Review

Urticarial vasculitis

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ABSTRACT

Urticarial vasculitis is a rare clinicopathologic entity that is characterized by chronic or recurrent episodes of urticarial lesions. Skin findings of this disease can be difficult to distinguish visually from those of chronic idiopathic urticaria but are unique in that individual lesions persist for ≥24 hours and can leave behind dusky hyperpigmentation. This disease is most often idiopathic but has been linked to certain drugs, infections, autoimmune connective disease, myelodysplastic disorders, and malignancies. More recently, some authors have reported associations between urticarial vasculitis and COVID-19, as well as influenza A/H1N1 infection. Urticarial vasculitis can extend systemically as well, most often affecting the musculoskeletal, renal, pulmonary, gastrointestinal, and ocular systems. Features of leukocytoclastic vasculitis seen on histopathologic examination are diagnostic of this disease, but not always seen. In practice, antibiotics, dapsone, colchicine, and hydroxychloroquine are popular first-line therapies, especially for mild cutaneous disease. In more severe cases, immunosuppressives, including methotrexate, mycophenolate mofetil, azathioprine, and cyclosporine, as well as corticosteroids, may be necessary for control. More recently, select biologic therapies, including rituximab, omalizumab, and interleukin-1 inhibitors have shown promise for the treatment of recalcitrant or refractory cases.

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Introduction

Urticarial vasculitis is a rare clinicopathologic entity that manifests as a result of inflammatory injury to the small vessels of the skin. This condition is characterized by chronic or recurrent episodes of urticarial lesions showing features of leukocytoclastic vasculitis on histopathologic specimens. Findings can be limited to the skin or extend systemically, affecting various organs, including the musculoskeletal, renal, pulmonary, gastrointestinal, and ocular systems (Fig. 1).

Epidemiology

Due to the rarity of this condition, the exact prevalence of urticarial vasculitis is unclear. One study conducted in Sweden estimated an annual incidence of 0.7% with a point prevalence of 9.5 per million as of December 2015 (Sjöwall et al., 2018). In patients presenting with chronic urticarial lesions, prevalence is estimated to range from 2% to 20%, and approximately 5% when the histologic criteria of leukocytoclastic vasculitis are met (O’Donnell and Black, 1995; Venzor et al., 2002).

Urticarial vasculitis condition more commonly affects women and peaks in frequency during the fourth and sixth decades of life (Jachiet et al., 2015; Sjöwall et al., 2018). Urticarial vasculitis very rarely affects children and even less frequently affects infants, with only two reported cases of the latter in the literature (Kaur and Thami, 2003; Koch et al., 2008).

Pathogenesis

Urticarial vasculitis is thought to be immune-complex mediated and as such is classified as a type III hypersensitivity reaction (Mehregan and Gibson, 1998). Antibodies complex with antigens, which may be autologous or of exogenous origin, and activate complement through the classical pathway. C3a and C5a are then generated, inducing mast-cell degranulation and the release of chemokines and cytokines (Venzor et al., 2002). This process may also explain the angioedema and true urticaria lesions lasting <24 hours that occur in approximately 50% of affected patients. These lesions are distinct from and should not be conflated with the hallmark lesions of urticarial vasculitis, which last >24 hours. Furthermore, proteolytic enzymes are released from neutrophils, and the sum of these changes leads to the characteristic tissue damage and edema in urticarial vasculitis. Eosinophils may also play a role in the pathogenesis of this disease because the infiltration of these cells is significantly greater than that seen in urticaria (Kamyab et al., 2019). Lastly, a role of interleukin 1 (IL-1) has been suggested given the utility of IL-1 inhibitors in the treatment of this disease (Bettuzzi et al., 2019; Krause et al., 2013).

