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Urticarial Vasculitis- Early Leucocytoclastic Vasculitis

1. Dr. Ravish Kumar Verma
Professor and Head of Department, Department of Internal Medicine,
Rama Medical College Hospital and Research Centre, Hapur,
Uttar Pradesh– 245304
Email: getravish1310@gmail.com
Mob.:9810695179

2. Dr. Neelam Anuragi
Professor, Department of Pathology
Rama Medical College Hospital and Research Centre, Hapur,
Uttar Pradesh– 245304
email: drneelamanuragi1978@gmail.com
Mob.9811148125

3. Dr. Arun Kumar Anuragi
Professor, Department of Internal Medicine,
Rama Medical College Hospital and Research Centre, Hapur,
Uttar Pradesh– 245304
Email: drakanuragiapi@gmail.com
Mob.9818590974

4. Dr. Prabhjot Singh S
PG Resident, Department of Internal Medicine,
Rama Medical College Hospital and Research Centre, Hapur,
Uttar Pradesh– 245304
Email: prabhjotsingh.5855@gmail.com
Mob:8610389042

5. Dr. Vinod Yadav
PG Resident, Department of Internal Medicine,
Rama Medical College Hospital and Research Centre, Hapur,
Uttar Pradesh– 245304

Abstract:

Urticarial vasculitis(UV) is a rare clinicopathologic entity that is characterized by chronic or recurrent episodes of urticarial lesions. This disease can be difficult to distinguish visually from those of chronic idiopathic urticaria but are unique individual lesions that persist for ≥24 hours, are often painful or burning rather than pruritic and can leave behind dusky hyperpigmentation. It is most often idiopathic but has been linked to certain drugs, infections, autoimmune connective disease, myelodysplastic disorders, and malignancies .UV can be classified into normocomplementemic and hypocomplementemic types, the latter often associated with systemic involvement and autoimmune diseases such as systemic lupus erythematosus. Diagnosis requires correlation of clinical features with investigations including complement levels, ESR/CRP, ANA, and a confirmatory skin biopsy showing small-vessel vasculitis.

Keywords:

Urticarial vasculitis, Hypocomplementemic urticarial vasculitis, Hypocomplementemic, Urticarial vasculitis syndrome, Leukocytoclastic vasculitis

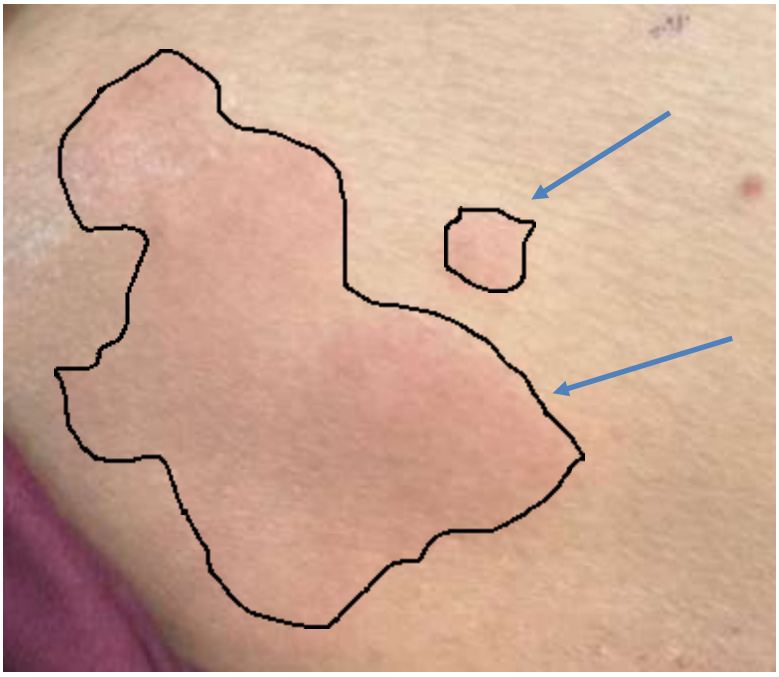
Case Report:

