Systemic Cutaneous Amyloidosis

A 55 years gardener presented with multiple pinch purpura and ecchymotic plaques predominantly over the periorbital region (Raccoon sign), neck, upper arms, forearms and penis. He also had extensive itchy, erythematous, waxy, infiltrated plaques, vesiculobullous lesions and hemorrhagic crusts over the neck, back and upper limbs with flexural predilection (Fig. 1). A few hemorrhagic vesicles and erosions were also present in the oral mucosa (Fig. 2). There were no other clinical findings of systemic amyloidosis in the form of macroglossia, renal dysfunction, heart blocks, congestive heart failure, autonomic dysfunction, peripheral neuropathy or myopathy. Abnormal laboratory investigations included anaemia (Hb - 9.5 gm%), elevated ESR (78 mm in 1st hour), reversal of serum albumin globulin ratio and proteinuria (1.2 gm in 24 hours). Skin biopsy showed presence of amorphous eosinophilic deposit both in papillary dermis and perivascular region. On congo red stain it was positive for amyloid. Presence of increased number of plasma cells (40%) in the bone marrow, a ‘M’ band on serum electrophoresis and Bence Jones proteins in the urine confirmed the presence of underlying multiple myeloma. Patient was initiated on vincristine, doxorubicin and dexamethasone (VAD) regimen for multiple myeloma.

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