Urticarial rash with leukocytosis and monoclonal gammopathy

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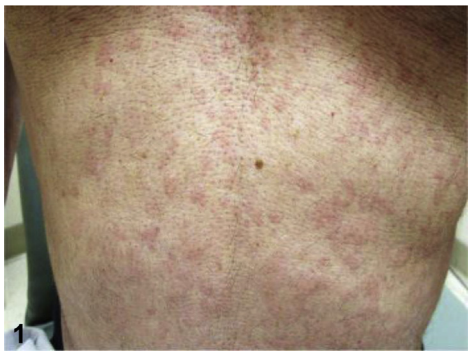

Fig 1. By Devea R. De, BS; Alicia Goldenberg, MD; Paul N. Bogner, MD; Susan Pei, MD, FAAD.

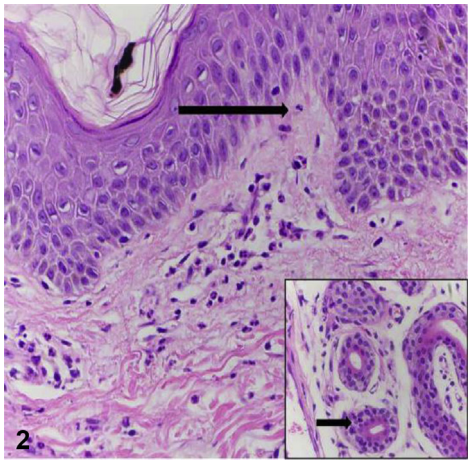

Fig 2. By Devea R. De, BS; Alicia Goldenberg, MD; Paul N. Bogner, MD; Susan Pei, MD, FAAD.

CASE PRESENTATION

A 58-year-old man is being evaluated for a 4-year history of a diffuse, intermittent, nonpruritic rash associated with intermittent arthralgia, myalgia, generalized lymphadenopathy, night sweats, and unintentional weight loss of 10 pounds over 8 months. The symptoms did not appear in a predictable pattern each day or week. The rash was not responsive to oral antihistamines. Laboratory tests over the 4 years showed persistent leukocytosis (range 13,000-20,200/μL[13.5-20.2$\times10^9$/L]) with neutrophilia (range 11.2-17.3$\times10^3/\mu$L [11.2-17.3 10^9/L]), worsening anemia (hemoglobin concentrations of 13.2-11.0 g/dL [132-110 g/L]), elevated erythrocyte sedimentation rate (92 mm/h), C-reactive protein (154 mg/L), and ferritin (494 ng/mL [494 μg/L]), and

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monoclonal IgM-κ gammopathy. Workup for Lyme disease, hepatitis B and C, HIV, tuberculosis, and syphilis was negative. Antinuclear antibody, anti—double- and single-stranded DNA, rheumatoid factor, anti-SSA, and anti-SSB were negative, and complement levels were within normal limits. Bone marrow and excisional lymph node biopsies were unrevealing. Physical examination revealed erythematous wheals coalescing into plaques scattered symmetrically on the trunk and extremities (Fig 1). A punch biopsy of the left arm was performed (Fig 2).

Question #1: What is the most likely diagnosis?

A. Adult-onset Still disease (AOSD)

B. Schnitzler syndrome (SS)

C. Systemic lupus erythematosus (SLE)

D. Cryopyrin-associated periodic syndrome

E. Urticarial vasculitis

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Conflicts of interest

The authors have no conflicts of interest to declare.