Brief Report:

Patterns of Vasculitis in Dermatological Practice

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Abstract

Background: Vasculitis is a rare disorder with a wide range of systemic and cutaneous manifestations. This study was aimed to identify the clinico-demographic characteristics of different patterns of vasculitis and determine their frequency in dermatological practice. **Methods:** This descriptive cross-sectional study was conducted on 48 cases of vasculitis diagnosed on the basis of clinical, histological, and laboratory parameters from January 2017 to December 2022 in the Department of Dermatology, BSMMU. Specific causes, types, and patterns of systemic and cutaneous manifestations were recorded and analyzed. **Results:** The seven diagnoses of vasculitis were cutaneous small vessel vasculitis (CSVV) (41.7%), polyarteritis nodosa (PAN) (14.6%), IgA vasculitis (IgAV) (12.5%), granulomatosis with polyangiitis (GPA) (12.5%), livedoid vasculopathy (8.3%), eosinophilic granulomatosis with polyangiitis (EGPA) (6.3%), and urticarial vasculitis (4.2%). Cutaneous findings included palpable purpura, urticaria subcutaneous nodule, livedo reticularis, erythematous plaque, hemorrhagic vesicle, and ulcer. Extracutaneous findings included fever, fatigue, weight loss, arthralgia, myalgia, abdominal pain, bloody stool, shortness of breath, wheezing, cough, chest pain, hemoptysis, sinusitis, nasal discharge, and hearing loss. **Conclusion:** The current study would be very helpful in the diagnosis and monitoring of patients with vasculitis having various cutaneous presentations, considering the future possibilities of other systemic involvement.

Keywords: Cutaneous small vessel vasculitis, Polyarteritis nodosa, IgA vasculitis, Granulomatosis with polyangiitis.

Introduction

Vasculitis is a relatively uncommon disorder, with a reported annual incidence of 40–54 cases per million people [1]. It is defined as the inflammation of blood vessels that can affect any part of the body. When vasculitis affects capillaries, post-capillary venules, and muscular arterioles in the superficial and mid dermis (<50 µm in diameter) [2], it is known as primary cuteneous vasculitis. Although approximately half of all cases of cutaneous vasculitis are skin-confined and self-limited, they can also appear as initial manifestations of systemic vasculitis or later progress to systemic vasculitis [3].

Preliminary skin presentation of vasculitis includes variable and active types of discoloration, swelling, bleeding, and necrosis. Physical signs of vasculitis include urticaria, purpura, purpuric papules, infiltrated erythema, ulcer, infarct, livedo reticularis, and nodules that affect the skin with varying intensities, depths, and distributions[4]. The classification of vasculitides has been a confusing and debate-provoking topic over the last half-century.[5] The 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides also has limitations [6]. Marzano et al. adopted a recently proposed working classification that focused on cutaneous small vessel vasculitis (CSVV), IgA vasculitis, urticarial vasculitis, and a variety of cutaneous manifestations that may be observed in the course of the main systemic vasculitides, such as polyarteritis nodosa (PAN), granulomatosis with polyangiitis (GPA), and eosinophilic granulomatosis with polyangiitis (EGPA). Livedoid vasculopathy, a cutaneous entity of non-frankly vasculitic origin, was also included in this classification [5].

CSVV is the most common form of vasculitis in dermatology that affects post-capillary venules, also known by the histologic term leukocytoclastic vasculitis

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(LCV) [6]. Palpable purpura is a hallmark of this disease [7]. Urticarial vasculitis presents histologically as fixed urticarial lesions with vasculitis [5]. IgA vasculitis is characterized by purpura, arthralgia, and abdominal and renal disease. IgA, C3, and fibrin deposition have been demonstrated in biopsies of both involved and uninvolved skin using immunofluorescence techniques [8]. Livedoid vasculopathy is a chronic, recurrent, painful disorder characterized by purpuric macules, papules, hemorrhagic crust, and irregular superficial ulcers around the ankle that heal with an atrophic scar [9].

