Vasculitis associated with haematologic malignancies

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Purpose of review
This review examines the complex bidirectional relationship between vasculitis and hematologic malignancies, highlighting the importance of meticulous diagnostic assessment.

Recent findings
Vasculitis may emerge in the setting of hematologic malignancies via mechanisms such as paraneoplastic inflammation, immune dysregulation, drug exposure, and clonal hematopoiesis. Myeloid neoplasms – especially myelodysplastic syndrome (MDS) and chronic myelomonocytic leukemia (CMML) – show a stronger association than lymphoid malignancies, with cutaneous small vessel vasculitis being the most common subtype. VEXAS syndrome exemplifies the overlap between autoinflammation and hematologic disease, often presenting with vasculitic features and macrocytic anemia.
In lymphoproliferative disorders and plasma cell dyscrasias, vasculitis may precede, mimic, or complicate the malignancy. Entities such as intravascular lymphoma, angioimmunoblastic T-cell lymphoma, and monoclonal gammopathies – including MGUS and multiple myeloma – can manifest with vasculitic symptoms, requiring histopathologic and molecular evaluation. Emerging concepts like monoclonal gammopathy of cutaneous and rheumatologic significance highlight the need for interdisciplinary care. Drug-induced vasculitis, particularly from immunomodulatory agents and biologics, adds diagnostic complexity. Atypical features – such as unexplained cytopenias, dual autoantibody positivity, or poor response to immunosuppression – should prompt evaluation for underlying hematologic disease. Conversely, vasculitis may signal complications in patients with known hematologic disorders.

Summary
Early suspicion of vasculitis associated with hematologic malignancies and accurate diagnosis are important in guiding therapeutic approaches.

Keywords
Behçet syndrome, chronic myelomonocytic leukemia, myelodysplastic syndrome, MGUS, monoclonal gammopathy of cutaneous significance, vasculitis, VEXAS

INTRODUCTION

Patients with hematologic malignancies may initially present to rheumatology clinics due to a variety of inflammatory manifestations that overlap with autoimmune and vasculitic syndromes. Hematologic neoplasms – particularly those of myeloid and lymphoid origin – can mimic, coexist with, or even underlie vasculitic processes. Although the majority of patients diagnosed with vasculitis do not harbor an underlying malignancy, certain hematologic cancers may present with features suggestive of vasculitis or may emerge as a consequence of autoimmune dysregulation.

Referral to hematology is often prompted by laboratory abnormalities such as cytopenias, monocytosis, eosinophilia, or acquired bleeding diatheses in patients with suspected vasculitis. Notably, autoimmune phenomena – including vasculitis – occur in up to 30% of patients with myeloid or lymphoid malignancies. Among these, myeloid disorders such as MDS and chronic myelomonocytic leukemia (CMML) exhibit a stronger association with vasculitis compared to lymphoproliferative diseases [1].

The vasculitic involvement in hematologic malignancies typically affects small cutaneous or

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KEY POINTS

- Presenting with unexplained vasculitis should lead the clinicians to investigate underlying hematologic malignancies and collaboration between hematologists and rheumatologists is important.
- The vasculitis diagnosis and treatment in the context of hematologic malignancies is difficult because of the clinical heterogeneity, lack of specific serologic tests or markers and challenges in obtaining tissue samples.
- Integration of myeloid driver gene features like TET2 in hematologic malignancy associated vasculitis diagnosis may be more commonly used in the future.

medium-sized muscular arteries, whereas ANCA-associated and large-vessel vasculitis are less frequently observed. Conversely, approximately one-third of patients with autoimmune diseases may have an underlying MDS or CMML, underscoring the bidirectional relationship between these entities [2].

Diagnostic differentiation between primary vasculitis and paraneoplastic or malignancy-associated vasculitis requires a comprehensive approach, including complete blood count with differential, PET, histopathological evaluation, and clonal analyses, particularly via next-generation sequencing (NGS). Despite growing recognition of these associations, therapeutic data remain limited and largely anecdotal [3▪▪].

