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Unusual presentation of cutaneous small vessel vasculitis

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Introduction: Cutaneous small vessel vasculitis (LCVV) is a disease characterized by inflammation of small vessels with characteristic clinical findings of petechiae and palpable purpura[1]. We are presenting a case of 48 years old female, presenting with large annular ulcero-necrotic lesions without any features of small vessel vasculitis like palpable purpura, hemorrhagic vesicles or urticarial plaques.

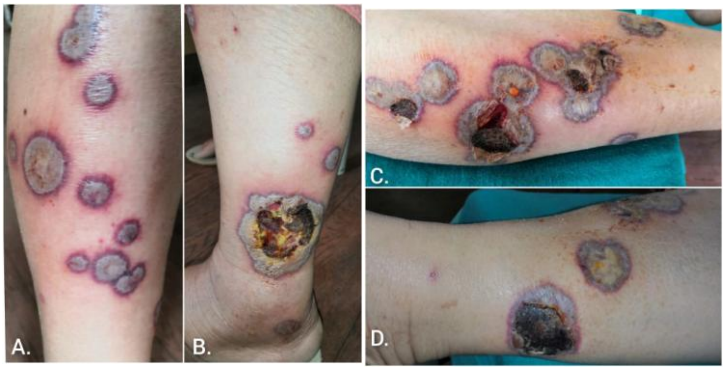
Case report: A 48 years old female patient preseted in dermatology OPD with multiple painful, ulcero-necrotic plaques and a large ulcer over bilateral lower limbs for last fifteen days. There was no history of fever, drug intake, local application of medicines or indeginous preparations prior to eruption of these lesions. The eruption was associated with generalized malaise and loss of appetite and edema of lower legs. Patient was known diabetic for last 5 years and was on metformin, there was no history of hypertention, thyroid disease, ATT intake in past or any other systemic illness.

On clinical examination, patient had multiple, annular plaques present over bilateral lower legs, discrete to coalescing at places with surface showing ulceration and variable degree of necrosis, with surrounding skin showing erythema. A large ulcer with yellowish granulation tissue and necrosed skin was present over left leg above lateral malleolus. There was pitting edema present over both legs.

General physical and systemic examination was within normal limits. The routine hematological and biochemical investigations were within normal limits except for eosinophilia and increased random blood sugar (158 mg/dl). ASO was negative and CRP was within normal limits. ANA and serology for HIV were both negative. No abnormality was detected in chest radiograph.

On the basis of morphology of lesions, differentials of medium vessel vasculitis, livedoid vasculopathy and dermatitis artefacta were considered and a punch biopsy was sent using 5mm biopsy punch. However the biopsy revealed features of leucocytoclastic vasculitis i.e segmental inflammation, fibrinoid necrosis and extravasation of RBC's along with karyorrhexis of nuclei.

Patient was started on oral deflazacort, prophylactic topical (fusidic acid) and oral antibiotics (co-amoxiclav) and was reviewed after seven days. There was significant improvement in patient's general condition and lesions were healing. No new lesions had erupted since the therapy had started.



Clinical photographs of patient showing multiple discrete to coalescent annular plaques with surrounding erythema(A), ulcer with necrotic slough and yellow granulation tissue(B), ulcero-necrotic plaques showing varying degree of necrosis(C and D).

Discussion: Leukocytoclastic vasculitis is an inflammatory vascular disease characterized by prominent involvement of the skin, the existence of a precipitating event (usually a drug), and by the infiltration of the small blood vessels with polymorphonuclear leukocytes and the presence of leukocytoclasia[2].

In a significant percentage of patients with CSVV, no precipitating event can be found; and secondly, a clinical and pathologic picture similar to that of CSVV can be observed as a manifestation of several processes such as infections, malignancies, other vasculitic disorders, and in most connective tissue diseases[3,4]. Cutaneous vasculitis is predominantly due to infections in 22%, drugs in 20%, connective tissue disorders in 12%, Henoch Schonlein purpura (HSP) in 10% and <5% each due to malignancy, primary systemic vasculitis or systemic inflammatory disease[5,6].

Yet, exact etiology may not be established in spite of exhaustive work up in many case scenarios and idiopathic nature predominates. The mean age of onset is 7 years in children, 47 years in adults, vasuclitis being, commoner in adults than children[7].

Clinical manifestations of CSVV include purpura, urticaria, vesiculobullous lesions and targetoid lesions[8]. Whereas in medium vessel vasculitis subcutaneous nodules, livedo reticularis, ulcers, infarcts, digital pitted scars and gangrene occur[6,7].

Investigations to be carried out in a suspected case of vasculitis include, hemogram, RFT, LFT, urine microscopy and proteins, ANCA, ANA, mauntoux, chest radiograph and skin biopsy. Treatment includes removal of triggering factors, if identified, supportive measures and systemic treatment in form of oral corticosteroids, colchicine and cytotoxic agents including azathioprine, ciclosporin andmethotrexate

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest:

There are no conflicts of interest.

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