Polyarteritis nodosa is a systemic necrotizing vasculitis that predominantly affects medium-sized muscular arteries, with an annual incidence rate of 0.9–8.0 per million and a prevalence of 31 per million [10]. The skin is involved in 50% of patients with systemic PAN [11]. ANCA-associated vasculitis (AAV) is a group of vasculitis predominantly affecting small vessels [12]. Cutaneous findings occur in 45% of patients with GPA and two-thirds of patients with EGPA [13,14].

From the perspective of dermatologists, it is always a major concern whether vasculitis is exclusively confined to the skin or a manifestation of more widespread systemic involvement. Identifying the pathognomonic cutaneous features of a certain type of vasculitis and further confirmation by histopathology, imaging, and relevant laboratory tests are crucial for proper diagnosis and management. The current study aimed to identify the variable clinical characteristics of different types of vasculitis and determine their frequency. This study was an approach used by dermatologists to work on both the cutaneous and systemic profiles of vasculitis. It may guide future dermatologists in the diagnosis, treatment, and proper referral of patients with vasculitis.

Methods

This descriptive cross-sectional study was conducted from January 2017 to December 2021 on 48 patients at the Department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka. Patients were conveniently selected from the Department of Dermatology and Rheumatology (who were referred for cutaneous evaluation and skin biopsy). All patients underwent lesional skin biopsy and histopathology reports with a direct immunofluorescence test (DIF) were provided by the Pathology Department. The diagnosis of vasculitis was ascertained by dermatologists based on history, clinical examinations, histopathology with direct immunofluorescence test (DIF), urinalysis, ANCA, routine laboratory parameters, and radiological findings [6].

Age, sex, disease duration, cutaneous and systemic features, and laboratory investigations were recorded by using a standard and pretested semi-structured questionnaire. The patients were plotted into seven patterns of vasculitis (CSVV, PAN, IgA vasculitis, GPA, Livedoid vasculopathy, EGPA and Urticarial vasculitis) according to the proposed working classification.5All of which were described based on age, sex, disease duration, and cutaneous and systemic presentation. Statistical analysis was performed using the Statistical Package for the Social Science (SPSS) software version 26.0 for Windows (SPSS Inc., Chicago, Illinois, USA). Quantitative variables (frequency of vasculitis, age, and duration) were expressed as mean ± SD. Qualitative data (sex, cutaneous, and extracutaneous presentation) were expressed as frequencies and percentages. Informed written consent to participate in the study has been obtained from all adult participants and all underaged participants' parent/legal guardian/next of kin. Patient privacy, safety, and proper treatment were ensured. Prior approval was obtained from the Institutional Review Board (IRB) (IRB-number-BSMMU/2016/10817) of BSMMU.

Result

The mean age of all study participants was 29.0±12.2 years and the median age was 30 years. The majority (41.7%) were diagnosed with CSVV, followed by PAN, IgA vasculitis, EGPA, Livedoid vasculopathy, and Urticarial Vasculitis (Fig. 1). The mean ages in years were 24±9.8 (CSVV), 26±9.8 (PAN), 21±10.1 (IgA vasculitis), 46±7.1 (GPA), 32±6.5 (Livedoid vasculopathy), 42±17.6 (EGPA) and 27±3.5 years (urticarial vasculitis) respectively.CSVV, EGPA, Livedoid vasculopathy, and urticarial vasculitis were more prevalent in females whereas PAN and GPA were common in males. The average disease duration was higher (64±54.1 years) in the EGPA and lower (1±0.3 months) in the urticarial vasculitis group (Table 1).

Table 1: Distribution of vasculitis stratified by age, gender and disease duration (n=48)

Characteristics	Age in years (Mean SD)	Gender		Duration of disease in months (Mean SD)
		Male %	Female %	
CSVV*	24 (9.8)	45.0	55.0	8 (13.8)
PAN*	26 (9.8)	57.1	42.9	15 (17.6)
IgA vasculitis	21 (10.1)	50.0	50.0	3 (4.5)
GPA*	46 (7.0)	100.0	0.0	24 (31.6)
Levedoid Vasculopathy	32 (6.4)	25.0	75.0	19 (21.4)
EGPA*	42 (17.6)	33.3	66.7	64 (54.1)
Urticarial Vasculitis	27 (3.5)	0.0	100.0	1 (0.3)
Age of the Study Participants	Mean (SD) =29.0 (12.2) Years Median=30 Years			

