

Cardiac Vasculitis in Henoch-Schönlein Purpura

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A 63-year-old man presented with a 1-week history of bloody diarrhea, abdominal pain, nausea, arthralgias, and fatigue. Physical examination revealed an acutely ill patient with a distended, tender abdomen. A purpuric skin rash was noted on the extremities and trunk (Figure 1). Laboratory tests showed leukocytosis, proteinuria, and elevated creatinine. A skin biopsy revealed small-vessel neutrophilic vasculitis. Immunofluorescence was positive for multifocal IgA deposits along the walls of dermal vessels (Figure 2). High-dose prednisone and azathioprine were started. On hospital day 4, the patient developed slow junctional rhythm with hypotension requiring transvenous ventricular pacing. Serum cardiac troponin T was elevated. Sinus rhythm never recovered, and ectopic low atrial

rhythm predominated (Figure 3). The subsequent course was marked by worsening renal failure, noncardiogenic pulmonary edema, and respiratory failure. The patient died despite maximal supportive care. At autopsy, the heart showed confluent ecchymoses involving the entire right atrium (Figure 4). Multiple sections from the atrium, including the area of the sinoatrial node, showed neutrophilic myocarditis and diffuse small-vessel leukocytoclastic vasculitis with fibrinoid necrosis (white arrow) and interstitial hemorrhages (black arrow) (Figure 5). The cardiac chambers and great vessels were spared. Other findings included intestinal serosal hemorrhages, bronchial mucosal ecchymoses, and focal segmental glomerulonephritis positive for IgA deposits.



Figure 1. Confluent purpuric skin lesions involving toes and distal part of foot.

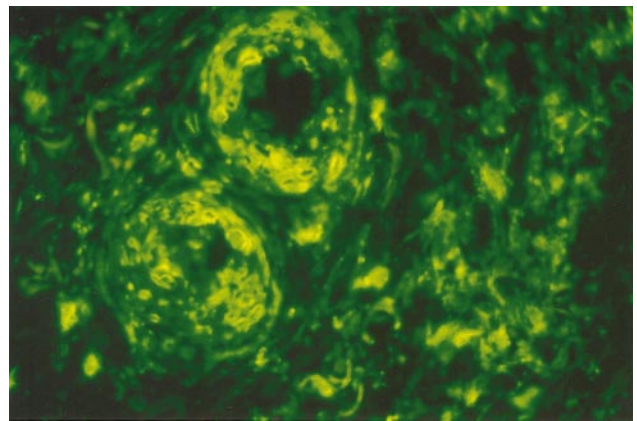


Figure 2. Immunofluorescence examination shows strong positivity for IgA deposits in walls of dermal small vessels.



Figure 3. Rhythm strip from lead II showing ectopic atrial rhythm.

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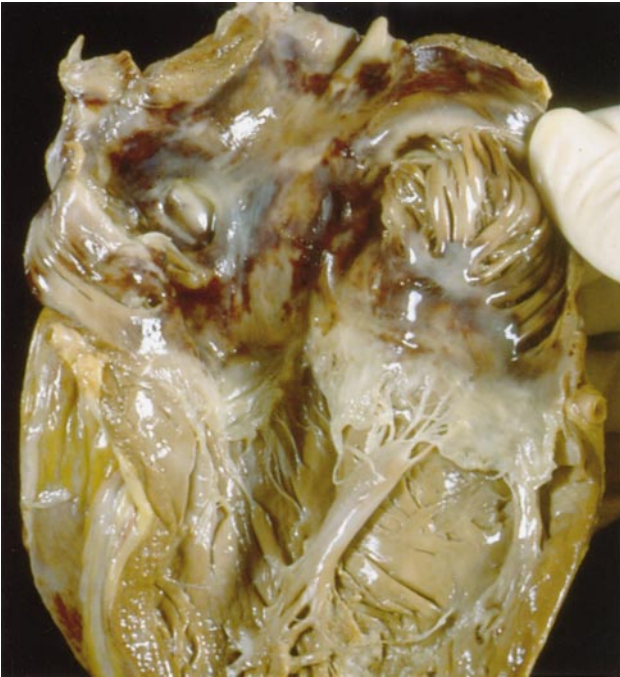


Figure 4. Exposed endocardial surface of right atrium and ventricle shows diffuse, confluent, subendocardial hemorrhages involving entire right atrium. Ecchymotic endocardium is sharply demarcated at tricuspid ring and at inlets of vena cava.

