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Urticarial Vasculitis and Hypereosinophilia Unmasking Coeliac Disease and Eosinophilic Granulomatosis with Polyangiitis in a 57-Year-Old Woman

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Abstract | **Case Report**

Introduction: Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare small-vessel vasculitis characterised by asthma, eosinophilia, and systemic involvement. Its association with coeliac disease (CD) is extremely rare but may share common immunological mechanisms. ***Case presentation:*** A 57-year-old woman with asthma presented with fixed purpuric and pruritic plaques, digestive symptoms, and distal paraesthesias. She had marked hypereosinophilia (6 000 cells/µL), eosinophilic vasculitis on skin biopsy, and axonal polyneuropathy. Further investigations revealed subtotal villous atrophy and positive coeliac serology, confirming coeliac disease. Treatment with corticosteroids and a gluten-free diet led to a favourable outcome. ***Discussion:*** This case illustrates how hypereosinophilia can serve as a clinical bridge between EGPA and coeliac disease. The coexistence of both conditions likely reflects a shared Th2-driven immunopathogenesis. Identifying this overlap is crucial for guiding both immunosuppressive and dietary management. ***Conclusion:*** In patients with unexplained hypereosinophilia and multisystem involvement, the combination of EGPA and coeliac disease, though rare, should be considered.

Keywords: Eosinophilic Granulomatosis with Polyangiitis, Hypereosinophilia, Urticarial Vasculitis, Coeliac Disease.

INTRODUCTION

Eosinophilic granulomatosis with polyangiitis is a rare systemic necrotising vasculitis affecting small to medium-sized vessels, characterised by asthma, peripheral eosinophilia, and eosinophilic tissue infiltration. Cutaneous manifestations are frequent and diverse, with urticarial vasculitis being a rare but potentially inaugural presentation [1]. Hypereosinophilia is a key feature of EGPA but may also reveal other immune-mediated conditions such as coeliac disease (CD), which, although primarily gastrointestinal, can present with systemic, including cutaneous and neurological, manifestations [2, 3]. The co-occurrence of EGPA and CD is exceedingly rare but biologically plausible, as both conditions share immunogenetic susceptibility and Th2-skewed immune responses. We report the case of a 57-year-old woman in whom fixed urticarial vasculitis and marked hypereosinophilia revealed the dual diagnosis of EGPA and previously unrecognised coeliac disease.

CASE PRESENTATION

A 57-year-old woman, previously treated and cured of pleuro-pulmonary tuberculosis (2019) and followed for inhaler-controlled asthma, presented with a one-month history of fixed, purpuric, intensely pruritic plaques distributed over the trunk, abdomen, back, and thighs (Figure 1). Each lesion persisted for several days and resolved with residual hyperpigmented macules (Figure 2), suggesting urticarial vasculitis rather than ordinary urticaria. During the week preceding admission, she developed diffuse abdominal pain, nausea, episodic vomiting, intermittent watery diarrhoea, abdominal bloating, and chronic distal paraesthesias of the upper and lower limbs.

Complete blood count showed marked hypereosinophilia at 6 000 cells μL^{-1} with otherwise unremarkable haematological indices. Complement levels were low. Liver and renal panels were normal. Serum IgE was elevated, and ANCA were negative. Bone-marrow aspiration showed 22 % eosinophils

(reference 2 – 3 %), and trephine biopsy confirmed mature eosinophilic hyperplasia.

A skin biopsy of an active plaque demonstrated a dense superficial and deep perivascular and interstitial infiltrate dominated by eosinophils with focal fibrinoid necrosis of vessel walls—features diagnostic of eosinophilic vasculitis. Direct immunofluorescence was negative.

Electroneuromyography revealed an axonal sensorimotor polyneuropathy involving both lower and upper limbs, consistent with vasculitic neuropathy.