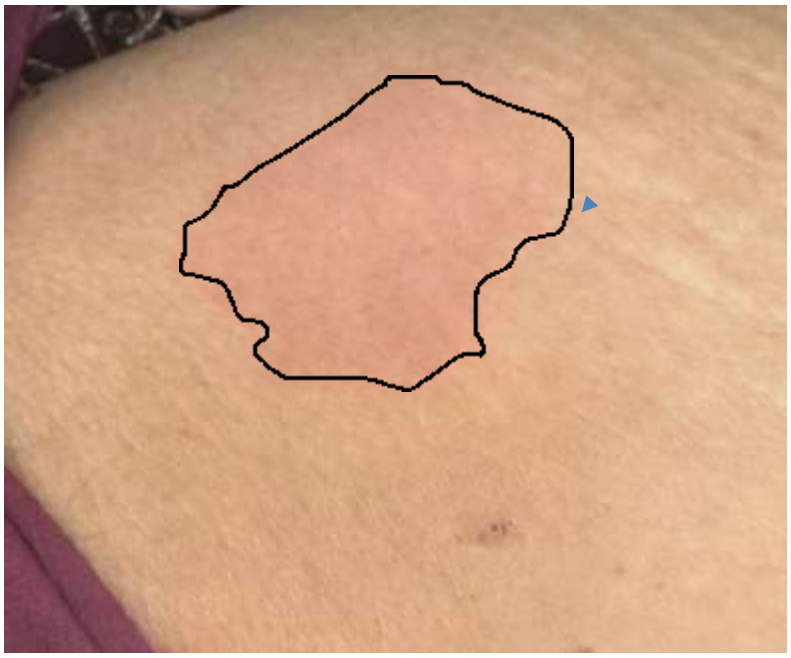
Patient Name: Gulerana
Age/Sex: 55-year-old Female
Place of Presentation: RAMA Hospital OPD
Chief Complaints:

- •Reddish lesions (wheals) over the trunk and bilateral upper and lower limbs
- •Burning sensation associated with the lesions
- •Duration: 10 days

Clinical Examination

- Multiple erythematous, non-blanchable wheals on trunk and all four limbs
- Presence of central dark red macules on several lesions
- No mucosal involvement
- No lymphadenopathy
- Systemic examination was unremarkable





Investigations

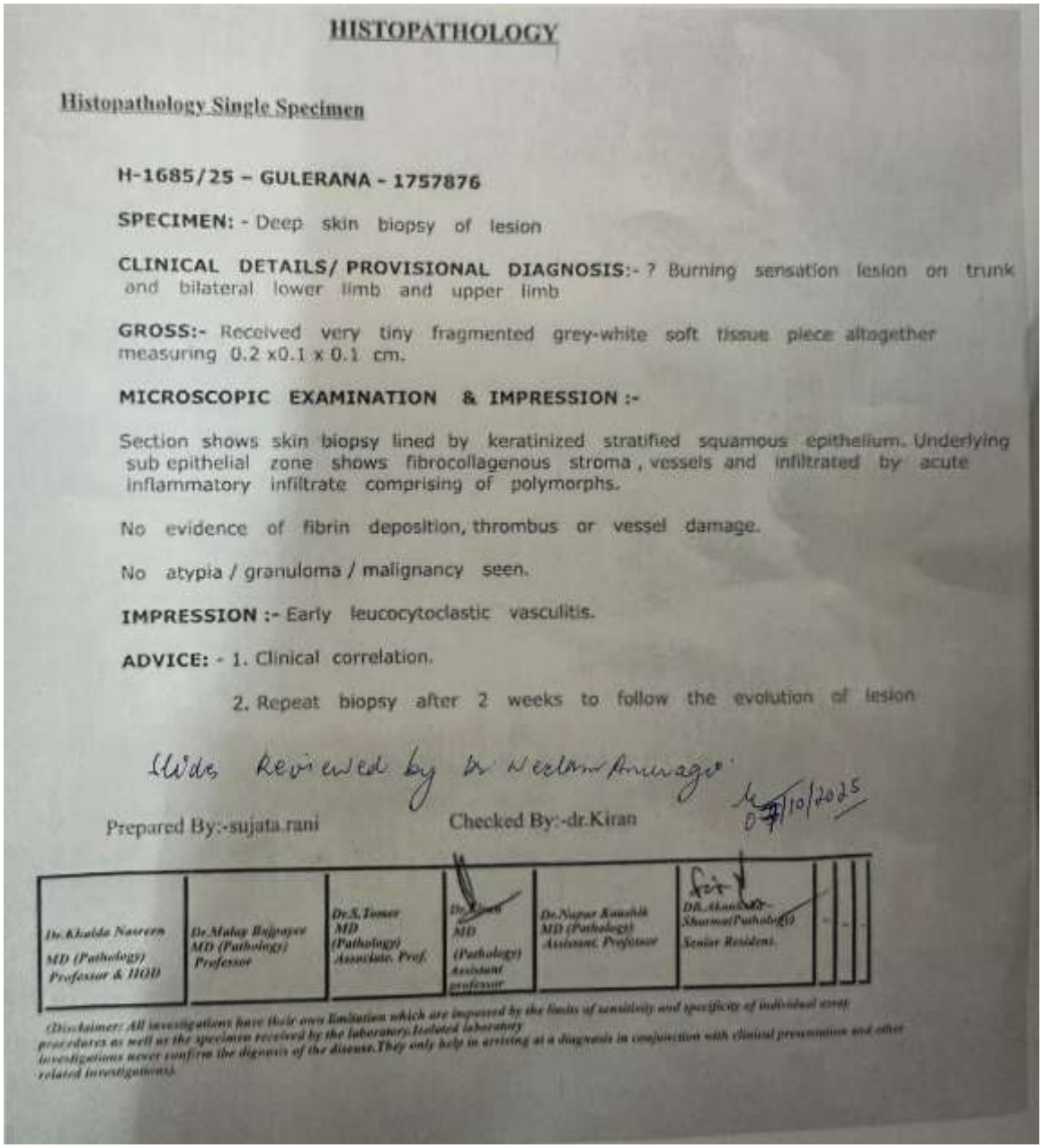
- ESR: Elevated
- Complement levels: Hypocomplementemia (low C3/C4)
- CBC: Within normal limits
- LFT/RFT: Within normal limits

