Chronic lymphocytic leukemia presenting as purpura fulminans

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A 61-year- old man presented with ecchymoses over his knees and ear lobes. Physical examination on admission reveled an ill-looking man. Multiple areas of ecchymoses were seen over his knees and ear lobes. There were remarkable necrotic areas on the ecchymoses (Figure: 1,2). Other pertinent physical findings inluded an enlarged spleen palpable 5 cm below the left costal margin, multiple palpable lymph nodes on bilateral axillary, submandibular, inguinal, and servical regions. Admission laboratory values revealed a white blood cell count of 42,200/µL with 15% segmented neutrophils, 85% lymphocytes. The platelet count was 62,000/µL and the level of hemoglobin was 12.0 g/dL. The erythrocyte sedimentation rate was 90 mm/hour. Other laboratory data included the following: Antitrombin III %47 (N:70-125), d-dimer 700 ug/L (normal <340), fibrinogen 190 mg/dL, PT 11.6 sec., aPTT 29.8 sec. Total bilirubin, alkaline phosphatase, aspartate aminotransferase, alanine aminotransferase were all in normal limits. Lactate dehydrogenase was 725 U/L. Abdominal ultrasound revealed para-aortic and para-caval enlarged lymph nodes. There was no ascites. Bone marrow aspiration and biopsy revelaed diffuse infiltration with small lymphocytes. There was no fibrosis. Immmunophenotypic tests with flow cytometry revealed 52% (CD5 + CD20) positivity. Final diagnosis was B-cell CLL with purpura fulminans (PF). The patient developed large full-thickness necrotic injury of the skin. He was treated with heparin and plasma; however he died after 5 days following admission. Purpura fulminans is at least in part a cutaneous manifestation of the syndrome of disseminated intravascular coagulation (DIC), characterized by microvascular thrombosis in the dermis followed by perivascular hemorrhage, necrosis, and minimal inflammation.[1] Laboratory findings are consistent with DIC. Although the pathogenesis is not fully understood, the DIC in purpura fulminans appears to involve the skin selectively. The patient reported here had CLL and chronic consumption coagulopathy, followed by catastrophic skin necrosis as the presenting symptom of his leukemia. PF is usually associated with sepsis or previous infection, homozygous Protein C and Protein S deficien-

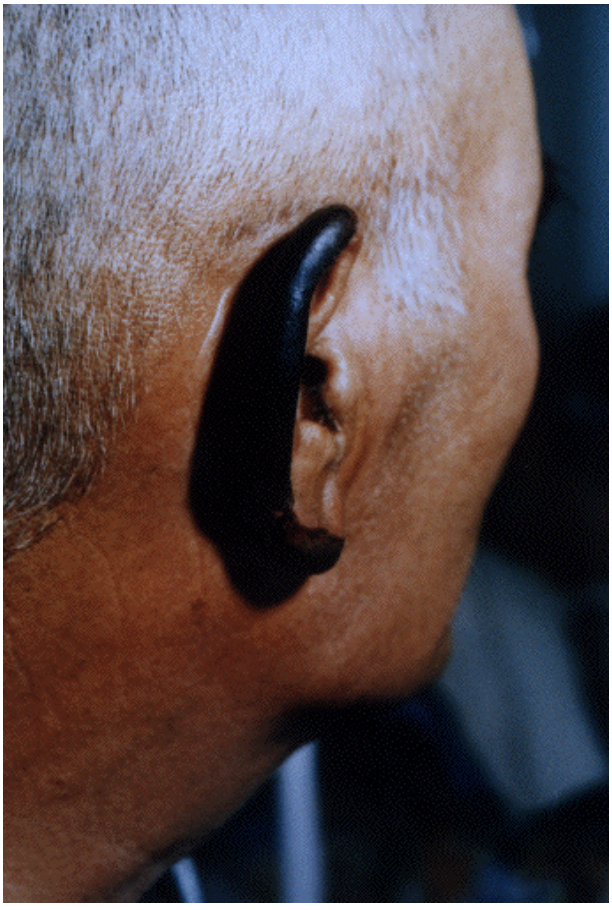

Figure 14. Remarkable necrosis on the right ear lobe.

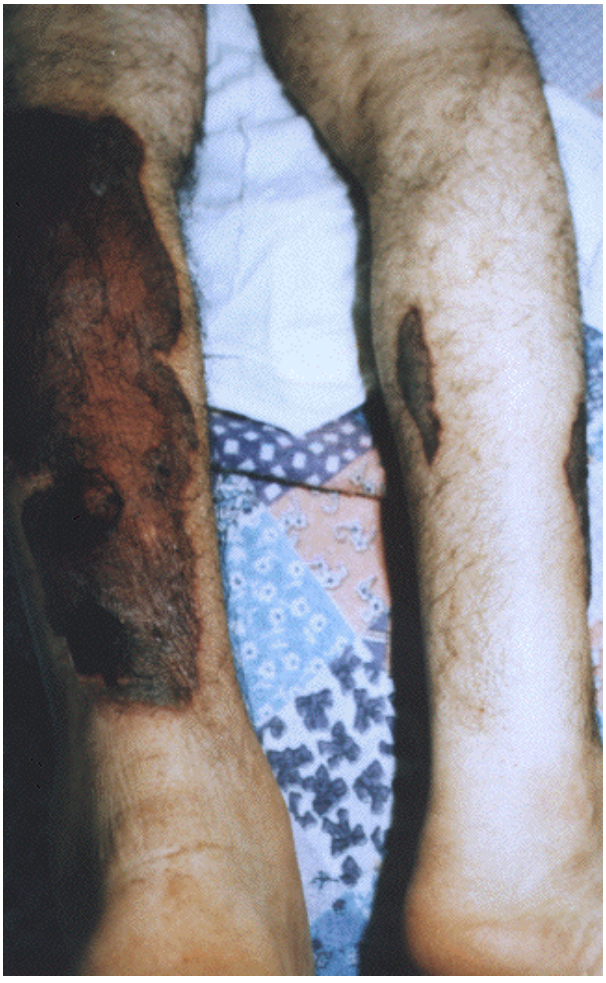

Figure 2. Multiple areas of ecchymoses over knees with necrotic areas.

cy.[2] Warfarin induced skin necrosis is a well known disease in patients with homozygous Protein C deficiency. Acute infectious purpura fulminans is a rapidly progressive syndrome of hemorrhagic skin necrosis associated with acute infection and disseminated intravascular coagulation.[3] Gram-negative organisms are the commonest cause of the acute infectious type, which is often associated with multi-organ failure. An idiopathic variety, often confined to the skin has also been reported. Idiopathic purpura fulminans produces rapidly progressive hemorrhagic necrosis of the skin with disseminated intravascular coagulation in individuals without known abnormalities of the protein C pathway or acute infections.[4] Idiopathic purpura fulminans usually occurs in young children and is frequently preceded by a preparatory viral or bacterial infection.[5] PF is also seen in patients with cholestasis and antiphospholipid syndrome. It has been reported in patients with malaria and ulcerative colitis.[6,7] There were no signs or symptoms of any infection in the case we report here. He had no deficinecy of PC or PS. He did not have any cholestasis, or antiphospholipid antibodies. As much as we know this is the first case with a presenting symptom of chronic lymphocytic leukemia.

K. Yamac,[1] T. Fen,[2] A. Erel[3]
[1]Department of Hematology, Faculty of Medicine, Gazi University;
[2]Department of Hematology, Oncology Hospital, Demetevler;
[3]Department of Dermatology, Faculty of Medicine, Gazi University, Besevler, Ankara, Turkey.

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