Upper endoscopy disclosed erosive antral gastritis and erosive bulboduodenitis. Histology of jejunal and duodenal biopsies revealed subtotal villous atrophy with crypt hyperplasia and intra-epithelial lymphocytosis (~30 %), highly suggestive of coeliac disease. Serology was positive for anti-tissue-transglutaminase, anti-gliadin, and anti-endomysium antibodies. Imaging of the abdomen showed moderate chylous ascites of undetermined origin; parasitological studies were negative.

The combination of asthma, marked peripheral and medullary eosinophilia, biopsy-proven eosinophilic vasculitis, axonal polyneuropathy, and multisystem involvement satisfied the 2022 ACR/EULAR criteria for eosinophilic granulomatosis with polyangiitis (EGPA). Concomitant histologically and serologically proven coeliac disease provided a remarkable association, positioning hypereosinophilia as the clinical bridge between cutaneous vasculitis and the underlying intestinal autoimmune disorder.

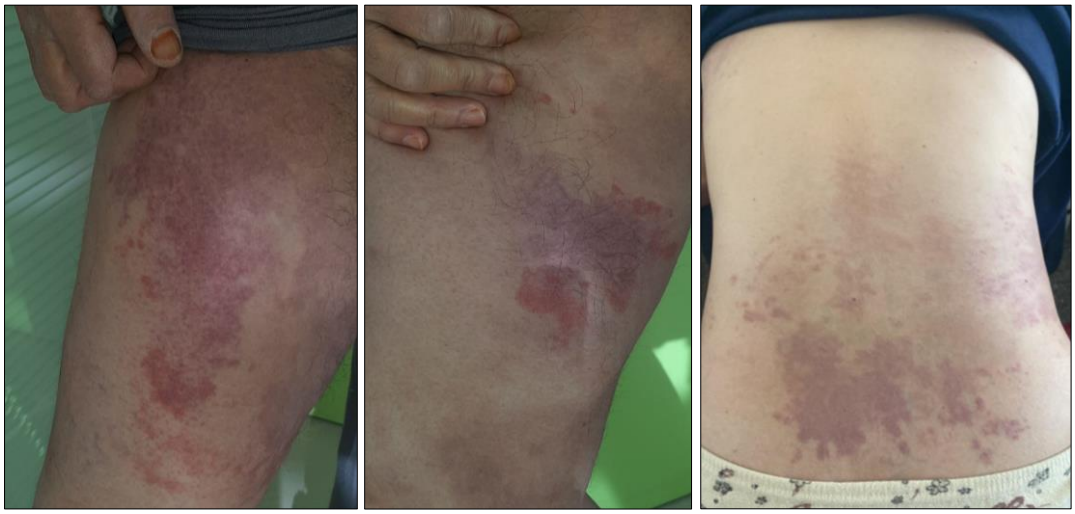

Figure 1: Initial Cutaneous Manifestations of Eosinophilic Vasculitis

Fixed, purpuric, and intensely pruritic plaques observed on the trunk, abdomen, back, and thighs at disease onset, consistent with urticarial vasculitis in the context of hypereosinophilic syndrome.

