Chapter 2

CUTANEOUS VASCULITIS

Adem ERTÜRK[1]

INTRODUCTION

Vasculitis is a condition characterized by partial or total occlusion of the vessel lumen, aneurysm formation, and inflammation of the vessel wall, resulting in impaired distal organ function. As a result of thinning of the vessel wall due to inflamed cells of the vessel wall, there is an increase in vascular permeability, vessel wall rupture, and hemorrhage in the affected organ (palpable purpura, pulmonary hemorrhage). There is narrowing or occlusion of the vessel lumen due to vascular intimal proliferation and intraluminal thrombus formation, and signs of ischemia or infarction develop in the affected organ, such as necrotic skin ulcers, mononeuritis multiplex, and major organ infarction. In cutaneous vasculitis, systemic vasculitis findings such as fever, weakness, fatigue, weight loss, widespread muscle pain, arthralgia, and oligoarthritis may also be seen (1).

In cases where cutaneous vasculitis is suspected, answers to the following questions should be sought:

-Is there a vasculitis-like (mimic) condition?

-Is there an underlying secondary cause?

-Is there organ involvement other than cutaneous?

-How can a diagnosis of vasculitis be confirmed?

-What type of vasculitis is present?

Cutaneous vasculitis typically targets the dermal and subcutaneous vessels of small or medium size, encompassing a diverse spectrum of conditions that vary in their extent, ranging from localized lesions to systemic manifestations (2).

The first International Chapel Hill Consensus Conference (CHCC) on the nomenclature of systemic vasculitides was held in 1994 to establish uniform terminology and definitions. In 2012, a revised version was developed (Table 1) (3,4).

[1] Asst. Prof. Dr. Afyonkarahisar Health Sciences University Faculty of Medicine Division of Rheumatology, Department of Internal Medicine, drademerturk@hotmail.com, ORCID iD: 0000-0001-8882-0692

CONCLUSION

Since skin lesions occurring in systemic and cutaneous vasculitis may show similar clinical and pathological features, systemic involvement should be excluded before diagnosing cutaneous vasculitis. As with systemic vasculitides, cutaneous vasculitides' causes are heterogeneous and generally classified as primary (idiopathic) and secondary. Infections, drugs, autoimmune diseases and malignancies are the most important secondary causes. As in systemic forms, cutaneous vasculitis is diagnosed by presenting clinicopathological and laboratory findings. The prognosis of vasculitis limited to the skin is generally favourable. However, caution should be exercised in painful, itchy or ulcerated skin lesions and chronic or recurrent cases. Treatment selection should be determined according to the severity of symptoms, potential risks and the patient's comorbidities. Randomized controlled studies on treatment in cutaneous vasculitis are insufficient; additional studies, including multicenter randomized studies, are needed on this subject.

REFERENCES

1. Callen JP, Requena L. Cutaneous vasculitis and panniculitis. In: Hochberg MC, Gravallese EM, Silman AJ (eds.) Rheumatology 7th edition. Philadelphia: Elsevier; 2019. p. 1412– 1421.
2. Chen KR, Carlson JA. Clinical Approach to Cutaneous Vasculitis. *American Journal of Clinical Dermatology*. 2008;9(2): 71–92. doi: 10.2165/00128071-200809020-00001
3. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of Systemic Vasculitides. *Arthritis & Rheumatism*. 1994;37(2): 187–192. doi: 10.1002/art.1780370206
4. Jennette JC, Falk RJ, Bacon PA, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis & Rheumatism*. 2013;65(1): 1–11. doi: 10.1002/art.37715
5. Sunderkötter CH, Zelger B, Chen K, et al. Nomenclature of Cutaneous Vasculitis. *Arthritis & Rheumatology*. 2018;70(2): 171–184. doi: 10.1002/art.40375
6. Caproni M, Verdelli A. An update on the nomenclature for cutaneous vasculitis. *Current Opinion in Rheumatology*. 2019;31(1): 46–52. doi: 10.1097/BOR.0000000000000563
7. Goeser MR, Laniosz V, Wetter DA. A Practical Approach to the Diagnosis, Evaluation, and Management of Cutaneous Small-Vessel Vasculitis. *American Journal of Clinical Dermatology*. 2014;15(4): 299–306. doi: 10.1007/s40257-014-0076-6
8. Filosa A, Verdelli A, Bianchi B, et al. Cutaneous vasculitidis: histology and immunofluorescence. *Giornale italiano di dermatologia e venereologia : organo ufficiale, Societa italiana di dermatologia e sifilografia*. 2015;150(2): 183–191.
9. Demirkesen C. Approach to cutaneous vasculitides with special emphasis on small vessel vasculitis: histopathology and direct immunofluorescence. *Current Opinion in Rheumatology*. 2017;29(1): 39–44. doi: 10.1097/BOR.0000000000000346

