (ANCA) antibodies.

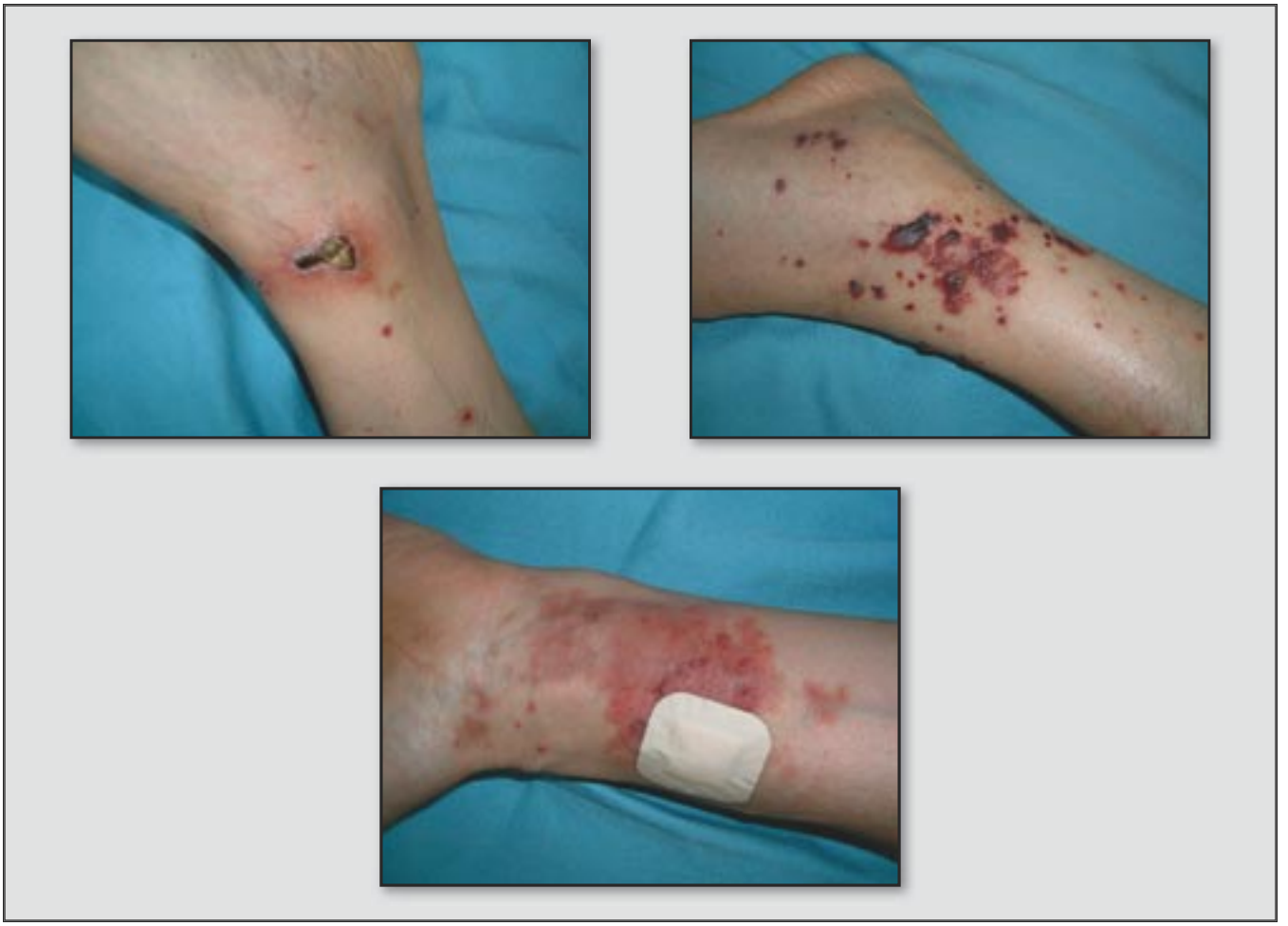
The etiology of CSS is not well understood. It is thought to involve an autoimmune response and proliferation of activated eosinophils.[3] According to the American College of Rheumatology, the presence of any four or more of the following six criteria has a sensitivity of 85% and a specificity of 99.7% for diagnosis of CSS:

