Leukocytoclastic Vasculitis: A Marker of Underlying Malignancy

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A 62-year-old man presents with a violaceous, nonpruritic eruption that arose 2 weeks earlier on the hands and feet, including the palms and soles, and spread to the arms and legs (Figure 1). Over the past 3 to 4 weeks, he has had malaise, nonproductive cough, and a decline in mental status but no fever, headache, nausea, light-headedness, hemoptysis, or melena.

The patient, a 30-pack-year smoker, has a history of hypertension, type 2 diabetes mellitus, congestive heart failure, and coronary artery disease. He admits to moderate alcohol consumption, denies injection drug use, and reports no recent outdoor activities or tick exposure. Two weeks earlier, he started taking amitriptyline and clopidogrel. Temperature is normal. No lymphadenopathy is noted. Mucous membranes are unaffected. There is no evidence of a tick bite.

The patient has numerous papules and plaques on the extremities that are nonblanchable on diascopy; hemorrhagic bullae and targetoid lesions are also present (Figures 2 and 3). A punch biopsy of the bullae reveals a mixed, predominantly neutrophilic, perivascular infiltrate obliterating vessel walls, with fibrinoid necrosis, leukocytoclasis, erythrocyte extravasation, and overlying epidermal necrosis (Figure 4). These findings support the diagnosis of acute leukocytoclastic vasculitis.

The patient is hospitalized and treated empirically with doxycycline, systemic corticosteroids, and supportive therapy. A chest radiograph and results of blood, urine, and cerebrospinal fluid cultures and serologic tests for Rickettsia rule out an underlying infectious cause. Because of the possibility of an allergic drug reaction, amitriptyline and clopidogrel are withheld. The patient is discharged 1 week after admission, and prednisone is continued.

A week later, the patient returns with worsening dyspnea, dry cough, pitting edema, and episodic delirium with fluctuating levels of consciousness. The skin eruption has spread to the trunk and face. A CT scan of the chest demonstrates a soft tissue mass in the right upper lobe of the lung, with bilateral hilar lymphadenopathy. Bronchoalveolar lavage reveals non-small-cell carcinoma of the lung. The cytologic features include single cells and cell clusters that demonstrate irregular hyperchromatic nuclei with a high nuclear-to-cytoplasmic ratio (Figure 5).

Two months after his initial evaluation, the patient died of complications of the lung carcinoma. The cutaneous vasculitis had persisted until his death.

**PARANEOPLASTIC CUTANEOUS LEUKOCYTOCLASTIC VASCULITIS**

**Leukocytoclastic vasculitis.** This uncommon small-vessel inflammatory disorder characteristically presents as nonpruritic, palpable purpura of the upper dermis in areas of dependency and localized pressure. Although more than half of the cases are idiopathic, the sudden appearance of leukocytoclastic vasculitis may signal an underlying systemic disease, such as infection, allergic drug reaction, autoimmune disorder, or malignancy.

**A paraneoplastic syndrome.** The pathogenesis of leukocytoclastic vasculitis as a paraneoplastic syndrome is not clearly understood. It has been speculated that neoplastic cells release unknown immunogenic factors into the circulation; this results in the deposition of immune complexes within vessel walls and the development of vasculitis. Circulating immune complexes occur in one third to one half of all patients with cancer, although neoplasms seldom lead to vasculitis. Leukocytoclastic vasculitis is more likely to be linked to malignancy in patients older than 50 years. Cutaneous vasculitis may precede the diagnosis of cancer by weeks, months, or even years and is generally associated with a worse prognosis. Although more frequently related to hematogenous malignancies (lymphoma and leukemia), paraneoplastic leukocytoclastic vasculitis...
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may also occur in association with solid tumors, such as squamous cell carcinoma of the lung.10-12

Making the diagnosis. Although this patient's leukocytoclastic vasculitis was initially thought to be an adverse drug reaction, the progression of the cutaneous eruption over 2 months, despite corticosteroid therapy and withdrawal of the suspicious agents, suggested a paraneoplastic dermatosis. Moreover, the concurrent onset and parallel course of the leukocytoclastic vasculitis and the lung neoplasm pointed to a paraneoplastic process.Key Point for Your Practice

Early recognition of leukocytoclastic vasculitis as a potential marker of malignancy may allow for more timely diagnosis, more successful treatment, and improved survival.8,10 Always consider an associated malignancy in patients who present with vasculitis of unknown cause, especially in those older than 50 years.8,10


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