The cause of urticarial vasculitis is often unknown, but there have been numerous reports of cases triggered by drugs, infections, autoimmune connective disease, myelodysplastic disorders, or malignancy. Some drugs that have been implicated include cimetidine, diltiazem, potassium iodide, fluoxetine, nonsteroidal anti-inflammatory drugs, methotrexate, telmisartan, enalapril, levetiracetam, and over-the-counter diet pills (Borcea and Greaves, 2000; Cherrez et al., 2015; Cicek et al., 2008; Koregol et al., 2015; Mahajan et al., 2015; Mangal and Kumaran, 2014). Of note, systemic medications may be over-implicated as the cause of urticarial vasculitis. For example, many patients start methotrexate, which takes 4 to 8 weeks to see full benefit, while simultaneously stopping prednisone, a regimen change that is associated with a rapid rebound of underlying diseases.

Many cases are also a manifestation of underlying infections; associations with streptococcus, tuberculosis, hepatitis B and C, Epstein–Barr virus, mycoplasma, COVID-19, influenza A/H1N1, trichomoniasis, and Lyme disease have all been reported (Baigrie et al., 2020; de Perosanz-Lobo et al., 2020; Gökçe et al., 2020; Kim et al., 2010; Kolkhir et al., 2019; Nasiri et al., 2020; Scott et al., 2014; Tsai et al., 2018). Links between urticarial vasculitis and various autoimmune diseases, including systemic lupus erythematosus, Sjögren’s syndrome, rheumatoid arthritis, inflammatory bowel disease, Schnitzler’s syndrome, and Muckle–Wells syndrome, similarly have been suggested (Baigrie et al., 2020; Swaminath et al., 2011; Tsai et al., 2018). In fact, urticarial vasculi-

Fig. 1. Diagnostic algorithm for evaluation of urticarial lesions.
tis can fulfill many diagnostic criteria for systemic lupus erythematosus, such that some authors have proposed a continuum of disease between these entities (Dincy et al., 2008). Urticarial vasculitis has also been reported to occur in patients with immunoglobulin (Ig) G-4 related diseases (Takao et al., 2016; Tokura et al., 2014; Wakamatsu et al., 2011).

In addition, urticarial vasculitis can manifest as a paraneoplastic finding. The associated malignancies in the literature include multiple myeloma, myelodysplastic syndrome, colonic adenocarcinoma, signet ring cell carcinoma, renal carcinoma, non-Hodgkin lymphoma, metastatic teratoma from testicular tumor, and chronic lymphocytic leukemia (Ducarme et al., 2003; Jachiet et al., 2018; Kassim et al., 2015; Shah et al., 2007; Wilson et al., 2002; Younis, 2018). Cold contact was noted in some reports to produce leukocytoclastic vasculitis locally at the site of application; however, cryoglobulinopathies can be associated with urticarial vasculitis, and cryoglobulins should be evaluated in such cases (Pérez-Bustillo and Sánchez-Sambucety, 2012).

Environmental exposures appear to play a significant role in the development and progression of this disease, but some reports have pointed to a genetic component as well. Familial cases of hypocomplementemic urticarial vasculitis syndrome, a more severe and systemic form of urticarial vasculitis, have been documented in a pair of identical twins, as well as among three siblings (Ozçakar et al., 2013a; Wisnieski et al., 1994). One genemapping study revealed an association between such cases and a homozygous frameshift mutation in DNASE1L3, which encodes for a protein in the deoxyribonuclease I family (Ozçakar et al., 2013b).

Clinical presentation

Cutaneous findings

Patients most often present with classical indurated wheals that can be difficult to distinguish clinically from chronic idiopathic urticaria (Fig. 2). However, unlike the latter, which is characterized by lesions that generally resolve within 2 to 8 hours and by definition within 24 hours, lesions of urticarial vasculitis last >24 h, often persisting for several days, and may leave behind a residual ecchymotic hyperpigmentation that is not seen in chronic idiopathic urticaria. These wheals are often non- or only partially blanchable with a central dark-red or brown macule. Additionally, individual urticarial vasculitis lesions tend to range from 0.5 to 5 cm in diameter, whereas those seen in true urticaria can coalesce and become very large (>10 cm) or serpiginous. When present, symptoms tend to be described as painful and/or burning, as opposed to true urticaria, which is more often frankly pruritic (Wisnieski, 2000).