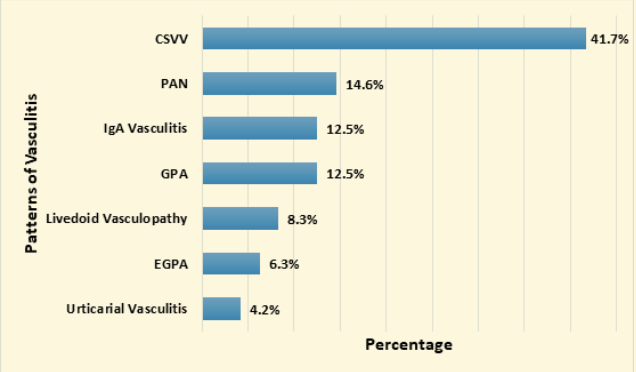

fig 1. Patterns of Vasculitis among the study participants (n=48)

*CSVV= Cutaneous Small Vessel Vasculitis; PAN= Polyarteritis Nodosa; GPA= Granulomatosis with Polyangiitis; EGPA= Eosinophilic Granulomatosis with Polyangiitis

Palpable purpura was present in all vasculitis patterns with variable frequencies. Urticaria was found in 100% of patients with urticarial vasculitis; a subcutaneous nodule was palpated in PAN (57.1%) and CSVV (10%); livedo reticularis was noticed in PAN (71.4%) and Livedoid Vasculopathy (25%); erythematous plaque was found in livedoid vasculopathy (75%), GPA(50%), and PAN (28.6%); hemorrhagic vesicles were observed in livedoid vasculopathy (50%) and PAN (14.3%); ulcers were found in all patients with livedoid vasculopathy and GPA, and in 85.7% of PAN patients. These results were statistically significant (Table 2).

Constitutional symptoms and involvement of the musculoskeletal system were observed in all patterns of vasculitis, with variable frequencies. GIT involvement was commonly seen in IgA vasculitis (66.7%); the nervous system was mostly (71.4%) affected by patients presenting with PAN; respiratory system involvement was a common presentation of EGPA (66.7%) and GPA (50%); sinusitis, nasal discharge, and hearing loss were found in 66.7% of patients with EGPA;eye changes were detected in 100% of patients with EGPA and 85.7% of patients with PAN. These results were statistically significant (Table 2).

Table 2: Clinical presentation of different patterns of vasculitis (n=48)

	CSVV*	PAN*	IgA vasculitis	GPA*	Leveoid Vasculo-pathy	EGPA*	Urticarial Vasculitis	P value
Cutaneous Presentation	%	%	%	%	%	%	%	
Palpable Purpura	95.0	57.1	100.0	50	25	66.7	100.0	0.01**
Urticaria	10.0	0.0	0.0	0.0	0.0	33.3	100.0	0.00**
Sub- cutaneous Nodule	10.0	57.1	0.0	0.0	0.0	0.0	0.0	0.01**
Livedo reticularis	5.0	71.4	0.0	0.0	25.0	0.0	0.0	0.00**
Erythematous plaque	15.0	28.6	0.0	50	75.0	0.0	0.0	0.04**
Hemorrhagic vesicle	0.0	14.3	0.0	0.0	50.0	0.0	0.0	0.01**
Ulcer	0.0	85.7	16.7	100	100	0.0	0.0	0.00**
Systemic Presentation								
Constitutional Symptoms (Fever, weight loss & fatigue)	20.0	100.0	33.3	83.3	25.0	66.7	100.0	0.00**
Musculoskeletal (Arthralgia & myalgia)	20.0	100.0	50.0	16.7	25.0	33.3	100.0	0.00**
GIT (Abdominal pain & bloody stool)	5.0	28.6	66.7	16.7	0.0	33.3	0.0	0.03**
Nervous System (Numbness, paresthesia, weakness of the limbs & headache)	5.0	71.4	0.0	0.0	25.0	0.0	0.0	0.06
Respiratory (Shortness of breath, wheeze, cough, chest pain & hemoptysis)	15.0	28.6	0.0	50.0	0.0	66.7	0.0	0.00**

