Recognizing Pinch Purpura as the First Manifestation of Light-chain Amyloidosis

Hafif Zincir Amiloidozun İlk Bulgu Olarak İzole Purpuradan Tanınması

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A 74-year-old female presented with a 6-month history of easy bruising as manifested by purpura after minor trauma to her face. Her physical examination was unremarkable except for the presence of pinch purpura scattered on her face (Figure 1). Laboratory tests showed leukocytes of 8100/µL, hemoglobin of 11.2 g/dL, platelets of 208,000/µL, prothrombin time of 11 s (normal range: 11.2-13.0 s), activated partial thromboplastin time of 25 s (normal range: 23.0-33.0 s), and erythrocyte sedimentation rate of 72 mm/h. Upon further workup, the presence of IgG lambda monoclonal gammopathy of the serum and lambda monoclonal light chain was found in urine immunofixation electrophoresis. Bone marrow biopsy revealed 7% lambda-restricted plasma cell infiltration, showing green birefringence with Congo red stain and vascular amyloid P deposition (Figure 2). There were no CRAB symptoms, organ dysfunction, or organomegaly. Echocardiography and pro-B-type natriuretic peptide results were normal. A diagnosis of amyloid light-chain (AL) amyloidosis initially presenting with purpura was made and a chemotherapy regimen of bortezomib and dexamethasone was started. Complete remission was achieved after six courses of chemotherapy and the purpuric lesions disappeared.

Figure 1. Purpura scattered on face (temporal region).

Cutaneous manifestations are reported in 30%-40% of AL amyloidosis cases [1]. The lesions usually reflect capillary

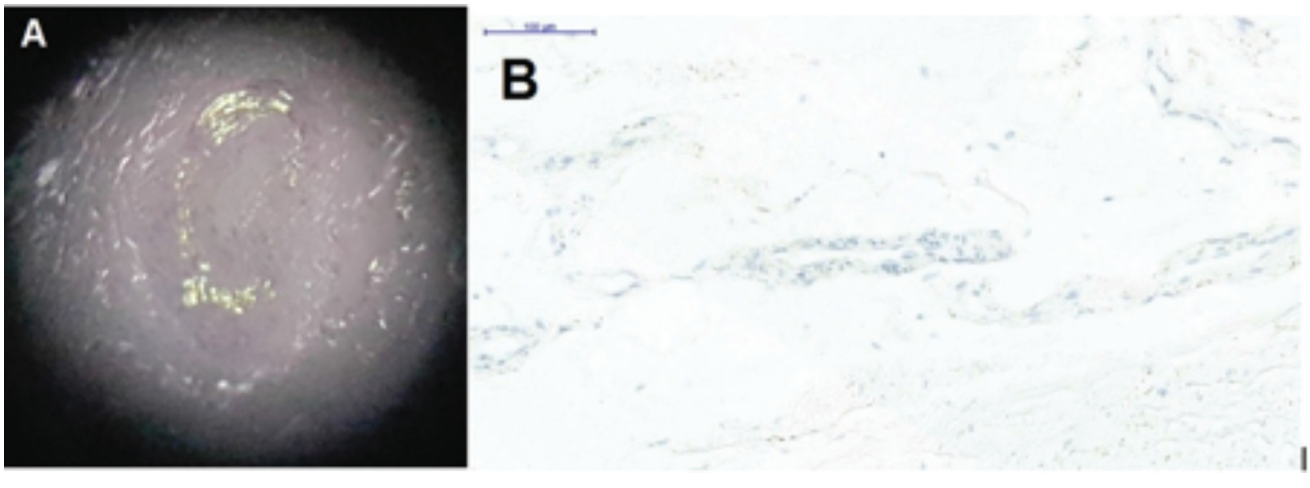

Figure 2. A) Microscopic section of the bone marrow stained with Congo red shows green birefringence under polarized light microscopy. B) Amyloid P with light microscopy.

infiltration and fragility with petechiae and purpura, characteristically affecting the eyelids, beard area, and upper chest [2]. Purpura as the initial manifestation leading to the diagnosis of AL amyloidosis is relatively rare [3,4]. Therefore, cutaneous findings are valuable in making a diagnosis of this challenging disorder since early diagnosis before development of organ failure is essential for improving the prognosis of AL amyloidosis patients.

Keywords: Amyloidosis, Purpura, Congo red

Anahtar Sözcükler: Amiloidoz, Purpura, Kongo kırmızısı

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Sclerosing Extramedullary Hematopoietic Tumor

Sklerozan Ekstramedüller Hematopoetik Tümör

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To the Editor,

Sclerosing extramedullary hematopoietic tumor (SEMHT) is a very rare disease. It was first described by Remstein et al. [1] in 2000 and few cases have been reported since then. It is associated with chronic myeloproliferative disorders. We describe a patient with SEMHT with bilateral renal involvement.

A 72-year-old man was admitted to the emergency clinic with dyspnea and abdominal pain. Physical examination revealed decreased breath sound over the right and left lower lung areas and severe edema in both lower extremities. He was clinically overhydrated. The rest of the physical examination was normal. Routine laboratory tests showed the following: serum levels of hemoglobin, 9.8 g/dL; white blood cell count, $31x10^9$/L; platelet count, $100x10^9$/L; blood urea nitrogen, 36 mg/dL; serum creatinine, 1.9 mg/dL; serum albumin, 2.2 g/dL. The peripheral blood smear was normal. A bone marrow biopsy was not performed. Abdominal ultrasound showed a perirenal hypoechoic mass compressing the bilateral kidneys (Figure 1a). Magnetic resonance imaging revealed heterogeneous renal capsular involvement with maximum thickness of 30 mm (Figure 1b). Renal Tru-cut biopsies were performed from both kidneys. Microscopically, both lesions showed myxoid and sclerotic stroma intermixed with large atypical cells (Figures 1c and 1d). Immunohistochemistry revealed positivity for factor-8 (Figure 1e) and CD41 in atypical cells (Figure 1f). Scattered mature myeloid cells were positive for myeloperoxidase, while CD34, CD117, S100, CD3, CD30, CD20, glycophorin, MDM2, keratin, EMA, desmin, and myogenin were all negative. The presence of CD41-positive atypical megakaryocytes within the tumor suggested the diagnosis of SEMHT. His previous history of primary myelofibrosis and splenectomy was learned after the histologic diagnosis of the renal tumor.

SEMHT is an uncommon lesion formerly known as fibrous hematopoietic tumor or myelosclerosis [1]. It is associated with