Pinch Purpura: A Clinical Clue for Primary Systemic Amyloidosis

Sir,

Primary systemic amyloidosis is a group of monoclonal plasma cell disorders, characterized by extracellular deposition of immunoglobulin light chain fibrils in multiple organs leading to progressive multi-organ dysfunction. It is a rare disease which usually occurs in elderly persons and has a poor prognosis.[1] We report a 73-year-old female with peri-orbital purpura as presenting feature of primary systemic amyloidosis.

A 73-year-old diabetic female presented with complaints of generalized weakness, arthralgia, and peri-orbital purpura of 3-month duration. The purpuric lesions developed spontaneously and were not associated with any recent trauma. There was no prior history of intake of drugs. On examination, there were extensive purpuric lesions on the neck, inframammary areas, anterior abdominal wall, inguinal area, and cubital fossa in addition to peri-orbital areas [Figure 1a]. No other cutaneous lesions were seen elsewhere over the body. Oral mucosa was congested with ruptured hemorrhagic bullae. The tongue was enlarged; fissured and hemorrhagic bullae were present on the surface with restricted movements. Teeth indentation marks were found around its lateral border [Figure 1b]. Systemic examination was normal. Her hemogram was normal except her hemoglobin was 8 g/dl. No abnormal cells were found in the peripheral smear. Bleeding time, clotting time, prothrombin time, activated partial thromboplastin time, and International normalized ratio (INR) were normal. Bone marrow aspiration from the iliac crest revealed increased number of plasma cells (average: 13%). All relevant biochemical and serological tests were normal. Urine 24 h protein was 132 mg/day. Urinary Bence Jones proteins were absent. High-resolution serum protein electrophoresis revealed the presence of monoclonal gammopathy M spike in the gamma globulin region. Serum immunofixation electrophoresis revealed the presence of monoclonal gammopathy (M spike) as IgG and Lambda. Skin biopsy from the purpuric lesions showed eosinophilic amorphous homogenous deposits in the dermis and surrounding thin blood vessels in the reticular dermis; extensive extravasated Red blood cells (RBCs) were present [Figure 2]. Congo red staining showed characteristic apple green birefringence on polarized microscopy. Abdominal fat pad aspiration showed apple green birefringence on polarized microscopy [Figure 3]. Chest radiograph, skeletal survey of the body, ultrasound, and Computed tomography (CT) of the abdomen were normal. Electrocardiogram (ECG) and 2D Echocardiography (2D-ECHO) showed no significant abnormalities.

Based on the clinical features, histopathology, and biochemical findings, a diagnosis of primary systemic amyloidosis of Amyloid light-chain (AL) type was made.

Skin manifestations are seen in almost all three groups of primary systemic amyloidosis, and its involvement is 30%–40% of cases. Cutaneous manifestations comprise of petechiae or hemorrhages, peri-orbital purpura, waxy papules, nodules, plaques, bullous lesions, mucocutaneous infiltrates, macroglossia, and nail dystrophy.[1] Purpura, petechiae, and ecchymoses are the common, and these are caused with or without trauma. This purpura is called as pinch purpura.[2] The purpura typically occurs above the nipple line and is often seen in the webbing of the neck, peri-orbital areas, and the eyelids. Amyloid deposition in the skin and blood vessel wall causes capillary fragility and eventually causing intracutaneous micro- and macrohemorrhages.[3] Factor X deficiency resulting from the binding of factor X to amyloid fibrils, is thought to be another cause of bleeding diathesis.[4]

In this patient, the key presenting feature was peri-orbital purpura and scattered nontraumatic ecchymoses, without systemic involvement with normal hematological findings. This feature (pinch purpura) led us to consider the diagnosis of primary systemic amyloidosis (AL type). In majority of the