10. Alberti-Violetti S, Berti E, Marzano A V. Cutaneous and systemic vasculitides in dermatology: a histological perspective. *Italian Journal of Dermatology and Venereology*. 2018;153(2). doi: 10.23736/S0392-0488.18.05886-8
11. Pina T, Blanco R, González-Gay MA. Cutaneous vasculitis: a rheumatologist perspective. *Current allergy and asthma reports*. 2013;13(5): 545-554. doi: 10.1007/s11882-013-0367-7
12. Marzano AV, Raimondo MG, Berti E, et al. Cutaneous Manifestations of ANCA-Associated Small Vessels Vasculitis. *Clinical Reviews in Allergy & Immunology*. 2017;53(3): 428–438. doi: 10.1007/s12016-017-8616-5
13. Jarrot PA, Kaplanski G. Pathogenesis of ANCA-associated vasculitis: An update. *Autoimmunity Reviews*. 2016;15(7): 704–713. doi: 10.1016/j.autrev.2016.03.007
14. Nakazawa D, Masuda S, Tomaru U, et al. Pathogenesis and therapeutic interventions for ANCA-associated vasculitis. *Nature Reviews Rheumatology*. 2019;15(2): 91–101. doi: 10.1038/s41584-018-0145-y
15. Morita TCAB, Criado PR, Criado RFJ, et al. Update on vasculitis: overview and relevant dermatological aspects for the clinical and histopathological diagnosis – Part II. *Anais Brasileiros de Dermatologia*. 2020;95(4): 493–507. doi: 10.1016/j.abd.2020.04.004
16. Ratzinger G, Zelger BG, Carlson JA, et al. Vasculitic wheel - an algorithmic approach to cutaneous vasculitides. *JDDG: Journal der Deutschen Dermatologischen Gesellschaft*. 2015;13(11): 1092–1117. doi: 10.1111/ddg.12859
17. Frumholtz L, Laurent-Roussel S, Lipsker D, et al. Cutaneous Vasculitis: Review on Diagnosis and Clinicopathologic Correlations. *Clinical Reviews in Allergy & Immunology*. 2021;61(2): 181–193. doi: 10.1007/s12016-020-08788-4
18. Lava SAG, Milani GP, Fossali EF, et al. Cutaneous Manifestations of Small-Vessel Leukocytoclastic Vasculitides in Childhood. *Clinical Reviews in Allergy & Immunology*. 2017;53(3): 439–451. doi: 10.1007/s12016-017-8626-3
19. Fraticelli P, Benfaremo D, Gabrielli A. Diagnosis and management of leukocytoclastic vasculitis. *Internal and Emergency Medicine*. 2021;16(4): 831–841. doi: 10.1007/s11739-021-02688-x
20. Marzano AV, Maronese CA, Genovese G, et al. Urticarial vasculitis: Clinical and laboratory findings with a particular emphasis on differential diagnosis. *Journal of Allergy and Clinical Immunology*. 2022;149(4): 1137–1149. doi: 10.1016/j.jaci.2022.02.007
21. Nakamura K, Tsunemi Y, Kaneko F, et al. Mucocutaneous Manifestations of Behçet's Disease. *Frontiers in Medicine*. 2021;7. doi: 10.3389/fmed.2020.613432
22. Espinoza GM, Wheeler J, Temprano KK, et al. Cogan's Syndrome: Clinical Presentations and Update on Treatment. *Current Allergy and Asthma Reports*. 2020;20(9): 46. doi: 10.1007/s11882-020-00945-1
23. Ikeda T. Recent topics related to etiology and clinical manifestations of cutaneous arteritis. *Frontiers in Medicine*. 2022;9. doi: 10.3389/fmed.2022.1022512
24. Turska M, Parada-Turska J. [Cutaneous polyarteritis nodosa]. *Wiadomosci lekarskie (Warsaw, Poland : 1960)*. 2018;71(1 pt 1): 73–77.
25. Karadag O, Jayne DJ. Polyarteritis nodosa revisited: a review of historical approaches, subphenotypes and a research agenda. *Clinical and experimental rheumatology*. 2018;36 Suppl 111(2): 135–142.
26. Morgan AJ, Schwartz RA. Cutaneous polyarteritis nodosa: a comprehensive review. *International Journal of Dermatology*. 2010;49(7): 750–756. doi: 10.1111/j.1365-4632.2010.04522.x