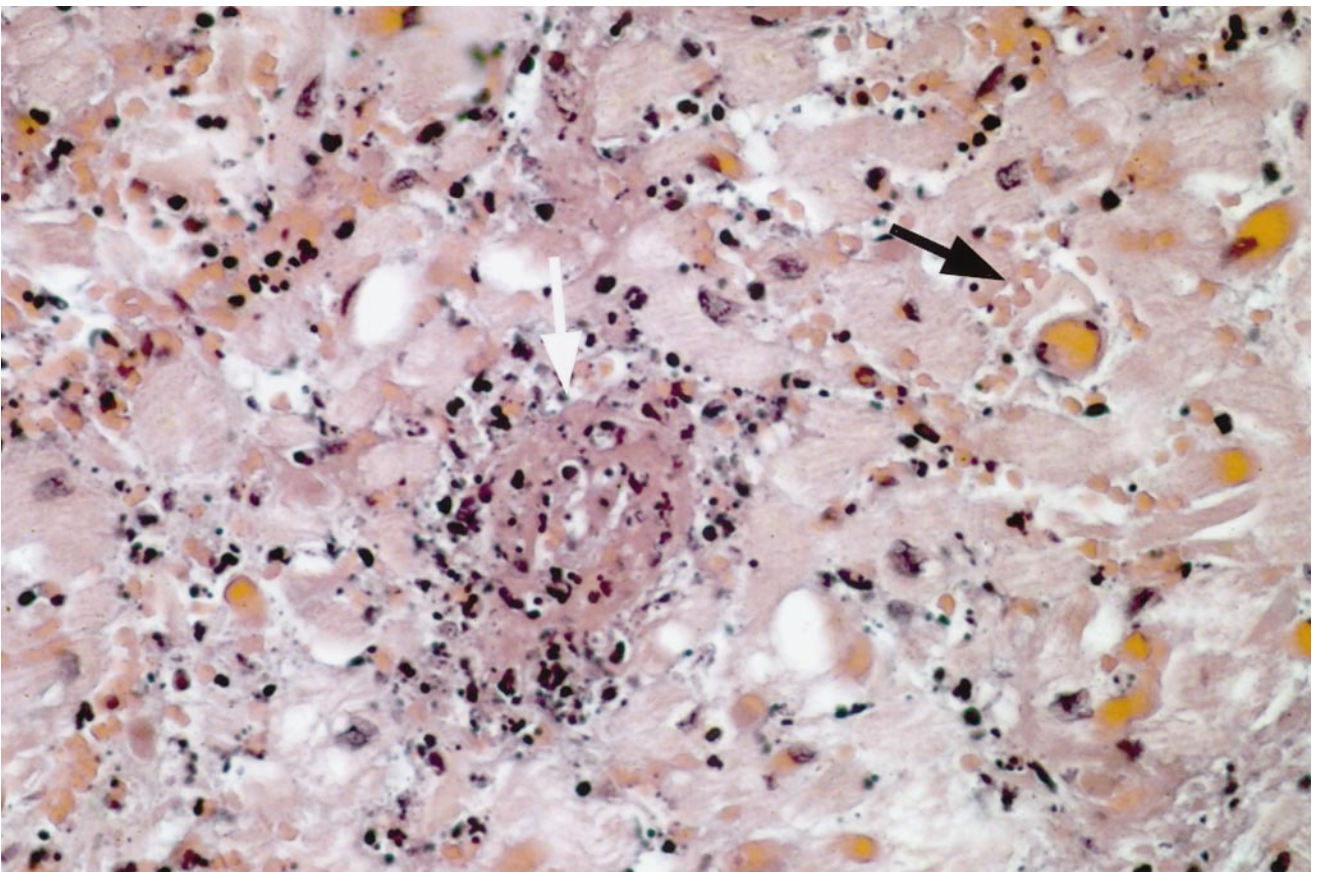


Figure 5. Hematoxylin-eosin–stained sections from right atrium showing necrotizing leukocytoclastic vasculitis of a small atrial vessel. Neutrophilic infiltrate and nuclear debris are seen in and around necrotic vessel (white arrow). Interstitial hemorrhages are present in background (black arrow).

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