VOLUME - 9, ISSUE - 8, August - 2020 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Dermatology

LIVEDOID VASCULOPATHY: AN UNCOMMON CAUSE OF LEG ULCER

Sanket Vashist	Department of Pediatrics, Zonal Hospital, Dharamshala, Himachal Pradesh.
Vikram K Mahajan	Department of Pediatrics, Zonal Hospital, Dharamshala, Himachal Pradesh.
K S Mehta	Department of Pediatrics, Zonal Hospital, Dharamshala, Himachal Pradesh.
P S Chauhan	Department of Pediatrics, Zonal Hospital, Dharamshala, Himachal Pradesh.
Vikas Sharma,	Department of Pediatrics, Zonal Hospital, Dharamshala, Himachal Pradesh.
Aashutosh Sharma*	Department of Pediatrics, Zonal Hospital, Dharamshala, Himachal Pradesh.*CorrespondingAuthor

KEYWORDS :

INTRODUCTION

Livedoid vasculopathy also referred to as Livedo reticularis with summer ulcerations[1] or Segmental hyalinizing vasculitis[2] is a rare cutaneous disease presenting with classic triad of livedo reticularis which refers to net like pattern of reddish blue skin discoloration, leg ulcers, and porcelain white atrophic scars which may have peripheral telangiectasias referred to as atrophie blanche. The chronic recurrent course is marked by episodic exacerbations. The pathogenesis is unclear and mostly attributed to dysregulated coagulation and Factor V Leiden.

Case report

This 42-yr-old man with left leg ulcer was referred for evaluation of persistent skin lesions. Historically, he had retiform erythema over fingers exacerbating during winters since childhood. He had developed retiform atrophied plaques and pigmentation over trunk, and upper extremities, and alopecia over the years.

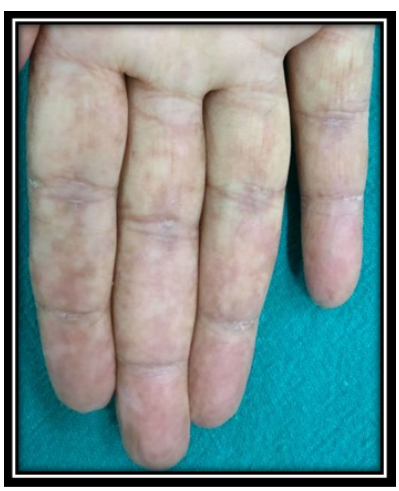

Figure 1: Retiform finger erythema.

He had recurrent painful ulcerations over lower legs healing with atrophic scars severely affecting quality of life. He was being treated for venous ulcer in last 7-8 yrs. Ligation of below knee perforator left leg was done recently due to a non-healing (?venous) ulcer present for 5 months.

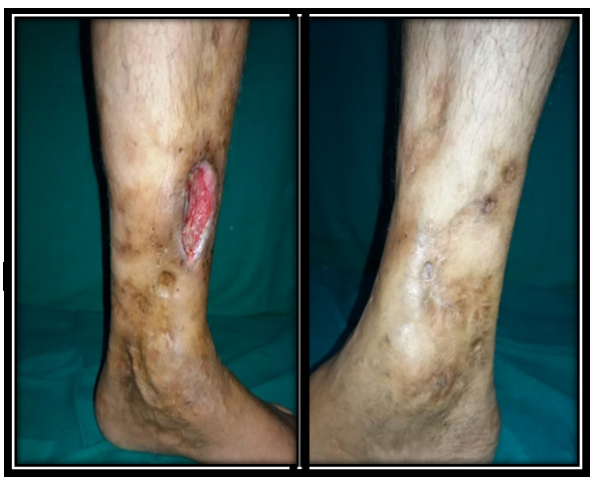

Figure 2, 3: A large ulcer with multiple atrophic scars.

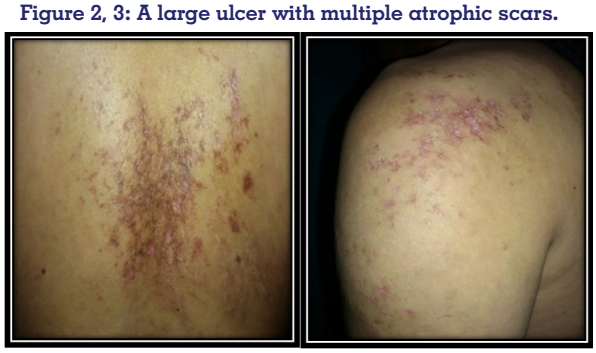

Figure 4, 5: Atrophic retiform plaques over back and shoulders.

Baseline investigations, coagulogram, viral markers, anticardiolipin Ab, D-dimer levels, RA factor were normal. ANCA, APLA, Protein C, S, Factor V Leiden, homocysteine levels were not done for want of affordability. ANA was mixed pattern 3+, titre 1:320.Doppler studies showed incompetent right Sapheno-Femoral junction but no incompetent perforators on affected leg were detected. Systemic examination was normal. Histopathology showed areas of epidermal necrosis and swollen endothelial cells (vasculopathy) .No vasculitis was seen.

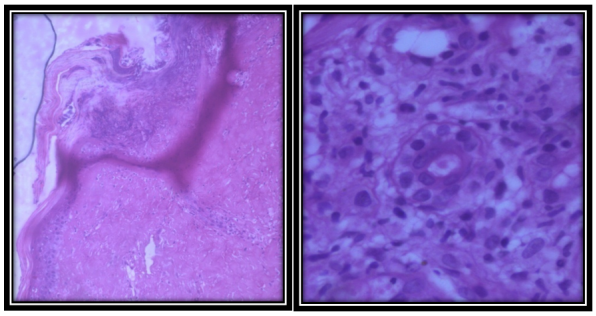

Figure 6 (a, b): Histopathology shows; (a) areas of epidermal necrosis, (b) swollen endothelial cells (vasculopathy). No vasculitis was seen.

Diagnosis of Livedoid Vasculopathy was made on basis of clinical history, characteristic morphology and corroborating histology. He was given tablet Nifedipine 10mg twice daily, tablet Clopidogrel 75mg daily, tablet Asprin 75mg thrice a day, tablet Folic Acid 5mg daily and sterile dressings for the ulcer. Marked improvement in pain and healing of ulcer occurred within 2-3 days. Patient is currently under follow up.

DISCUSSION

Leg ulcers can have multiple etiologies and Livedoid vasculopathy is one such relatively rare cause which poses diagnostic challenges as it may be idiopathic or occur as a secondary phenomenon. It may be seen in patients with or without identifiable coagulation abnormalities. A wide array of differential diagnosis but not limited to include peripheral articular disease, peripheral vascular disease, vasculitis especially poly arteritis nodosa, systemic lupus erythematosus, scleroderma, microscopic polyangitis, granulomatous vasculitis and cryoglobulinemias. Treatment includes anti-platelet drugs, systemic anticoagulants and fibrinolytic therapy. Colchicine, Hydroxychloroquine, vasodilators, Intravenous immunoglobulins, folic acid, immunosuppressive drugs, hyperbaric oxygen therapy, supportive measures like bed rest , limb elevation, compression stockings and smoking cessation may be helpful.[3,4] Large ulcers may require additional excision and grafting.

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