Case 18

J.P. is a 63-year-old woman who was admitted to CPMC for evaluation of daily fevers to 102°F, rash, and diffuse arthralgias (joint pains). Physical examination was notable for BP 160/95 mm Hg and the presence of a tender, palpable, purpuric rash on her lower extremities (Fig. 1). She had no lymphadenopathy (enlarged lymph nodes) or joint swelling. Laboratory examination revealed Hct 34.5% (low), WBC 7.9 ($X10^9$/l) (normal). Serum creatinine was 2.0 mg/dl (elevated). Serum bilirubin, transaminases and alkaline phosphatase (liver function tests) were all slightly elevated. Her ESR (erythrocyte sedimentation rate) was 74 (very high; consistent with inflammation). A urinalysis revealed no protein, minimal microscopic hematuria (3-4 RBC per high power field) and occasional WBCs and numerous granular casts. A test for Hepatitis B surface antigen was positive. The following tests were negative or normal: Coombs, heterophile agglutinins, anti-streptolysin 0 (ASLO), latex fixation, ANA (anti-nuclear antibodies), cryoglobulins and SPEP (serum protein electrophoresis). All bacterial and fungal cultures were negative. Skin tests to tuberculin, fungal, mumps were all non-reactive. A chest X-ray was normal. During her hospitalization, she developed paresthesias (tingling) of the first three fingers on the right hand followed by intermittent burning pain in the same distribution. She also complained of sciatica which, in retrospect, she noted intermittently in the prior 6 months. A neurological examination was notable for muscular weakness in the L2, 3 distribution. EMG (electromyogram) of the median nerve and L2, L3 was consistent with a neuropathy. A biopsy of the skin is shown below (Fig. 2). She was treated with pulse intravenous methylprednisolone (15 mg/kg/dose IV X 3 days), followed by oral prednisone (1 mg/kg/day) and her fever resolved slowly. She remained afebrile but her neuropathy persisted and Cytoxan (cyclophosphamide) was added to her regimen with resultant improvement in her neurological symptoms.

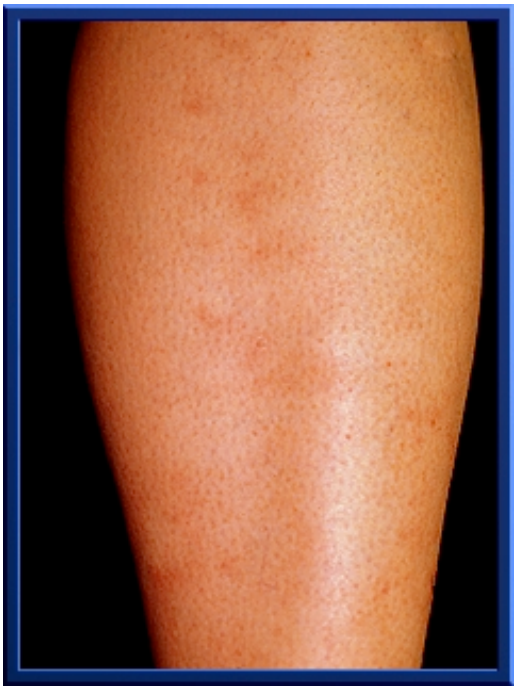

Fig. 1. Anterior tibial surface of J.P. demonstrates a slightly raised, tender, purpuric rash.

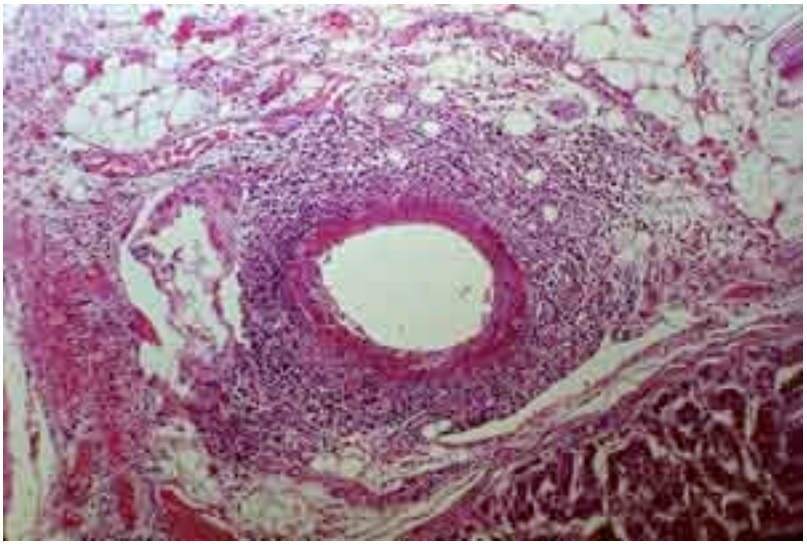

Fig. 2. H&E stain of a section from a skin biopsy reveals lymphocyte infiltration around a medium-sized artery.

Questions for Case 18

(1) What antigen was most likely involved in the etiology of this patient's immune vascular disease?

(2) What immunologic mechanisms caused the renal and neurologic abnormalities?

(3) What are the similarities and differences between the renal disease in this patient and in patients with SLE? If a renal angiogram were obtained in J.P., what would the results likely be?

(4) What other immunologic and clinical parameters distinguish J.P. from patients with SLE?

(5) The classification of vasculitis is complex. Mention three other types of vasculitis and describe how they are different from the current case.