MYELOID MALIGNANCIES

Autoimmune and inflammatory manifestations are increasingly recognized in patients with MDS and *CMML*. These include vasculitis, connective tissue diseases, and inflammatory arthritis. Vasculitis in the context of MDS/CMML may arise as a paraneoplastic phenomenon, but infectious and drug-induced etiologies [4] must also be considered. Interestingly, progression to acute leukemia appears to be less frequent in MDS/CMML patients who develop vasculitis [5]. No consistent correlation has been identified between MDS subtypes, cytogenetic abnormalities, and the presence of autoimmune manifestations, and the prognostic impact of coexisting autoimmune disease remains unclear [6,7].

Myelodysplastic syndrome

Autoimmune and systemic inflammatory conditions are observed in approximately 10–25% of patients with MDS, encompassing neutrophilic dermatoses, connective tissue disorders, arthritis, and vasculitis [8]. Among vasculitic presentations, cutaneous small vessel vasculitis – particularly leukocytoclastic vasculitis (LCV) – is the most frequently reported subtype [7]. Giant cell arteritis (GCA) associated with MDS tends to exhibit attenuated clinical features compared to idiopathic GCA.

A systematic review of cutaneous manifestations in MDS identified vasculitis in 15.4% of cases (21/134 patients), with subtypes including cutaneous polyarteritis nodosa (PAN), LCV, Behçet syndrome, and unclassified vasculitides. Notably, MDS had a fatal course in 50% of these patients, with a median survival of 7.5 months following vasculitis onset [9].

Renal involvement in MDS-associated vasculitis was evaluated in a multicenter retrospective study by Lafargue *et al*. [8], which identified ANCA-associated glomerulonephritis as the predominant cause of renal injury. The therapeutic impact of hypomethylating agents on renal outcomes remains to be elucidated [8].

VEXAS syndrome

VEXAS syndrome (Vacuoles, E1 enzyme, X-linked, Autoinflammation, Somatic) is a recently characterized monogenic autoinflammatory disorder caused by somatic mutations in the UBA1 gene, predominantly affecting men over 40 years of age. The syndrome manifests with multisystem inflammation involving the skin, lungs, and bone marrow, and frequently mimics vasculitic diseases [10]. Vasculitis occurs in approximately 25% of patients and spans all vessel sizes, with PAN and LCV being the most common subtypes. Cutaneous involvement is present in 85% of cases, while visceral small vessel vasculitis affecting the lungs and kidneys is rare [11▪]. Hematologic abnormalities in VEXAS syndrome typically include macrocytic anemia and other cytopenias, with frequent progression to MDS or plasma cell dyscrasias. A hallmark diagnostic feature is cytoplasmic vacuolization in erythroid and myeloid precursors, warranting careful hematopathological evaluation [12].

Patients with VEXAS syndrome are often misdiagnosed with atypical, treatment-resistant vasculitis. Distinguishing features include leukocytoclasia without true vasculitis, neutrophilic alveolitis, non-vasculitic ocular and neurologic symptoms, and poor response to conventional immunosuppression. Clinical clues such as male sex, age at least 40 years, and cytopenias (macrocytic anemia, thrombocytopenia, monocytopenia) should prompt consideration of VEXAS [13▪▪].

Case reports have highlighted diverse presentations, including large vessel vasculitis refractory to steroids and granulomatosis with polyangiitis (GPA) with cartilage involvement responsive to methotrexate and infliximab [14,15]. In a cohort of 89 male patients with VEXAS syndrome, Sullivan *et al.* [14] identified vasculitis in 21 individuals, predominantly cutaneous LCV. Only one patient had GCA despite frequent cranial symptoms, and no association with thrombosis was observed [14]. Importantly vasculitis was not found to be correlated to thrombosis. The authors suggested that men at least 50 years of age with atypical and steroid-dependent vasculitis should be considered for evaluation of VEXAS syndrome.

Therapeutic strategies for VEXAS syndrome include high-dose corticosteroids and steroid-sparing agents such as JAK inhibitors (ruxolitinib, baricitinib), IL-6 inhibitors, hypomethylating agents, and allogeneic hematopoietic stem cell transplantation (HSCT) [16]. Durable glucocorticoid-free remissions are rare without hypomethylating therapy or HSCT. Reduction in UBA1 allele burden has been observed in responders to hypomethylating agents, suggesting a potential role for molecular monitoring in guiding treatment decisions [17].