MATERIALS AND METHODS :

persisting for more than 2This prospective observational study was conducted in the Department of Internal Medicine at Rama Medical College Hospital and Research Centre, Hapur, over a seven-month period from 10/11/2023 to 16/06/2024, including a total of 40 consecutive patients clinically suspected of having urticarial vasculitis. Inclusion criteria included adults aged 18–70 years presenting with urticarial lesions 4 hours, presence of purpura, post-inflammatory hyperpigmentation, burning or painful wheals, or systemic symptoms suggestive of vasculitis such as arthralgia or fever. Exclusion criteria included

patients with acute urticaria, chronic spontaneous urticaria without vasculitic signs, drug-induced urticaria unrelated to immune-complex mechanisms, systemic vasculitides of other etiologies, and immunosuppressed individuals. Detailed demographic data, clinical history, duration of symptoms, medication history, comorbidities, and systemic manifestations were recorded. A thorough physical examination was conducted, documenting morphology, distribution, number of lesions, presence of angioedema, and persistence beyond 24 hours. Routine laboratory investigations included complete blood count, ESR, CRP, liver and renal function tests, thyroid function, fasting blood glucose, and urinalysis. Autoimmune profiles comprising ANA, anti-dsDNA, rheumatoid factor, complement levels (C3, C4, CH50), and viral markers (HBsAg, anti-HCV, HIV) were assessed. All patients underwent a 4-mm punch biopsy from an active lesion present for at least 24–48 hours. Biopsies were processed using standard hematoxylin and eosin staining and examined by an experienced dermatopathologist. Histological features assessed included leucocytoclasia, neutrophilic infiltration, fibrinoid necrosis, endothelial swelling, perivascular infiltrates, erythrocyte extravasation, and immune-complex deposition where immunofluorescence was available. Complement deposition via direct immunofluorescence (DIF) was performed in selected cases to support diagnosis. Treatment decisions were individualized based on severity. Mild cases received non-sedating antihistamines, NSAIDs for arthralgia, and topical symptomatic therapy. Moderate to severe cases received systemic corticosteroids (prednisolone 0.5–1 mg/kg/day) tapered gradually. Patients with chronic or recurrent symptoms were considered for hydroxychloroquine, colchicine, or azathioprine. Follow-up was done every two weeks to monitor response, lesion recurrence, systemic involvement, and adverse effects. Outcomes were measured based on symptom resolution, improvement in lesion duration, normalization of inflammatory markers, and reduction in systemic manifestations. Statistical analysis was performed using SPSS version 25. Descriptive statistics were used for frequency distribution. Continuous variables were expressed as mean ± SD and categorical variables as percentages. Correlation analysis evaluated the relationship between complement levels, symptom severity, and histopathological findings. The study adhered to ethical guidelines and obtained approval from the Institutional Ethical Committee.

