A previously healthy 36-year-old man presented with a 1-month history of fever and pain in both shoulders and knees, which had been preceded by a sore throat 2 weeks before the onset of fever. Laboratory studies were notable for a white-cell count of 13,800 per cubic millimeter (85% neutrophils), a C-reactive protein level of 26 mg per deciliter (reference value, ≤0.3), and an antistreptolysin O titer of 1478 IU per milliliter (reference value, <241). Transthoracic echocardiography revealed mild aortic regurgitation. His fever and arthralgias abated after the administration of a nonsteroidal antiinflammatory drug. One week later, painless, non-pruritic, red annular macules appeared on the upper limbs and abdomen (Panel A shows the right forearm; black ink dots indicate the diameter of one lesion 10 hours before the time that the photograph was taken). The rash migrated within hours and then faded over the course of a few days while new lesions appeared (Fig. S1 in the Supplementary Appendix, available at NEJM.org). Skin biopsy revealed perivascular infiltration of lymphocytes and neutrophils in the dermis (Panel B, hematoxylin and eosin). Acute rheumatic fever with erythema marginatum was diagnosed. Erythema marginatum, an evanescent nonpruritic macular rash, is one of the major Jones criteria for the diagnosis of acute rheumatic fever. The patient began taking amoxicillin for secondary prophylaxis of rheumatic heart disease. The rash disappeared completely 4 months after presentation, and the antistreptolysin O titer decreased to 246 IU per milliliter 12 months after presentation.

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