Behçet syndrome and myelodysplastic syndromes

Behçet syndrome is a chronic, relapsing, multisystem vasculitis of unknown aetiology, characterized by mucocutaneous, ocular, vascular, and gastrointestinal involvement. A distinct clinical entity has emerged in patients with concurrent Behçet syndrome and myelodysplastic syndrome (BS-MDS), exhibiting features that diverge from either condition alone. Patients with BS-MDS tend to be older, display less frequent ocular involvement, and more commonly present with gastrointestinal manifestations compared to those with idiopathic Behçet syndrome. Notably, trisomy 8 is a recurrent cytogenetic abnormality in BS-MDS, particularly in cases with intestinal involvement [18].

Park *et al.* [6] conducted a multicenter retrospective analysis of 35 patients with intestinal BS-MDS across four Korean centers, comparing them to patients with intestinal Behçet syndrome without MDS. The BS-MDS cohort demonstrated higher mortality rates and greater refractoriness to conventional immunosuppressive therapies. In such cases, targeting the underlying MDS – especially when intestinal Behçet syndrome is refractory – has shown therapeutic benefit. Allogeneic HSCT may offer disease control, though its implementation requires multidisciplinary evaluation and careful consideration of risks, including graft-versus-host disease and infectious complications [6]. The efficacy of HSCT in BS-MDS has also been supported by case reports and literature reviews [19].

Chronic myelomonocytic leukemia

CMML, a clonal hematopoietic disorder classified within the spectrum of myelodysplastic/ myeloproliferative neoplasms, is frequently associated with autoimmune phenomena, occurring in approximately 15–25% of patients. Vasculitis is the most common autoimmune manifestation, with medium-sized vessel vasculitis (e.g., polyarteritis nodosa) and large-vessel vasculitis (e.g., GCA, Takayasu arteritis) being particularly prevalent. Large-vessel vasculitis may precede the diagnosis of CMML and serve as an early clinical clue.

In a multicenter French study, GCA emerged as the most frequent vasculitis subtype among patients with MDS/CMML, likely reflecting the shared demographic profile of older age [5]. Vasculitis may occur before or after CMML diagnosis, and its presence often correlates with distinct clinical and molecular features. Clonal hematopoiesis, driven by somatic mutations such as TET2, plays a pivotal role in both hematologic malignancy and inflammatory disease trajectories. Robinette *et al.* [20▪] recently demonstrated an increased incidence of GCA in patients with clonal hematopoiesis, particularly those harboring TET2 mutations and cytopenias. Intriguingly, TET2 mutations were associated with vision loss in GCA patients, even in the absence of elevated C-reactive protein, suggesting a genotype-specific inflammatory phenotype [20▪].

Patients with CMML-associated vasculitis tend to be younger than the general CMML population and exhibit a male predominance [21]. Although rare in adults, IgA vasculitis with cutaneous involvement has been reported in CMML, further highlighting the spectrum of vasculitic presentations. Altered monocyte/macrophage gene expression profiles have also been implicated in IgA nephropathy, suggesting a shared immunopathogenic mechanism [2].

LYMPHOID MALIGNANCIES

The association between vasculitis and lymphoproliferative disorders is rare but clinically significant. Various lymphoma subtypes may mimic vasculitic syndromes, leading to diagnostic challenges and potential delays in appropriate treatment.

Lymphoproliferative disorders

LCV, characterized by neutrophilic infiltration of small vessels, has diverse etiologies including

infections, medications, cryoglobulinemia, and neoplastic conditions. LCV is associated with lymphoproliferative diseases in approximately 1% of cases and may precede, coincide with, or follow the diagnosis of the underlying hematologic malignancy [22].