*CSVV= Cutaneous Small Vessel Vasculitis; PAN= Polyarteritis Nodosa; GPA= Granulomatosis with Polyangiitis; EGPA= Eosinophilic Granulomatosis with Polyangiitis; **Statistically significant

Discussion

Vasculitis is a rare disorder, and related data are still lacking from various parts of the world, including the Indian subcontinent [15]. In the current study, CSVV was the leading variant (41.7%) of vasculitis affecting young adults, predominantly females. Bilateral palpable purpura were frequently observed (95%). The clinico-demographic findings of CSVV found in this study were in agreement with previous studies [5,6,7]. A multi-ethnic cohort study from the UK showed that the incidence of IgA vasculitis was higher in young people of Indian subcontinent origin than in Caucasians and Afro-Caribbeans [8]. In this study, patients with IgA vasculitis reflected the same age range, and their cutaneous and systemic characteristics were similar to those in a previous study [8].

Here, urticarial vasculitis is a less frequent variant of vasculitis, affecting mainly women, which is consistent with a recently published article [16]. Urticaria can also be a presenting feature of vasculitides; in the present study, it was observed in urticarial vasculitis, CSVV, and EGPA [17].

The higher frequency of PAN in comparison to GPA and EGPA and their male predominance in this study were a reflection of another previous study [18]. However, the recommended age group in this study was older than that in the present study. Patients with PAN may present with

palpable purpura, tender subcutaneous nodules, livedo reticularis, hemorrhagic bulla and ulcers [19]. These cutaneous findings were also observed in this study.

Livedo reticularis is an important cutaneous feature of PAN and ANCA-associated vasculitis [20]. In the present study, it was identified in CSVV, PAN and Livedoid vasculopathy. Necrotic ulcer is another striking feature of vasculitis, which was detected in the majority of patients with PAN and all patients with GPA in our study [13].Though the presence or absence of any particular cutaneous manifestation is not entirely specific to a particular ANCA type, some distinguishing patterns are common.

Conclusion

Cutaneous small-vessel vasculitis is the most common type of vasculitis observed in dermatological practice. Palpable purpura is the usual cutaneous presentation of every pattern of vasculitis. Urticaria and ulcers are the most common manifestations of urticarial vasculitis and granulomatosis with polyangiitis. Because cutaneous features can be present even before any obvious features of internal involvement, each cutaneous feature should be investigated in depth to reach a final diagnosis of vasculitis.

Statements

This study was conducted in accordance with the principles of the Declaration of Helsinki. The protocol was approved by the Institutional Review Board (IRB) of Bangabandhu Sheikh Mujib Medical University (Memo no: BSMMU/2016/10817).

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Statement of Ethics

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Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Conception and design: MSIB, FW, RN

Acquisition, analysis and interpretation of data: FW, MZS, KNS

Manuscript drafting and revising it critically: MSIB, FW, MZS, FA

Approval of the final version of the manuscript: MSIB, FW, MZS, and KNS

Guarantor accuracy and integrity of the work: MSIB, FW, MZS

Data Availability Statement

The data that support the findings of this study are openly available at MendeleyDOI:10.17632/m2rg66r88p.1. Further inquiries can be directed to the corresponding authors.

References

1. Reinhold-Keller E, Herlyn K, Wagner-Bastmeyer R, Gross WL. Stable incidence of primary systemic vasculitides over five years: results from the German vasculitis register. Arthritis Rheum. 2005 Feb 15;53(1):93-9. doi: 10.1002/art.20928.

2. Caproni M, Verdelli A. An update on the nomenclature for cutaneous vasculitis. Curr OpinRheumatol. (2019) 31:46–52. 10.1097/BOR.0000000000000563

3. Alpsoy E. Cutaneous vasculitis; An algorithmic approach to diagnosis. Front Med (Lausanne). 2022 Sep 21;9:1012554. doi: 10.3389/fmed.2022.1012554.