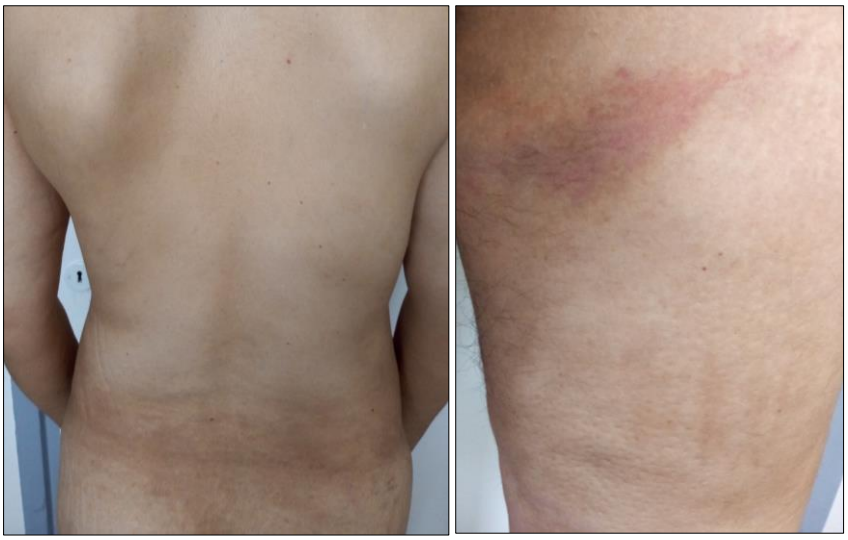

Figure 2: Post-inflammatory Sequelae of Cutaneous Vasculitis

Residual hyperpigmented macules following resolution of the purpuric plaques, reflecting the chronicity and vasculitic nature of the cutaneous lesions.

DISCUSSION

The striking hypereosinophilia that dominated our patient's blood (6 000 cells μL^{-1}) and marrow (22 %) is pathognomonic of the eosinophilic phenotype of eosinophilic granulomatosis with polyangiitis (EGPA). Eosinophils, driven primarily by interleukin-5 (IL-5) and other Th2-cytokines, are now recognised as central effectors rather than mere epiphenomena in EGPA pathogenesis, orchestrating tissue damage in skin, nerves and the gastrointestinal tract [4,5]. The dense eosinophilic vasculitis on skin biopsy, together with asthma and vasculitic neuropathy, fulfilled the 2022 ACR/EULAR criteria for EGPA and explains the urticarial vasculitis that heralded disease onset.

Celiac disease (CD), confirmed here by subtotal villous atrophy and triple-positive serology, is itself an immune-mediated enteropathy characterised by a mucosal Th1/Th2 imbalance. IL-5 transcription within duodenal eosinophils of untreated CD patients directly links gluten exposure to eosinophil activation and systemic recruitment [6]. Clinically, peripheral or tissue eosinophilia accompanies a minority of adult CD cases, but when present it can be profound and occasionally mimics hypereosinophilic syndromes [7]. Epidemiological series also document an excess of eosinophil-driven disorders such as eosinophilic oesophagitis among CD cohorts, underscoring a shared immunopathological terrain [8].

Several lines of evidence help to rationalise the exceptional coexistence of EGPA and CD. First, genome-wide association studies have revealed overlapping susceptibility loci on chromosome 5 (IL-4/IL-5/IRF1 cluster) and within the HLA-DQ region, implicating common pathways of mucosal barrier dysfunction and Th2 skewing in ANCA-negative EGPA and in CD [5]. Second, gluten-induced epithelial stress in genetically primed individuals amplifies IL-5 production. Third, chronic intestinal permeability could expose the systemic immune system to luminal antigens, fostering the extravascular granulomatous vasculitis typical of EGPA.

Our case illustrates how hypereosinophilia can serve as a clinical bridge between two seemingly disparate entities: urticarial vasculitis evolving into full-blown EGPA and silent CD unmasked by digestive symptoms. Recognition of this relationship is clinically relevant because a gluten-free diet, alongside corticosteroids, not only induced mucosal healing but also paralleled the rapid normalisation of eosinophil counts and cutaneous remission. Whether gluten withdrawal alone can modify the natural history of EGPA remains uncertain, yet the favourable early outcome in our patient encourages systematic CD screening in adults presenting with hypereosinophilic EGPA, especially when gastrointestinal complaints are prominent.

CONCLUSION

Certainly! Blood hypereosinophilia is a complex aspect of hematological pathology, revealing its immunological and clinical intricacies. Recognizing it as a potentially revealing sign, especially when symptoms are vague, emphasizes the need for a thorough diagnostic approach, highlighting the various possible etiologies ranging from parasitic causes to tumor related and drug-related implications.

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