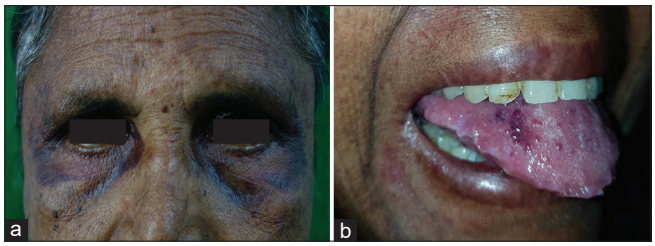

Figure 1: (a) Peri-orbital purpura (b) Hemorrhagic blisters and teeth indentation marks over the tongue

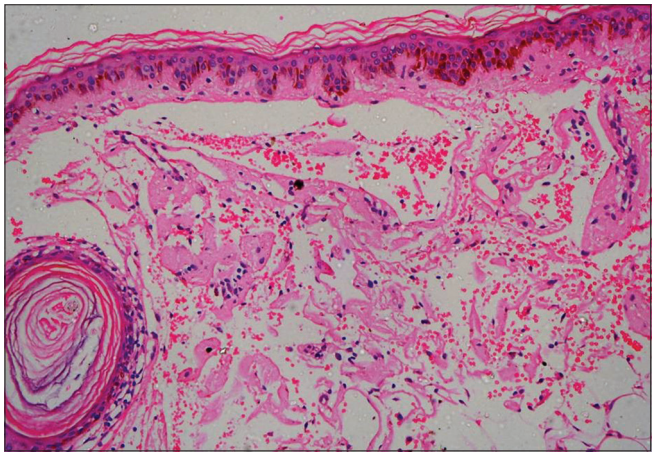

Figure 2: (H and E × 400) Normal epidermis with thin-walled blood vessels, seen with surrounding eosinophilic amorphous homogenous deposits. The deposits are also present in the dermis. The reticular dermis shows extensive extravasated RBCs. There is focal homogenization of the reticular dermis

published cases[5-7] also, pinch purpura is the early presenting manifestation. Recently, Zoe *et al*. reported an interesting case of pinch purpura with systemic amyloidosis, multiple myeloma, and subclinical cardiomyopathy.[8] Another case of peri-orbital inframammary purpura with left ventricular hypertrophy without any clinical history of hypertension was also reported.[3] The heart is the second most commonly affected organ next to renal involvement, which accounts for 75% of deaths in AL amyloid patients. Therefore, pinch purpura can be considered as an early cutaneous marker of this systemic disease.

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Conflicts of interest

There are no conflicts of interest.

Ashwini R. Mahesh, T. Satyaprakash[1], Deepak Joshi, Gandikota Raghurama Rao, Kollipara Haritha[2], A. Prasad Chowdary

Department of DVL, GSL Medical College, [1]Department of Pathology, GSL Medical College, Rajahmundry, [2]Department of DVL, GITAM Medical College, Visakhapatnam, Andhra Pradesh, India

Address for correspondence: Dr. Gandikota Raghurama Rao, Department of DVL, GSL Medical College, Rajanagaram, Rajahmundry - 533 296, Andhra Pradesh, India.
E-mail: graghuramarao@gmail.com

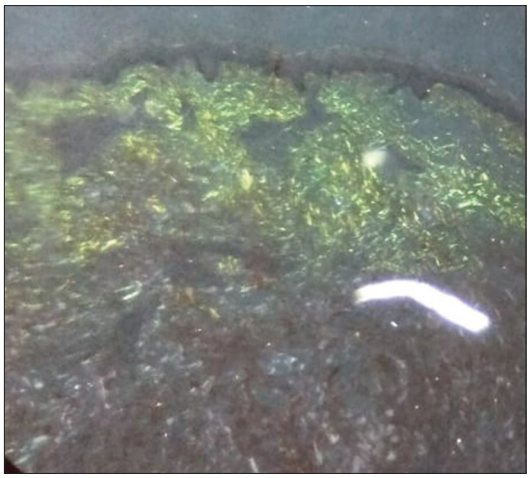

Figure 3: (×100) Congo red shows apple green birefringence under polarized light

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