Recent reports demonstrate the usefulness of dermoscopy in evaluating lesions because the extent of vascular necrosis is usually small enough where frank purpura is not apparent, thus requiring the usage of magnification for accurate visualization (Dahl, 1994; García-García et al., 2020). Episettes of urticarial vasculitis persist on average from 4 to 8 weeks and resolve within 1 year in 30% to 40% of patients (Koç et al., 2017; Zuberbier and Maurer, 2014). Chronic versions that last months to years have also been described (Koç et al., 2017).

Laboratory findings

An elevated erythrocyte sedimentation rate and hypocomplementemia are the most common laboratory abnormalities observed in urticarial vasculitis. Unlike erythrocyte sedimentation rate, which is generally nonspecific, hypocomplementemia is a useful predictor for this disease (Venzor et al., 2002; Zuberbier and Maurer, 2014). Patients with normal complement levels are classified as having normocomplementemic urticarial vasculitis and generally have a better prognosis with few systemic complications. Patients with depressed levels of complement have a higher risk of complications and are either classified as having hypocomplementemic urticarial vasculitis when systemic involvement is little to none or hypocomplementemic urticarial vasculitis syndrome when systemic involvement is significant (Buck et al., 2012). Notably, hypocomplementemia is often indicative of an underlying systemic disease (Chang and Carr, 2007). Circulating immune complexes and autoantibodies involved in the disease process can also be detected, including anti C1q, which is found in 50% of patients with hypocomplementemic urticarial vasculitis and 100% of patients with hypocomplementemic urticarial vasculitis syndrome (Damman et al., 2020). In addition, some studies have shown an elevation in antithyroid antibodies at a prevalence higher than in chronic spontaneous urticaria, although the relevance of this finding remains unclear (Cherrez-Ojeda et al., 2019).

Other appropriate laboratory tests include complete blood count, chemistry profile, creatinine, hepatitis and liver studies, urinalysis, renal function tests, and antinuclear antibody (Koç et al., 2017; Zuberbier and Maurer, 2014). Anti-DNase B may be a useful metric as well because postinfectious leukocytoclastic vasculitis can occur and, when present, is most often due to a streptococcal infection (Baigrie et al., 2020). A chest x-ray and pulmonary function tests are also recommended in patients demonstrating relevant symptoms because pulmonary involvement is the leading cause of mortality in this disease (Chang and Carr, 2007). Serum levels of C-reactive protein may also be elevated (Criado et al., 2013; Roy et al., 2013). Additionally, antinuclear antibodies are detectable in approximately half of patients with hypocomplementemic urticarial vasculitis, which further supports the speculation that the severe end of the urticarial vasculitis disease spectrum overlaps with and meets many criteria for lupus (Dincy et al., 2008; Jachiet et al., 2015).
Systemic involvement

In addition to cutaneous abnormalities, patients with urticarial vasculitis can exhibit symptoms in multiple organ systems, which result in a myriad of systemic findings. Overall, these tend to occur more frequently in hypocomplementemic patients or among those expressing anti-C1q antibodies (Jachiet et al., 2015). Musculoskeletal symptoms, including arthralgias and myalgias, are the most common extracutaneous finding in patients (Koç et al., 2017; Kolkhir et al., 2020).

Renal involvement is estimated to occur in approximately 20% of patients with hypocomplementemic urticarial vasculitis, but this figure can range from 9% to 60% depending on the case series used for analysis. Patients most frequently show evidence of glomerular impairment, primarily resulting from membranoproliferative glomerulonephritis (Boyer et al., 2020). Other observed renal histologies include extracapillary, extramembranous, mesangial, crescentic, focal-proliferative, and segmental hyalinosis glomerulonephritis (AliHermi et al., 2017). Some patients have diffuse interstitial involvement in association with the aforementioned pathologies, but very rarely do these occur in isolation. The most common renal symptoms are hematuria and proteinuria, with a small minority eventually developing kidney failure requiring dialysis. Fortunately, even patients with extensive kidney involvement maintain a fairly good prognosis (Boyer et al., 2020).

