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CASE REPORT

A Rare Case Report of Primary Skin-Limited Leucocytoclastic Vasculitis: A Diagnostic and Therapeutic Challenge

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ABSTRACT

Leucocytoclastic vasculitis (LCV) is a histopathologic term referring to small vessel vasculitis characterized by the infiltration of neutrophils and immune complex deposition. Clinically, LCV presents with palpable purpura, often involving the lower extremities, and may be associated with systemic involvement. We hereby report a rare case of LCV in a 56 year female, highlighting clinical presentation and management strategies for the subset of LCV that remains confined to the skin without systemic manifestations posing diagnostic challenges.

KEYWORDS:

• Leucocytoclastic vasculitis • Small vessel vasculitis • Immune complex • Palpablepurpura

INTRODUCTION

Leucocytoclastic vasculitis (LCV) is a small-vessel vasculitis predominantly affecting postcapillary venules. The condition is commonly immune-mediated and can be triggered by infections, drugs, autoimmune disorders, and malignancies. The clinical spectrum ranges from isolated cutaneous involvement to systemic vasculitis affecting the kidneys, gastrointestinal tract, and nervous system.[1] The exact incidence of primary skin-limited LCV remains unclear due to its variable presentation and self-limiting nature. Studies estimate that approximately 10-30% of LCV cases are confined to the skin without systemic involvement.[2] However, under reporting and misdiagnosis contribute to

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challenges in determining the true prevalence.

This case report presents a unique presentation of LCV in a 56 year old female with an idiopathic underlying trigger.

CASE PRESENTATION

A 56-year-old female known case of type 2 diabetes and hypertension presented with a complain of 2 month history of multiple joint pain all over the body initially in bilateral knee and then slowly progressing to involve upper limb joints starting from shoulder and elbow Joint. There was no history of morning stiffness along with complain of rash which was non blanchable since 4 days, sudden in onset that started from the trunk (Figure 1) and gradually progressed to lower limbs (Figure 2) over 2 to 3 days, not associated with pruritis or burning sensation, no similar complaint of rash in the past. The patient denied recent infections, native medication changes, or autoimmune symptoms. Initial laboratory tests showed normal blood counts, elevated C-reactive protein, mildly elevated AST, ALT and normal renal function.

Autoimmune serology, including RA, ANA and ANCA, were all negative. Two skin punch biopsy were taken from abdomen and sent for histopathology to department of pathology.

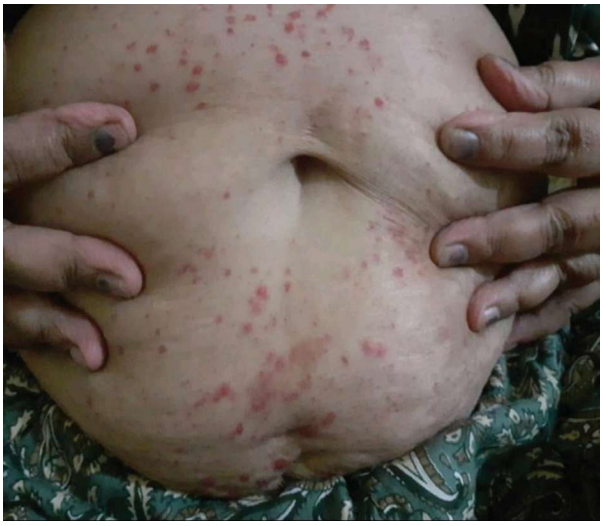

Figure 1: Clinical image of non-blanching petechial rash on abdomen

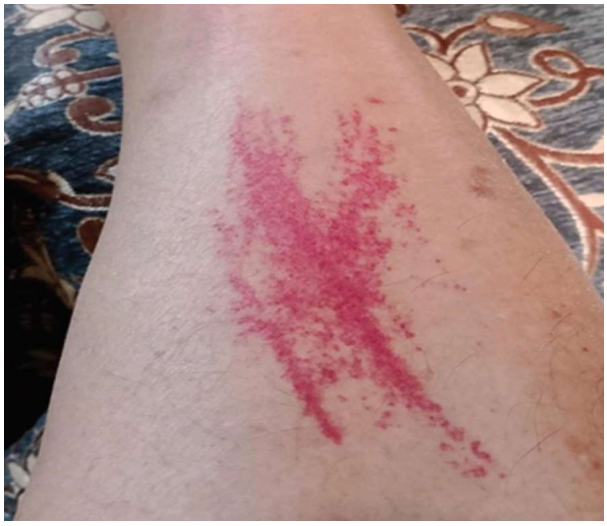

Figure 2: Clinical presentation of non-blanching petechial rash on lower extremity

Grossly: We received two grey white partially skin covered tiny tissue bits altogether measuring 0.8x0.4x0.2cm.

On Microscopy: Section examined from skin punch biopsy show keratinized stratified squamous epithelium (Figure 3a) and mild basal layer pigment incontinence (Figure 3b).