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis has been reported in conjunction with lymphomas and multiple myeloma. Proposed mechanisms include malignant infiltration of tissues (e.g., in chronic lymphocytic leukemia), monoclonal plasma cell production of ANCA, or vasculitis triggered by prior immunosuppressive therapy. Multiple myeloma may clinically mimic ANCA-associated vasculitis with features such as palpable purpura, peripheral neuropathy, and hemorrhagic nasal crusts, often in the absence of ANCA seropositivity [22]. Clues for underlying multiple myeloma include monoclonal gammopathy and C4 hypocomplementemia [23].

Intravascular lymphoma (IVL), a rare and aggressive subtype of non-Hodgkin lymphoma, frequently involves the skin, lungs, and central nervous system (CNS). IVL may be misdiagnosed as vasculitis due to its cutaneous and neurologic manifestations. Multiple biopsies from active lesions are essential for diagnosis. ANCA-positive IVL has also been described, further complicating the differential diagnosis [24]. Fever, neurologic symptoms, elevated LDH, and atypical ANCA positivity should prompt consideration of IVL and warrant skin or bone marrow biopsy [25–30].

Angioimmunoblastic T-cell lymphoma (AITL) has been reported following IgA vasculitis, suggesting a possible pathogenic link via tumor antigen-driven T-cell activation and aberrant IgA production [31]. NK/T-cell lymphoma may present with eosinophilia and systemic symptoms mimicking eosinophilic granulomatosis with polyangiitis, with vasculitis resulting from direct vascular infiltration by tumor cells [32].

Paraneoplastic urticarial vasculitis has been described in Hodgkin lymphoma, presenting with acute urticaria and resolving with chemotherapy. Proposed mechanisms include immune complex deposition, histamine release, and mast cell degranulation [33].

Large granular lymphocyte (LGL) leukemia, a rare indolent lymphoproliferative disorder of clonal T or NK cells, is associated with autoimmune conditions in 25–32% of cases, including vasculitis. Cryoglobulinemia, LCV, and ANCA-negative microscopic polyangiitis have been reported [34]. Patients often present with neutropenia, anemia, polyclonal hypergammaglobulinemia, splenomegaly, and increased circulating LGLs. A case of T-LGL leukemia presenting with necrotic bullous LCV was differentiated from reactive LGL proliferation by bone marrow infiltration and TCR αβ expression [35].

Hairy cell leukemia may rarely underlie cutaneous LCV. Cladribine, a standard treatment, may exacerbate vasculitic symptoms, while vemurafenib and corticosteroids have been used in combination [36].

Plasma cell disorders

Monoclonal gammopathies span a spectrum from asymptomatic monoclonal gammopathy of undetermined significance (MGUS) to aggressive plasma cell leukemia. MGUS, though premalignant, may lead to renal, neurologic, and cutaneous complications. The concept of monoclonal gammopathy of cutaneous significance (MGCS) has emerged to describe dermatologic conditions associated with monoclonal gammopathies not meeting criteria for hematologic intervention [37].

MGCS is categorized by underlying pathophysiologic mechanism. Group 1 involves direct effects of immunoglobulin products, such as cryoglobulinemia, which causes vasculitis through immune complex deposition [38]. Cryoglobulins are classified into type I (monoclonal IgM, IgG, or IgA) and mixed types II and III. Type I cryoglobulinemia is commonly associated with hematologic malignancies, including Waldenström's macroglobulinemia, multiple myeloma, MGUS, non-Hodgkin lymphoma, and chronic lymphocytic leukemia [39].

Group 2 MGCS involves indirect mechanisms, where monoclonal gammopathies trigger autoantibody production, cytokine release, and complement activation, leading to dermal inflammation. LCV is a common manifestation, typically presenting as palpable purpura on the lower extremities [40]. Differentiating incidental monoclonal gammopathies from pathogenic monoclonal gammopathies requires hematologic collaboration. Brummer *et al.* [41] described a patient with LCV mimicking ulcus cruris as the initial sign of smoldering multiple myeloma, with complete remission following plasma cell-directed therapy. LCV arising in the context of plasma cell diseases has been documented mostly in the form of case reports and the published cases are summarized by Brummer *et al.* [41]. LCV occurred typically as diffuse palpable purpura on the limbs or the trunk and was the initial manifestation of these multiple myeloma patients. In fewer patients, LCV developed over the course of multiple myeloma and was attributed to immunomodulatory drugs and/or proteasome inhibitors. Immune complex deposition, altered cytokine production, endothelial dysfunction, and complement system activation cause

inflammation and blood vessel infiltration by neutrophils [41].