SKIN BIOPSY:



HISTOPATHOLOGY

Histopathology Single Specimen

H-1685/25 – GULERANA - 1757876

SPECIMEN: - Deep skin biopsy of lesion

CLINICAL DETAILS/ PROVISIONAL DIAGNOSIS:- ? Burning sensation lesion on trunk and bilateral lower limb and upper limb

GROSS:- Received very tiny fragmented grey-white soft tissue piece altogether measuring 0.2 x0.1 x 0.1 cm.

MICROSCOPIC EXAMINATION & IMPRESSION :-

Section shows skin biopsy lined by keratinized stratified squamous epithelium. Underlying sub epithelial zone shows fibrocollagenous stroma , vessels and infiltrated by acute inflammatory infiltrate comprising of polymorphs.

No evidence of fibrin deposition, thrombus or vessel damage.

No atypia / granuloma / malignancy seen.

IMPRESSION :- Early leucocytoclastic vasculitis.

ADVICE: - 1. Clinical correlation.

2. Repeat biopsy after 2 weeks to follow the evolution of lesion

Slide Reviewed by Dr Neelam Anuraga

Prepared By:-sujata rani

Checked By:-dr.Kiran

07/10/2025

Dr.Khalda Naveen MD (Pathology) Professor & HOD	Dr.Malay Bajpayee MD (Pathology) Professor	Dr.S.Tomar MD (Pathology) Associate. Prof.	[illegible] MD (Pathology) Assistant professor	Dr.Nupur Kaushik MD (Pathology) Assistant Professor	DR. [illegible] Sharma(Pathology) Senior Resident

Disclaimer: All investigations have their own limitation which are imposed by the limits of sensitivity and specificity of individual assay procedures as well as the specimen received by the laboratory.Isolated laboratory investigations never confirm the diagnosis of the disease.They only help in arriving at a diagnosis in conjunction with clinical presentation and other related investigations.

Discussion:

Urticarial vasculitis is characterized by urticarial lesions lasting >24 hours with histopathological features of small-vessel vasculitis. Hypocomplementemic UV is more commonly associated with systemic features and autoimmune disorders. Elevated ESR and

low complement levels help support the diagnosis. Differentiation from chronic spontaneous urticaria is essential, as management and prognosis differ significantly.

Histologic findings:

Urticarial vasculitis is a leukocytoclastic vasculitis and most often affects the postcapillary venules of the skin. Because the presentation of this condition can vary, a lesional biopsy is considered the gold standard for diagnosis among clinicians (Kolkhir et al., 2020). As is the case with small vessel vasculitis, demonstration of the full histologic picture requires biopsy of the lesion at the exact stage of evolution and is influenced by therapy. Although not always seen, common diagnostic features include damage to the dermal vessels (including endothelial swelling and associated luminal occlusion), karyorrhexis of neutrophils with production of nuclear dust, and extravasation of erythrocytes into the dermis. Fibrinoid changes of the vessel walls are also often seen (Zuberbier and Maurer 2014). Inflammatory infiltrate can be detected in the walls of the vessels and perivascularly and is generally composed of neutrophils, eosinophils, and/or lymphocytes. As lesions age, the infiltrate tends to shift from neutrophil or eosinophil dominant to primarily lymphocytic (Damman et al., 2020). Neutrophil extracellular traps have also been found in some patients, and investigators postulate that this feature is potentially reflective of the severity of the disease (Bonnekoh et al., 2019). Deposits of IgM, IgG, and less frequently of IgA, C1q, C4, C3, or fibrinogen, are commonly found within the vessel walls on immunopathology (Fig. 3; Zuberbier and Maurer 2014).

Conclusion:

Urticarial vasculitis is a rare clinicopathological entity that most often presents cutaneously as classic indurated wheals. These lesions have some distinguishing features, such as a duration in excess of 24 hours and the presence of a residual dusky hyperpigmentation, that aid with diagnosis, but biopsy should be obtained for an accurate diagnosis. Clinicians should be judicious when selecting the lesion for evaluation, and patient history should be used to contextualize the results. Histopathologic specimens for urticarial vasculitis often exhibit some features of leukocytoclastic vasculitis. Systemic involvement can occur as well, with pulmonary complications being the primary cause of mortality.

This disease is often idiopathic, but it can also be linked to some infections, drugs, autoimmune disorders, and malignancies. When a cause is known, treatment of the underlying disease or disorder or removal of the complicit antigen should be completed before any other therapies are administered. Currently used medications for the treatment of urticarial vasculitis include dapsone and colchicine, hydroxychloroquine, immunosuppressives, corticosteroids, and select biologics.

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