



BULLOUS LEUKOCYTOCLASTIC VASCULITIS – AN UNUSUAL PRESENTATION

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ABSTRACT

Leukocytoclastic vasculitis (LCV) also known as ‘Hypersensitivity vasculitis’ or ‘angitiis’ used to denote small vessel vasculitis. LCV classically presents as palpable purpura, but has polymorphic presentations clinically which can be misleading in the diagnosis. Bullous LCV is relatively a rare presentation. We report a case of a 52-year-old man who presented with hemorrhagic blisters and ulcers over lower limbs for one-month duration. On cutaneous examination, there were multiple hemorrhagic bullae and shallow ulcers over both legs. Histological examination confirmed the diagnosis. Other lesions of LCV are often cited in literature, but very few cases of bullous LCV have been reported so far. Hence we present this case of ‘Bullous LCV’ for its rarity.

Keywords: Bullous LCV, Vasculitis.

INTRODUCTION :

Leukocytoclastic vasculitis is an immune complex mediated cutaneous small vessel vasculitis, mainly affecting the cutaneous post-capillary venules and is characterised by vascular damage and inflammatory infiltration. LCV is idiopathic in 50% of cases and the secondary causes include drugs, infections and malignancies. (1) The usual clinical manifestation is palpable purpura but sometimes it may progress to a wide array of lesions like nodules, plaques, vesicles, bullae or pustules with secondary ulceration and necrosis. (2) Bullous LCV is relatively rare manifestation of LCV which manifests when there is pronounced edema in dermis.

CASE REPORT :

A 52 -year-old businessman presented with one month history of multiple reddish blisters over both legs, which erupted simultaneously over the dorsum of both feet and progressed upto mid tibial level in a span of 1 week which was associated with occasional pruritus. Patient did not have systemic symptoms and denied over the counter drug intake in the recent past. Patient is a known diabetic, hypertensive with CAD for which he is on insulin, metformin, atenolol, aspirin, statins, furosemide for 3 years. Physical examination revealed multiple, hemorrhagic bullae of size 2 to 5cms, distributed bilaterally over dorsum of feet, medial and lateral ankle and lower legs (figure 1). Few bullae had ruptured already and the intact ones on rupture using sterile needle discharged hemorrhagic non foul smelling fluid. The skin lesions eventually ulcerated in few days. The ulcers had well defined margins, sloping edges, and hemorrhagic crusts over the floor. Scalp, nail, mucosa were normal.

Laboratory evaluation revealed raised ESR of 60 mm at one hour and random blood sugar of 145mg/dl. Complete blood count, urea, creatinine, electrolytes, liver function tests, lipid profile were within normal limits. ANA, p-ANCA, CRP, ASO Anti HBV and Anti HCV Antibodies were negative. Tzanck smear, Gram staining and culture of the bullous fluid were unremarkable. Skin biopsy from a bullous lesion over right leg revealed sub epidermal blister with a dermal perivascular infiltrate of neutrophils and their nuclear dusts (degenerated neutrophils) with fibrinoid necrosis of blood vessels (figure 2). Based on the history, clinical findings and histopathology, patient was diagnosed as a case of ‘Bullous LCV’ and was started on azathioprine 50 mg once daily along with calcium dobesilate, analgesics, antihistamines and glycemic control measures. The ulcers were treated with topical zinc oxide cream and saline soaks. The lesions responded well in 6 weeks and there were no new eruptions (figure 3).

DISCUSSION :

Leukocytoclastic vasculitis is the commonest form of cutaneous vasculitis where the term leukocytoclasia refers to the histological pattern. It is a type 3 hypersensitivity reaction in which immune complex deposition in post capillary venule results in a destructive inflammatory vasculitis. The resultant complement activation leads to influx of inflammatory cells like neutrophils and eosinophils which induce mast cell degranulation and neutrophil chemotaxis. This in turn release proteolytic enzymes and free oxygen radicals that damage vessel walls. (3,4) Increased expression of adhesion molecules also plays a role in the pathogenesis. Histopathology shows leukocytoclasia, perivascular inflammatory infiltrate, fibrinoid necrosis and fibrin deposition in and around affected vessels. Sub epidermal bullae may form due to marked edema in

superficial dermis resulting in bullous LCV.(5)All these feature were seen in our patient. The differential diagnosis include arthropod bites, bullous drug eruptions, erythema multiforme, Sweet's syndrome, cryoglobulinemic vasculitis and bullous pemphigoid. Skin biopsy is confirmative and the ideal time to perform is within 18-36 hours after the onset of the lesions. Direct immunofluorescence shows IgM, C3 and fibrin deposits in the vessel wall.ANCA is rarely (<5%) positive in LCV.Serum complement are lowered. (6)For mild cases Analgesics are sufficient and for symptomatic cases corticosteroids (e.g. up to 1-2 mg/kg of prednisolone), colchicine (0.6 mg twice daily), dapsone, azathioprine, cyclophosphamide, methotrexate, cyclosporine or, IVIg has been tried. (7)

CONCLUSION :

This case is being reported because of the rarity and also to stress the importance of keeping a high index of clinical suspicion in context of bullous dermatosis for early diagnosis and prompt management.



Figure 1: Hemorrhagic bullae over left leg



Figure2: healing ulcers after treatment

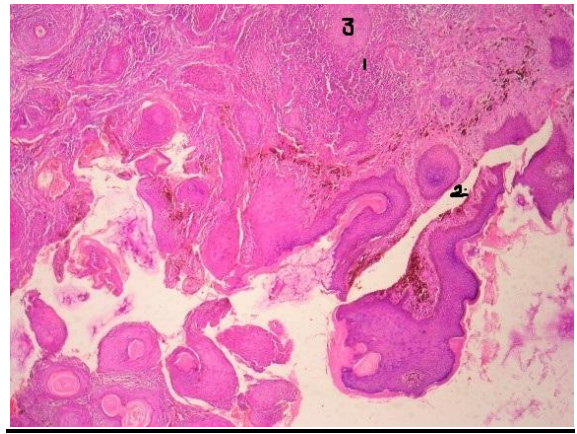


Figure 3: HPE(10x) showing 1.perivascular neutrophilic infiltrate, 2.subepidermal bulla and 3.fibrinoid degeneration of blood vessels.

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