27. Sánchez-Guerrero J, Gutiérrez-Ureña S, Vidaller A, et al. Vasculitis as a paraneoplastic syndrome. Report of 11 cases and review of the literature. *The Journal of rheumatology*. 1990;17(11): 1458–1462.
28. Solans-Laqué R, Bosch-Gil JA, Pérez-Bocanegra C, et al. Paraneoplastic vasculitis in patients with solid tumors: report of 15 cases. *The Journal of rheumatology*. 2008;35(2): 294–304.
29. Fain O, Hamidou M, Cacoub P, et al. Vasculitides associated with malignancies: Analysis of sixty patients. *Arthritis & Rheumatism*. 2007;57(8): 1473–1480. doi: 10.1002/art.23085
30. Khan TA, Cuchacovich R, Espinoza LR, et al. Vasculopathy, hematological, and immune abnormalities associated with levamisole-contaminated cocaine use. *Seminars in arthritis and rheumatism*. 2011;41(3): 445–454. doi: 10.1016/j.semarthrit.2011.04.010.
31. Sandhu JK, Albrecht J, Agnihotri G, et al. Erythema elevatum et diutinum as a systemic disease. *Clinics in Dermatology*. 2019;37(6): 679–683. doi: 10.1016/j.clindermatol.2019.07.028
32. Doktor V, Hadi A, Hadi A, et al. Erythema elevatum diutinum: a case report and review of literature. *International Journal of Dermatology*. 2019;58(4): 408–415. doi: 10.1111/ijd.14169
33. Hasbún C, Moya N, Morales C. Eritema indurado de Bazin como una reactivación de una tuberculosis latente en una paciente con tratamiento biológico anti-factor de necrosis tumoral. *Revista chilena de infectología*. 2022;39(1): 100–102. doi: 10.4067/S0716-10182022000100100
34. Fonseca Aizpuru EM, Rodríguez Ávila EE, Arias Miranda I, et al. Revisión de 11 casos de eritema indurado en un hospital de segundo nivel. *Anales de Medicina Interna*. 2008;25(7). doi: 10.4321/S0212-71992008000700018
35. Corrà A, Verdelli A, Mariotti EB, et al. Cutaneous vasculitis: Lessons from COVID-19 and COVID-19 vaccination. *Frontiers in Medicine*. 2022;9. doi: 10.3389/fmed.2022.1013846
36. Maronese CA, Zelin E, Avallone G, et al. Cutaneous vasculitis and vasculopathy in the era of COVID-19 pandemic. *Frontiers in Medicine*. 2022;9. doi: 10.3389/fmed.2022.996288
37. Genovese G, Moltrasio C, Berti E, et al. Skin Manifestations Associated with COVID-19: Current Knowledge and Future Perspectives. *Dermatology*. 2021;237(1): 1–12. doi: 10.1159/000512932
38. McGonagle D, Bridgewood C, Ramanan AV, et al. COVID-19 vasculitis and novel vasculitis mimics. *The Lancet Rheumatology*. 2021;3(3): e224–e233. doi: 10.1016/S2665-9913(20)30420-3
39. Fraticelli P, Benfaremo D, Gabrielli A. Diagnosis and management of leukocytoclastic vasculitis. *Internal and Emergency Medicine*. 2021;16(4): 831–841. doi: 10.1007/s11739-021-02688-x
40. Sunderkötter C, Bonsmann G, Sindrilaru A, et al. Management of leukocytoclastic vasculitis. *Journal of Dermatological Treatment*. 2005;16(4): 193–206. doi: 10.1080/09546630500277971
41. Tsampau D, Buggiani G, Hercogova J, et al. Cutaneous necrotizing vasculitis: a rational therapeutic approach. *Dermatologic Therapy*. 2012;25(4): 335–339. doi: 10.1111/j.1529-8019.2012.01480.x

42. Russell JP, Gibson LE. Primary cutaneous small vessel vasculitis: approach to diagnosis and treatment. *International Journal of Dermatology*. 2006;45(1): 3–13. doi: 10.1111/j.1365-4632.2005.02898.x
43. Martinez-Taboada MVM, Blanco MR, Garcia-Fuentes MM, et al. Clinical Features and Outcome of 95 Patients With Hypersensitivity Vasculitis. *The American Journal of Medicine*. 1997;102(2): 186–191. doi: 10.1016/S0002-9343(96)00405-6
44. Lotti T, Ghersetich I, Comacchi C, et al. Cutaneous small-vessel vasculitis. *Journal of the American Academy of Dermatology*. 1998;39(5): 667–690. doi: 10.1016/S0190-9622(98)70039-8
45. Cupps TR. Chronic, Recurrent Small-Vessel Cutaneous Vasculitis. *JAMA*. 1982;247(14): 1994. doi: 10.1001/jama.1982.03320390056044
46. Caplan A, Fett N, Rosenbach M, et al. Prevention and management of glucocorticoid-induced side effects: A comprehensive review. *Journal of the American Academy of Dermatology*. 2017;76(1): 1–9. doi: 10.1016/j.jaad.2016.01.062
47. Callen JP. Colchicine is effective in controlling chronic cutaneous leukocytoclastic vasculitis. *Journal of the American Academy of Dermatology*. 1985;13(2): 193–200. doi: 10.1016/S0190-9622(85)70158-2
48. Sais G, Vidaller A, Jucglà A, et al. Colchicine in the treatment of cutaneous leukocytoclastic vasculitis. Results of a prospective, randomized controlled trial. *Archives of dermatology*. 1995;131(12): 1399–1402.
49. Micheletti RG, Werth VP. Small Vessel Vasculitis of the Skin. *Rheumatic Disease Clinics of North America*. 2015;41(1): 21–32. doi: 10.1016/j.rdc.2014.09.006
50. Micheletti RG, Pagnoux C. Management of cutaneous vasculitis. *La Presse Médicale*. 2020;49(3): 104033. doi: 10.1016/j.lpm.2020.104033
51. Ghazanfar MN, Thomsen SF. Omalizumab for Urticarial Vasculitis: Case Report and Review of the Literature. *Case reports in dermatological medicine*. 2015;2015: 576893. doi: 10.1155/2015/576893