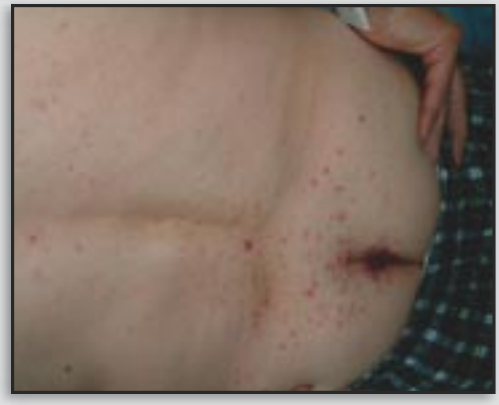
- asthma,
- eosinophilia greater than 10% on a differential white blood cell count,
- mononeuropathy or polyneuropathy,
- non-fixed pulmonary infiltrates,
- paranasal sinus abnormality, and
- extravascular eosinophils.[4]

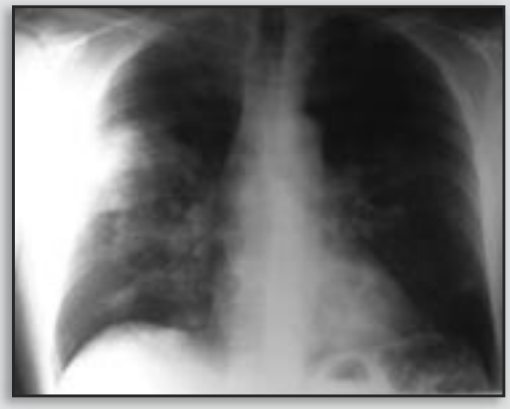
Necrotizing glomerulonephritis is very common in WG and MP, but uncommon in CSS. Cardiac, renal, gastrointestinal, and central nervous system involvement are associated with a worse prognosis.

Three types of skin lesions have been described in CSS: an erythematous, maculopapular eruption, resembling erythema multiforme; hemorrhagic lesions that vary from petechiae to ecchymoses and are often associated with urticarial wheal formation; and subcutaneous and cutaneous nodules.[5] Several types of lesions can occur simultaneously on the same patient. Common sites of involvement are the flexor and extensor surfaces of the extremities, back, and abdomen.

Subcutaneous nodules tend to occur on the scalp and extremities but occur less frequently on the trunk. Often, they are tender, persist for up to 3 months, and resolve with a scar.[1] Patients might also have ulcers, livedo reticularis, vesicles, and facial edema.[2]







Differential diagnosis is very broad and can incorporate polyartheritis nodosa, WG, MP, rheumatoid arthritis, cryoglobulinemia, subacute bacterial endocarditis, lymphoproliferative disorders, and chronic active hepatitis.[6]

Laboratory investigations should include hematology workup for anemia and eosinophilia; renal tests for elevated serum blood urea nitrogen and creatinine, abnormal urine sedimentation, proteinuria, microscopic hematuria, and red blood cell casts; erythrocyte sedimentation rate and rheumatoid factor (usually elevated); and ANCA (positive in 50% to 70% of cases).[7] Other tests might include chest x-ray examination, electrocardiography and echocardiography, and gastrointestinal endoscopy if bleeding is suspected.

Biopsy is usually helpful in making the diagnosis. Biopsy sites include skin, lung, kidney, muscle, and sural nerve. Three histologic findings are typical in CSS: necrotizing granuloma, necrotizing vasculitis, and eosinophilic infiltration.[2]

Prognosis is unfavourable when major organ systems are involved. Corticosteroids have been standard therapy for CSS, with an initial dose of prednisone of 60 to 100 mg daily. Cyclophosphamide can be added to the corticosteroid regimen to prevent or reduce end organ damage.[2] Erythrocyte sedimention rate and eosinophil count can be used to monitor response to therapy.[1] Patients usually need long-term immunosuppressive medications. Rheumatology consultation is strongly recommended.

References

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