Fig. 3. (A) Papillary edema with perivascular neutrophil-predominant infiltrate associated with leukocytoclastic vasculitis, characterized by karyorrhexis, extravasated erythrocytes, and swelling of endothelial cells. Numerous scattered interstitial eosinophils and mast cells can be seen (hematoxylin and eosin, ×100). (B) Perivascular neutrophils, eosinophils (green arrow) and mast cells (red arrow; hematoxylin and eosin, ×300).

The lungs are another common site of involvement, with approximately 20% to 30% of patients developing chronic obstructive pulmonary disease (COPD; Venzor et al., 2002). This number increases to up to 50% in patients with hypocomplementemic urticarial vasculitis (Zuberbier and Maurer 2014). This finding is most common in young patients who smoke tobacco and is more extensive in patients with hypocomplementemic urticarial vasculitis than would be expected based on the degree of smoking alone (Koç et al., 2017). As such, screening for COPD in afflicted patients is crucial and smoking cessation in all patients who present with this disease is strongly recommended. The exact role urticarial vasculitis plays in the development of COPD remains unclear, but the binding of C1q precipitins to pulmonary alveoli surfactant proteins has been proposed as a contributing factor (Buck et al., 2012).

Pleuritis is another possible involvement, often manifesting as chest pain or shortness of breath (Koç et al., 2017). These types of complications are particularly recalcitrant to treatments, with many patients requiring lung transplantation, and tend to worsen with disease progression. Overall, pulmonary involvement, especially in smokers, is associated with poor short-term vital prognosis and is the leading cause of mortality for urticarial vasculitis (Jara et al., 2009; Raoufi et al., 2016). Upper-respiratory tract afflictions, including laryngeal edema, may occur as well (Alomari et al., 2019).

Gastrointestinal symptoms occur in up to 30% of patients. These commonly manifest as abdominal pain, nausea, vomiting, and diarrhea (Davis and van der Hilst, 2018). Cases of intestinal ischemia secondary to urticarial vasculitis have also been reported (Wong et al., 2016). Ocular inflammation has been observed as well, frequently manifesting as uveitis, most often posterior, episcleritis, and conjunctivitis (Zuberbier and Maurer 2014). These complications occur in approximately 10% of patients and up to 30% of those with hypocomplementemic urticarial vasculitis syndrome (Jachiet et al., 2015). Other rare extracutaneous pathologies include pericarditis, pericardial effusion, pseudotumor cerebri, cranial nerve palsies, and transverse myelitis (Koç et al., 2017).

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).

These classic histopathologic findings of this disease have been shown to last only 24 to 48 hours, making selection of the ideal lesions for biopsy an important clinical endeavor (Koç et al., 2017).
Clinicians should also realize that systemic therapy may influence histologic findings. Some reports suggest histology other than that described herein, but these other findings can be explained by lesions that are too early, too late, or influenced by systemic therapy (Kamyab et al., 2019; Lee et al., 2007).

Although there are characteristic features to look for on histopathologic examination, patients will not necessarily demonstrate all these findings and among those who do, a substantial amount of variation can exist. In mild cases, the infiltrate can be sparse and perivascular with minimal leukocytoclasia and little to no evidence of fibrinoid deposits. This can be quite disparate from more severe cases, where frank leukocytoclasia and fibrin deposition can readily be found (Dincy et al., 2008).

As with disease severity, hypocomplementemia is a useful predictor for histologic features of urticarial vasculitis. In hypocomplementemic urticarial vasculitis, perivascular and interstitial eosinophil and neutrophil infiltration, neutrophil count, erythrocyte extravasation, and nuclear dust formation are all more extensive when compared with findings typical for normocomplementemic urticarial vasculitis (Damman et al., 2020). Some authors also found that eosinophilic predominant infiltrates occurred more often in normocomplementemic patients, whereas those that were neutrophil dominant were more typical of hypocomplementemic patients (Mehregan et al. 1992).