4. Carlson JA, Ng BT, Chen KR. Cutaneous vasculitis update: diagnostic criteria, classification, epidemiology, etiology, pathogenesis, evaluation and prognosis. Am J Dermatopathol. 2005 Dec;27(6):504-28. doi: 10.1097/01.dad.0000181109.54532.c5.

5. Marzano AV, Vezzoli P, Berti E. Skin involvement in cutaneous and systemic vasculitis. Autoimmun Rev. 2013 Feb;12(4):467-76. doi: 10.1016/j.autrev.2012.08.005.

6. James WD, Elston DM, McMahon PJ. Andrews' Diseases of the Skin: Clinical Dermatology. 13th Edition. Publisher: Elsevier Inc. 2020.

7. Baigrie D, Goyal A, Crane JS. Leukocytoclastic Vasculitis. [Updated 2023 Aug 8]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK482159/.

8. Xu L, Li Y, Wu X. IgA vasculitis update: Epidemiology, pathogenesis, and biomarkers. Front Immunol. 2022 Oct 3;13:921864. doi: 10.3389/fimmu.2022.921864.

9. Eswaran H, Googe P, Vedak P, Marston WA, Moll S. Livedoid vasculopathy: A review with focus on terminology and pathogenesis. Vasc Med. 2022 Dec;27(6):593-603. doi: 10.1177/1358863X221130380.

10. Watts RA, Lane SE, Bentham G, Scott DG. Epidemiology of systemic vasculitis: a ten-year study in the United Kingdom. Arthritis Rheum. 2000 Feb;43(2):414-9. doi: 10.1002/1529-0131(200002)43:2<414::AID-ANR23>3.0.CO;2-0.
11. Puéchal X. Polyarteritis Nodosa: State of the art. Joint Bone Spine. 2022 Jul;89(4):105320. doi: 10.1016/j.jbspin.2021.105320.
12. Kitching AR, Anders HJ, Basu N, Brouwer E, Gordon J, Jayne DR, Kullman J, Lyons PA, Merkel PA, Savage COS, Specks U, Kain R. ANCA-associated vasculitis. Nat Rev Dis Primers. 2020 Aug 27;6(1):71. doi: 10.1038/s41572-020-0204-y.
13. Greco A, Marinelli C, Fusconi M, Macri GF, Gallo A, De Virgilio A, Zambetti G, de Vincentiis M. Clinic manifestations in granulomatosis with polyangiitis. Int J ImmunopatholPharmacol. 2016 Jun;29(2):151-9. doi: 10.1177/0394632015617063.
14. Emmi, G., Bettiol, A., Gelain, E. et al. Evidence-Based Guideline for the diagnosis and management of eosinophilic granulomatosis with polyangiitis. Nat Rev Rheumatol 19, 378–393 (2023). https://doi.org/10.1038/s41584-023-00958-w.
15. Watts RA, Hatemi G, Burns JC, Mohammad AJ. Global epidemiology of vasculitis. Nat Rev Rheumatol. 2022 Jan;18(1):22-34. doi: 10.1038/s41584-021-00718-8.
16. Gu SL, Jorizzo JL. Urticarial vasculitis. Int J Womens Dermatol.
2021 Jan 29;7(3):290-297. doi:10.1016/j.ijwd.2021.01.021.
17. Morita TCAB, Criado PR, Criado RFJ, Trés GFS, Sotto MN. Update on vasculitis: overview and relevant dermatological aspects for the clinical and histopathological diagnosis - Part II. An Bras Dermatol. 2020 Jul-Aug;95(4):493-507. doi: 10.1016/j.abd.2020.04.004.
18. Watts RA, Scott DG. Epidemiology of the vasculitides. Current Opinion in Rheumatology. 2003 Jan;15(1):11-16. DOI: 10.1097/00002281-200301000-00003.
19. Stanton M, Tiwari V. Polyarteritis Nodosa. [Updated 2023 Feb 22]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK482157/
20. Micheletti RG, Chiesa Fuxench Z, Craven A, Watts RA, Luqmani RA, Merkel PA; DCVAS Investigators. Cutaneous Manifestations of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. Arthritis Rheumatol. 2020 Oct;72(10):1741-1747. doi: 10.1002/art.41310.