

A 73 year old woman with new onset, spontaneous purpura. What is the most likely diagnosis?

IMAGES IN CLINICAL MEDICINE

Amyloid Purpura



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Carmel Medical Center Haifa 34362, Israel ben@usha.org.il 73-YEAR-OLD WOMAN PRESENTED WITH NEW-ONSET PERIORBITAL PURpuric, nonblanching, nonpruritic lesions. The lesions appeared spontaneously and were not associated with any recent trauma. She did not take aspirin, nonsteroidal antiinflammatory drugs, or any other anticoagulant agents. On physical examination, no similar skin lesions were found elsewhere. In addition, laboratory studies revealed mild impairment of renal function and nephritic-range proteinuria. The blood count showed mild thrombocytopenia (platelet count, 80,000 per cubic millimeter), whereas the prothrombin time and partial-thromboplastin time were normal. Approximately 3 years earlier, the patient had received a diagnosis of multiple myeloma and acquired monoclonal immunoglobulin light-chain amyloidosis, for which she had not required any treatment. Therapy with cyclophosphamide and dexamethasone was initiated. The skin lesions waxed and waned repeatedly, with no apparent response to therapy. The patient died from pneumonia 6 months after the lesions first appeared.

Amyloid purpura appears in a minority of patients with amyloidosis. The purpura typically occurs above the nipple line and is often seen in the webbing of the neck, the face, and the eyelids. Factor X deficiency, resulting from the binding of factor X to amyloid fibrils, is thought to be one cause of the bleeding diathesis that may occur in patients with amyloidosis.

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