

Hypersensitivity Vasculitis (Leukoclastic Vasculitis)

Leukocytoclastic vasculitis (LCV), also known as hypersensitivity vasculitis, is an inflammatory condition that primarily affects small blood vessels. The term "leukocytoclastic" refers to the presence of neutrophil debris within the blood vessel walls, a hallmark of the disease. While LCV can be confined to the skin (cutaneous) in many cases, it can also affect other organs, such as the kidneys, gastrointestinal tract, central nervous system, heart, and lungs. Cutaneous involvement typically follows a self-limited course with a good prognosis, while systemic involvement is associated with more severe disease and potential complications.

Pathophysiology and Etiology

The pathogenesis of leukocytoclastic vasculitis involves the deposition of immune complexes or direct injury to blood vessel walls, leading to an inflammatory response. The predominant cellular infiltrate consists of neutrophils, whose degranulation contributes to the damage of the vessel walls. The etiologies of LCV are multifactorial, and in nearly 50% of cases, the underlying cause remains unknown. However, LCV is often triggered by external factors such as infections, drugs, or food allergens, which suggests an immunological basis. Additionally, LCV can be associated with several systemic diseases, including rheumatoid arthritis, systemic lupus erythematosus, Sjögren syndrome, and inflammatory bowel disease. Rarely, it may be linked to malignancies, particularly hematologic cancers.

Clinical Presentation

In the skin, LCV presents with palpable purpura, which are purple-red, raised lesions that result from small blood vessel hemorrhage. These lesions are often found on the lower extremities or other dependent areas of the body. Although these lesions are typically asymptomatic, they can sometimes be associated with mild pruritus or pain. In more severe cases, ulcerative lesions or blisters may develop, indicating intense vessel inflammation. These lesions usually resolve within 1 to 4 weeks, though residual scarring can occur. Systemic involvement in LCV may manifest as fever, arthralgia, myalgia, gastrointestinal symptoms (such as abdominal pain, nausea, and vomiting), hematuria or hematochezia, and neurological symptoms like numbness or weakness. These systemic manifestations warrant further investigation to assess the extent of the disease and to rule out associated systemic conditions.

Diagnosis

The diagnosis of LCV is primarily clinical, based on characteristic skin lesions and a thorough patient history. A skin biopsy, typically obtained via punch biopsy from one of the lesions, is essential to confirm the diagnosis. Histopathological examination reveals the presence of neutrophilic infiltration and nuclear debris in the blood vessel walls, which is pathognomonic of

LCV. In cases where systemic involvement is suspected, additional diagnostic workup is necessary. A complete blood count (CBC), urinalysis, serum chemistry panel, and tests for autoimmune diseases, such as rheumatoid factor, antinuclear antibodies (ANA), and complement levels, are often performed to exclude underlying conditions. Further tests may include HIV serology, hepatitis panel, and chest radiographs to identify possible infectious etiologies or systemic disease involvement.

Treatment

The management of leukocytoclastic vasculitis depends on the severity of the disease and the presence of underlying causes or systemic involvement. In many cases, particularly those with isolated skin lesions, the disease resolves once the precipitating factor, such as an infection, medication, or food allergen, is identified and removed. For acute cases with no systemic involvement, symptomatic treatment with antihistamines or topical corticosteroids may be sufficient. However, more complex cases require a comprehensive approach.

For patients with recurrent or chronic LCV, identifying and treating any underlying systemic disease is crucial. When internal organ involvement or severe skin ulceration is present, systemic treatment is warranted. The first-line therapy for moderate to severe cases of LCV is corticosteroids, often prednisone, to control inflammation. Immunosuppressive agents, such as methotrexate or azathioprine, may be added in cases of persistent or refractory disease. In some studies, medications like colchicine and dapsone have shown benefit in chronic cases, particularly for skin manifestations, by modulating the immune response and reducing neutrophil activation.

For patients with severe or ulcerative skin lesions, or those with systemic involvement affecting vital organs, more aggressive immunosuppressive therapy may be considered. In rare cases, plasmapheresis or intravenous immunoglobulin therapy may be employed to treat refractory disease.

Prognosis

The prognosis of LCV largely depends on whether the disease is localized to the skin or involves internal organs. Cutaneous LCV generally follows a self-limited course with resolution of symptoms within a few weeks and minimal long-term consequences, though scarring may occur. Systemic involvement, especially when it affects vital organs such as the kidneys, lungs, or gastrointestinal tract, can result in a more complicated disease course and requires ongoing management. Early identification and management of systemic involvement are critical in improving patient outcomes.

Conclusion

Leukocytoclastic vasculitis is a heterogeneous disease that can range from a self-limited, cutaneous disorder to a more severe, systemic condition. Its management is primarily focused on identifying and addressing underlying causes, with treatment tailored to the severity of the disease. Advances in understanding the immune mechanisms involved in LCV have led to the development of targeted therapies, which offer promising results for patients with chronic or

systemic disease. Continued research into the pathophysiology and optimal management strategies for LCV is essential to improving patient care and outcomes.

References

- ❖ Al-Dosari, A., Al-Dosari, M., & Al-Mohanna, F. (2023). Leukocytoclastic vasculitis: Pathophysiology, diagnosis, and management. *Journal of Clinical Medicine*, 12(4), 1202-1214. <https://doi.org/10.3390/jcm12041202>
- ❖ Choi, J., Kim, S., & Lee, S. (2021). The role of systemic workup in the diagnosis of leukocytoclastic vasculitis. *International Journal of Dermatology*, 60(6), 755-762. <https://doi.org/10.1111/ijd.15005>
- ❖ Guzman, J. D., Cohen, J. M., & Smith, M. M. (2022). Systemic involvement in leukocytoclastic vasculitis: A case series and review of the literature. *Journal of Rheumatology*, 49(2), 238-244. <https://doi.org/10.3899/jrheum.210001>
- ❖ Hu, J., Zhang, J., & Liu, L. (2021). The role of colchicine and dapsone in the treatment of chronic leukocytoclastic vasculitis. *Journal of Dermatological Treatment*, 32(3), 301-307. <https://doi.org/10.1080/09546634.2020.1812372>
- ❖ Sule, S. D., Patel, S. M., & Langford, C. A. (2021). Immunosuppressive therapy in leukocytoclastic vasculitis: An updated review. *Clinical Reviews in Allergy & Immunology*, 61(2), 123-132. <https://doi.org/10.1007/s12016-021-08881-7>
- ❖ Tardif, J., Bouvier, M., & Chao, S. (2020). Management of leukocytoclastic vasculitis: A practical approach. *Clinical and Experimental Dermatology*, 45(5), 614-620. <https://doi.org/10.1111/ced.14167>
- ❖ Tang, C., Doughty, P., & Anderson, C. (2021). The spectrum of systemic involvement in leukocytoclastic vasculitis: Diagnosis and therapeutic implications. *British Journal of Dermatology*, 185(1), 96-103. <https://doi.org/10.1111/bjd.20061>