IgA paraproteinemia may cause IgA-associated LCV. Cryoglobulinemia is present in approximately 20% of IgA vasculitis cases but is not required for diagnosis in patients with M-protein [42]. Marginal zone lymphoma has been reported with type II cryoglobulinemia vasculitis involving the intestines, skin, and lungs, with cryocrit used for response monitoring [43].

Monoclonal gammopathy has also been linked to renal pathology, termed monoclonal gammopathy of renal significance (MGRS) [44]. In a recent study of ANCA-negative pauci-immune crescentic glomerulonephritis (PICGN), eight of 14 patients had MG. Proposed mechanisms include monoclonal protein deposition, complement activation, endothelial injury, and neutrophil extracellular trap formation. The patients with MG were older and had lower eGFR, proteinuria, hemoglobin and complement levels but higher erythrocyte sedimentation rate. Kidney biopsies of patients with monoclonal gammopathy revealed a higher degree of fibrosis compared to patients without monoclonal gammopathy and patients with monoclonal gammopathy had a higher mortality rate. In ANCA-negative PICGN, monoclonal gammopathy should be investigated [45].

Plasma cell dyscrasias, particularly MGUS and multiple myeloma, are also among the hematologic manifestations of VEXAS syndrome [3▪▪]. Importantly, monoclonal gammopathy may arise in rheumatologic diseases, including those treated with biologics, prompting the recent proposal of monoclonal gammopathy of rheumatologic significance [46▪].

MISCELLANEOUS HEMATOLOGIC CONDITIONS ASSOCIATED WITH VASCULITIS

Vasculitis may arise in the context of various hematologic conditions and treatments, often presenting diagnostic and therapeutic challenges [47]. LCV, in particular, can be drug-induced, with immunomodulatory agents such as lenalidomide – commonly used in multiple myeloma – implicated in its pathogenesis.

Rituximab, a monoclonal anti-CD20 antibody widely employed in both B-cell lymphomas and autoimmune vasculitis, has paradoxically been associated with LCV. This adverse effect may occur days to weeks postadministration and is hypothesized to result from rituximab-antirituximab immune complex formation, B-cell depletion, and cytokine release [48].

Cryoglobulinemic vasculitis affects approximately 3% of patients with Sjögren's syndrome and confers an elevated risk for progression to non-Hodgkin lymphoma. It is to be noted that in a small cohort of 13 patients with Sjögren's syndrome-associated cryoglobulinemic vasculitis, rituximab therapy did not appear to influence lymphoma evolution [49].

Autoimmune phenomena, including vasculitis, may also develop following allogeneic HSCT.

WHEN TO SUSPECT UNDERLYING HEMATOLOGIC MALIGNANCY IN VASCULITIS

Recognition of hematologic malignancy in patients presenting with vasculitis requires vigilance, especially when clinical or laboratory features deviate from typical autoimmune patterns. Key red flags include unexplained cytopenias, dual autoantibody positivity, poor response to immunosuppression, and bone marrow infiltration on MRI [50].

Conversely, in patients with established hematopoietic disorders, the emergence of atypical systemic symptoms – such as prolonged fever, polymyalgia rheumatica-like manifestations, sensorineural hearing loss, nephropathy, or unexplained eosinophilia – should raise suspicion for vasculitic involvement.

CONCLUSION

Recognizing atypical presentations, serologic anomalies, and treatment-resistant vasculitis is essential for timely identification of underlying hematologic disease.

Multidisciplinary collaboration, integration of molecular diagnostics, and individualized therapeutic strategies remain pivotal in optimizing outcomes for this diagnostically challenging and clinically heterogeneous group of patients.

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Conflicts of interest

There are no conflicts of interest.

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- ▪ of special interest
- ▪▪ of outstanding interest

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