Differential diagnosis

There are several potential differential diagnoses for urticarial vasculitis. As mentioned earlier, chronic idiopathic urticaria can present similarly to urticarial vasculitis, but these two conditions can be distinguished by the disparate duration of their individual lesions. Schnitzler’s syndrome, an autoimmune inflammatory condition, is another possibility to consider. On biopsy, lesions of Schnitzler’s syndrome are similar to urticarial vasculitis in that they have a primarily neutrophilic infiltrate; however, unlike urticarial vasculitis, they do not demonstrate any evidence of vasculitis on histopathologic examination (Dingli and Camilleri, 2015). Other differential diagnoses include Well’s syndrome, erythema migrans, tumid lupus erythematosus, and urticarial multiforme (Table 1; Emer et al., 2013).

Table 1

| Differential diagnosis          | Features                                                                 |
|--------------------------------|--------------------------------------------------------------------------|
| Chronic idiopathic urticaria   | Individual lesions lasting <24 hours                                      |
| Well’s syndrome                | Usually a small number of lesions with intensively eosinophilic infiltrate on histology |
| Erythema migrans               | Annular erythema and other features of Lyme borreliosis                  |
| Tumid lupus erythematosus      | Annular plaques on sun-exposed areas that are fixed and have a characteristic histology |
| Urticarial phase of bullous pemphigoid | Evolves to characteristic blistering lesions with diagnostic routine microscopic and immunomicroscopic changes |
| Schnitzler syndrome            | Chronic urticarial rash showing neutrophilic dermal infiltrate on histology; no evidence of vasculitis present |
| Urticarial multiforme           | Classic target lesion with violaceous or dusky center that may blister   |

Usage of corticosteroids is often necessary for control of cutaneous or systemic symptoms but is generally reserved for moderate-to-severe cases or when other first-line treatments have failed. This therapy is effective in resolving cutaneous disease in approximately 80% of patients and has been shown to significantly decrease joint, ocular, gastrointestinal, and pulmonary symptoms (Kolkhir et al., 2019). Patients have also exhibited immunological changes, including depression of inflammatory markers and increase in complement levels, which correlate with cutaneous improvement (Jachiet et al., 2018). However, usage of glucocorticoids comes with its own set of challenges, and many patients experience a relapse in symptoms when treatment is discontinued.

Several biologic medications have also been promising for the treatment of urticarial vasculitis and are especially useful in refractory cases when conventional avenues of therapy have been exhausted. Rituximab (anti-CD20), which is often paired with corticosteroids, hydrochloroquine, or cyclosporine, has been shown to yield higher response rates and is associated with an increased time to treatment failure when compared with corticosteroids and conventional immunosuppressives (Jachiet et al., 2018;
Swaminath et al., 2011). Omalizumab (anti-IgE) is another option, and several recent case reports have supported the efficacy of this medication in the treatment of urticarial vasculitis (Cherrez-Ojeda et al, 2018; Nucera et al., 2017). Omalizumab seems to be especially effective in treating normocomplementemic patients, with investigations into its utility for hypocomplementemic urticarial vasculitis yielding inconsistent results (de Brito et al., 2018). Successful usage of this medication in conjunction with methotrexate has also been documented (Garbayo-Salmons et al., 2020). The mechanism of action of omalizumab and its role in the pathogenesis of urticarial vasculitis has yet to be elucidated and may or may not be the same mechanism by which it exerts its efficacy in the treatment of chronic urticaria (Fueyo-Casado et al., 2017).

Usage of IL-1 inhibitors (anakinra and canakinumab) as treatment has also shown promising results. Studies have demonstrated a concurrent improvement in both serological markers and clinical disease in response to this therapy. The efficacy of these medications has led some authors to propose a role of IL-1 in the development of urticarial vasculitis, but the exact link remains unclear (Bettuzzi et al., 2019; Botsios et al., 2007; Krause et al., 2013). Intravenous immunoglobulin has also been effective in treating cutaneous and systemic symptoms in some patients (Staubach-Renz et al., 2007; Yamazaki-Nakashimada et al., 2009; Table 2).

**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 complicating 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.

**Conflicts of interest**

None.

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None.

**Study approval**

The author(s) confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies.

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