

LetterTJH-2018-0016.R2

Submitted: 9 January 2018

Accepted: 19 March 2018

Recognizing Pinch Purpura As The First Manifestation Of AL Amyloidosis

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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 in her face. Her physical examination was unremarkable except for the presence of pinch purpura scattered on her face (figure 1). Laboratory tests showed leukocytes 8100/ μ l, hemoglobin 11.2 g/dl, platelets 208000/ μ l, PT 11 sec (N:11.2–13.0 sec), aPTT 25 sec (N: 23.0–33.0 sec) and erythrocyte sedimentation rate 72 mm/h. On further workup, presence of IgG lambda monoclonal gammopathy on serum and lambda monoclonal light chain at urine immunofixation electrophoresis were found. 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 and organomegaly. Echocardiography and pro-BNP were normal. A diagnosis of AL amyloidosis initially presenting with purpura was made and chemotherapy regimen of bortezomib and dexamethasone was started. Complete remission was achieved after six courses of chemotherapy and purpuric lesions disappeared.

Cutaneous manifestations are reported in 30-40% of AL amyloidosis [1]. The lesions usually reflect capillary infiltration and fragility with petechiae and purpura, characteristically affecting the eyelids, beard area and upper chest [2]. Purpura as the initial manifestation

leading to 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 prognosis of AL amyloidosis patients.

References:

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

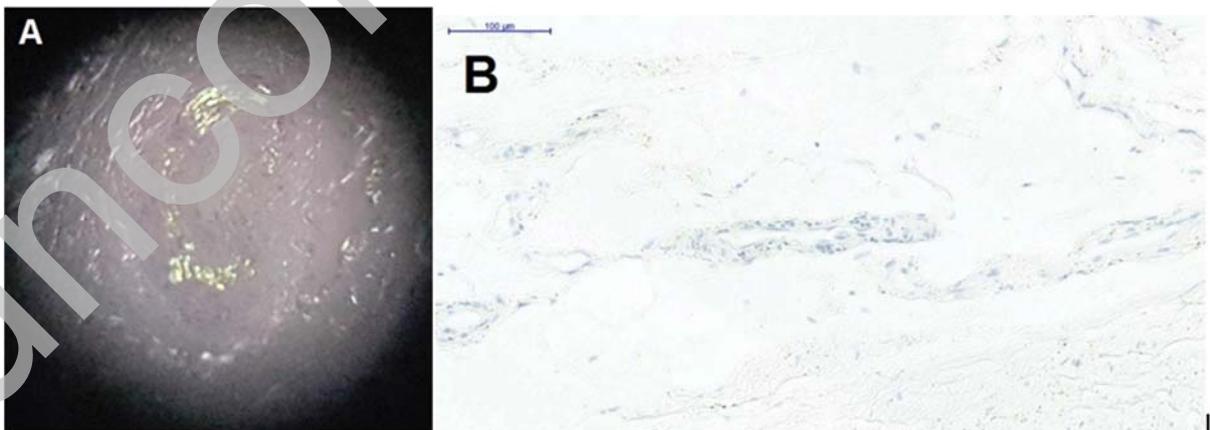


Figure 2: A. Microscopic section of the bone marrow stained with Congo red shows green birefringence under polarized light microscopy. B. Amyloid P on light microscopy