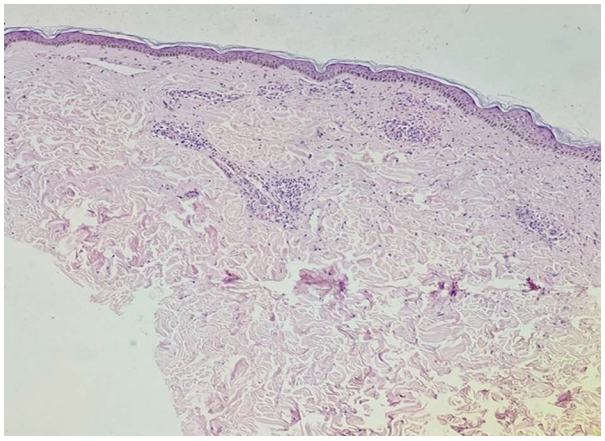

Figure 3a: Photomicrograph of skin punch biopsy showing keratinized stratified squamous Epithelium. Dermis showing perivascular inflammatory infiltrate. (Scanner, H&E)

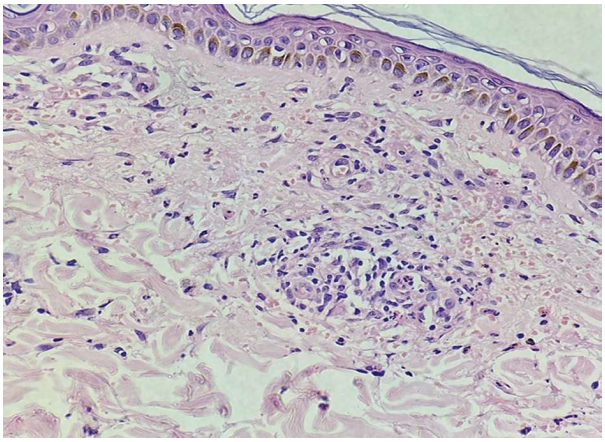

Figure 3b: Photomicrograph of skin punch biopsy showing keratinized stratified squamous epithelium with mild basal layer pigment incontinence. (10x, H&E)

Both upper and deeper dermis revealed few dilated blood vessels showing mild mixedperivascular infiltration comprising of polymorph, lymphocytes, and few eosinophils alongwith cellular debris. (Figure 3c) No granuloma/atypia was seen.

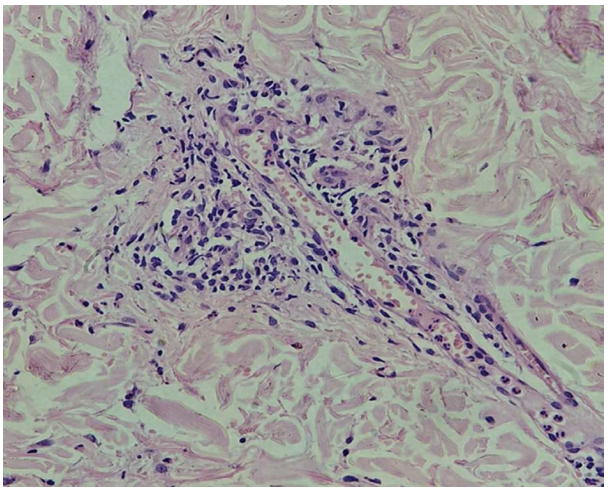

Figure 3c: High power view exhibiting upper and deeper dermis with few dilated blood vessels showing mild mixed perivascular infiltration comprising of polymorph, lymphocytes, and few eosinophils along with cellular debris. **(40x, H&E)**

The classic features of LCV, including perivascular neutrophilic infiltration, cellular debris, and endothelial proliferation clinched on the histological diagnosis of Cutaneous Leucocytoclastic Vasculitis.

Further on short course of systemic corticosteroids, resulted in significant clinical improvement within four weeks.

DISCUSSION

Leucocytoclastic vasculitis is an immune-mediated small vessel vasculitis presenting predominantly as palpable purpura. Cutaneous Vasculitis (CV) refers to a spectrum of disorders characterized by inflammation of blood vessels in the skin, leading to various clinical manifestations ranging from mild, self- limiting conditions to severe systemicinvolvement. In about 30% of individuals with LCV, extracutaneous involvement occurs, implying that the majority approximately 70% have disease limited to the skin.[3] It is a key dermatologic manifestation of systemic vasculitic syndromes and may also occur as a primary skin-limited disease.

The etiology of primary skin-limited LCV is often idiopathic, though hypersensitivity reactions, infections, or mild immune dysregulation may play a role. Diagnosis requires a thorough evaluation to rule out systemic vasculitis, and biopsy remains the gold standard for confirmation.[4]

Diagnosing primary skin-limited LCV presents several challenges.[5]

Histopathologic Variability: While skin biopsy remains the gold standard, findings can overlap with other vasculitic and inflammatory conditions.

Exclusion of Systemic Involvement: Extensive laboratory and imaging studies may be required to rule out systemic vasculitis.

Identification of Triggers: Medications, infections, and underlying autoimmune disorders must be carefully evaluated.

Intermittent and Self-Limiting Nature: Some cases resolve spontaneously, making long-term follow-up essential to ensure no progression to systemic disease.

Treatment is guided by disease severity, with immunosuppressive therapy reserved for systemic involvement. The favorable response to corticosteroids and trigger avoidance further supports the immune-mediated pathogenesis of LCV.[6]

CONCLUSION

Primary skin-limited leucocytoclastic vasculitis is a benign and self-limiting condition in most cases. A thorough diagnostic approach, including histopathology and laboratory workup, is essential to exclude systemic involvement. This case underscores the importance of recognizing skin-limited LCV as a distinct entity, guiding appropriate management strategies. Our case underscores the importance of a detailed history and comprehensive evaluation in diagnosing primary skin limited LCV. Although common triggers include infections and medications, this report highlights emphasizing the need for awareness among clinicians. A thorough diagnostic approach, including histopathology and serologic testing, is crucial for identifying underlying causes.

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