40 year old female presented with non-healing ulcers on both the legs since 5 years. She was being treated by general surgeon with broad spectrum antibiotic coverage and repeated debridements. Plastic surgeon was also consulted. On examination the ulcers were around the malleolus, painful, irregular in shape with erythematous margins and the floor was covered with necrotic slough. The surrounding skin showed hyperpigmentation. The patient was investigated for vasculitic pathology and also the possibility of thrombophilic state was considered. Her baseline investigations were normal and the workup for ANA, p-ANCA, c-ANCA, protein C, protein S, anti-cardiolipin antibody was negative. To resolve the diagnostic dilemma, skin biopsy was undertaken which was consistent with livedoid vasculopathy. The patient was started on aspirin and dipyridamole. Marked improvement was observed in the ulcers with resolution of pain. The patient is currently on aspirin and dipyridamole.

Livedoid vasculopathy (also called atrophie blanche) is a form of occlusive vasculopathy. It is most common in young to middle aged females either as idiopathic or secondary syndrome.\(^1\) It can be associated with chronic venous hypertension but those ulcers are not painful or with livido reticularis.

It is a thrombotic vasculopathy where occlusion of small dermal vessels with fibrin thrombi is the primary event. Multiple pathophysiological abnormalities have been implicated including platelet activation, hyperhomocysteinemia, altered fibrinolysis, antiphospholipid antibodies and factor V Leiden.\(^2\) Microscopic examination of the skin biopsies reveal hyalinization and thrombosis of blood vessels. Inflammatory infiltrate is scanty unlike vasculitis syndromes. Hence immunosuppressive therapy is not as effective and controlling the hypercoagulable state is the aim of treatment.\(^3\)

Ulcers are seen usually on the lower limbs; around the ankles and dorsum of feet; rarely is the upper extremity involved. Telangiectatic and purpuric papules progress to painful ulcers which heal with stellate, white scars.

Antiplatelet antithrombotic therapies form the mainstay of the treatment.\(^4\) Aspirin, dipyridamole, oral pentoxyphylline may be used. Other treatment strategies include dapsone, PUVA therapy, intravenous immunoglobulin, danazol, nicotinic acid and hyperbaric oxygen therapy.

References

4. Weedon D. The Vasculopathic Reaction Pattern. Weedon’s Skin Pathology 3rd ed; Section 2: 198.