Pustular Vasculitis: Different Names for Same Entity?

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Sir,

Classic leukocytoclastic vasculitis presenting as palpable purpura is a commonly encountered entity in dermatology. Here, we report a case of pustular leukocytoclastic vasculitis which is a relatively rare clinical presentation with only a few cases reported in the literature. Clinically, it predominantly presents on acral sites as pustules or bullae on a purpuric base. Several terms in the literature have been used in the past to describe pustular vasculitis, and here we briefly review the introduction and evolution of this case.

Pustular vasculitis is a rare clinical entity which was first reported by Jorizzo *et al.* in 1983. Clinically, it is characterized by pustules or pustular-plaques on a purpuric base and histologically depict features of leukocytoclastic vasculitis.[1] It is commonly misdiagnosed and often treated as an infectious process. Alternative diagnoses are considered only when the patient fails to respond to antibiotics. Here, we report a case of classic pustular vasculitis along with a brief review of evolution of this distinct entity.

A 64-year-old male presented to the Mount Sinai Hospital in New York with the chief complaint of bilateral lower extremity pain for 3 days and multiple painful fluid-filled lesions on the legs of 1-day duration. His past medical history was significant for hepatitis C-induced liver cirrhosis, HIV, end-stage renal disease, hypertension, and deep vein thrombosis (DVT). His medications included rifaximin, ant-retroviral treatment (Darunavir, Dolutegravir, Ritonavir), nadolol, lactulose, sucralfate, and lisinopril. In addition, he was on hemodialysis for end-stage renal disease, and had an IVC filter for DVT. He had fever (100.4°F) and diffuse polyarthralgias, with no other systemic symptoms, recent travel, or new medications. There was no history of similar lesions in the past. He smoked 10 cigarettes/day for >30 years but denied alcohol or illicit drug use. His family history was not medically relevant. On physical examination, multiple pustules and bullae with turbid fluid were noted on a purpuric base on bilateral lower legs [Figure 1]. No oral or genital involvement was noted. His systemic examination was unremarkable. His basic work up was significant for anemia, thrombocytopenia, deranged liver and kidney function tests owing to his cirrhosis, and end-stage renal disease. His viral load was undetectable and CD4 count was 153/µl. Wound and blood cultures were negative for bacteria, virus, or fungi. Doppler ultrasound was negative for any deep venous thrombosis.

Two skin biopsies were performed from the lesional and peri-lesional skin for histopathology and direct immunofluorescence testing (DIF) testing, respectively. Skin biopsy was noted to have perivascular mixed infiltrate of mononuclear

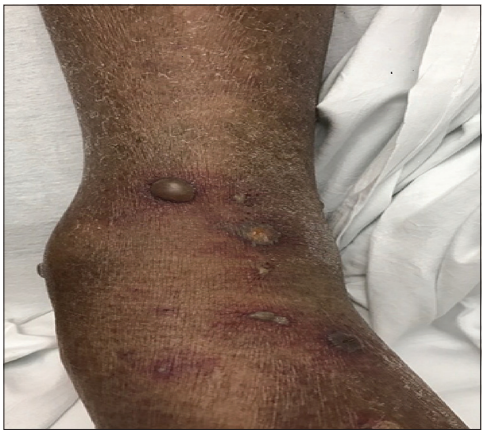

Figure 1: Multiple pustules and bullae with turbid fluid were noted on a purpuric base bilaterally; here on left leg

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cells, neutrophils with focal fragmentation of their nuclei, and extravasated erythrocytes. The endothelial cells were swollen and demonstrated fibrin in their walls. DIF showed granular perivascular deposition of C3 and fibrinogen; sparse granular perivascular deposits of IgG were seen without any significant deposition of IgM or IgA. Based on these findings, a diagnosis of pustular leukocytoclastic vasculitis was made. Because there was no evidence of systemic vasculitis in our patient, he was managed with supportive care and all his lesions resolved by the end of 2 weeks. The trigger for his acute outbreak remains unclear [Figures 2-4].

Various terms have been used in the literature to describe conditions that are clinically characterized by pustular acral lesions and histologically by a neutrophilic infiltrate with or without features of vascular damage. Jorizzo *et al.* introduced the term pustular vasculitis in 1983, describing a group of conditions that exhibited overlapping histologic changes of Sweet's syndrome and leukocytoclastic vasculitis. In their review, they use the term pustular vasculitis to include Behcet's syndrome, gonococcal dermatosis-arthritis-syndrome, bowel-associated dermatosis-arthritis-syndrome, and chronic meningococcemia.[1-3] Thereafter, McNeely *et al.* described an entity called primary idiopathic pustular vasculitis.[4] There are reports of another condition called acute generalized pustular bacterid which presents as a single episode of pustular vasculitis following streptococcal upper respiratory tract infections.[5,6] In 1995, Strutton *et al.* described six patients with symmetric lesions on bilateral hands. Clinically, these lesions resembled Sweet's syndrome but biopsy specimens showed severe leukocytoclastic vasculitis.[7] They called it a distinct entity termed as pustular vasculitis of the hands. This was followed by reports of clinically similar lesions on the hands, but they lacked evidence of vasculitis on histology, which led to the introduction of the term neutrophilic dermatosis of the dorsal hands.[8-10] Later in 2004, Weenig *et al.* excluded the word dorsal from the term as they observed several patients with pustular lesions involving the palmar aspect of hands as well and termed it neutrophilic dermatosis of the hands.[11] Pustular lesions with purpuric base on the legs were described by Selvan *et al.*[12] Most recently, the term acral Sweet's syndrome has been used to describe similar lesions.[13]

Another interesting case report by Lazarov *et al.* described a patient with disseminated pustular eruption compatible with pustular vasculitis and concomitant bullous lesions along with the features of superficial bullous pyoderma gangrenosum during an exacerbation of ulcerative colitis. The authors suggested that pustular vasculitis is another skin manifestation of ulcerative colitis.

All the entities described above have common clinical features of pustules on purpuric bases, pustular plaques, or bullous lesions and histological features of neutrophilic dermatosis with or without features of leukocytoclastic vasculitis. Occurrence of these lesions on both hands and feet have been described in the literature. Skin biopsy is imperative in making the diagnosis. The term neutrophilic dermatoses is preferred if there is no histological evidence of vasculitis, however, it should be diagnosed as pustular vasculitis if there are signs of vessel damage in addition to features of neutrophilic dermatoses.[4,14,15] Controversy remains whether pustular vasculitis is a variant of neutrophilic dermatoses, but it can be concluded that

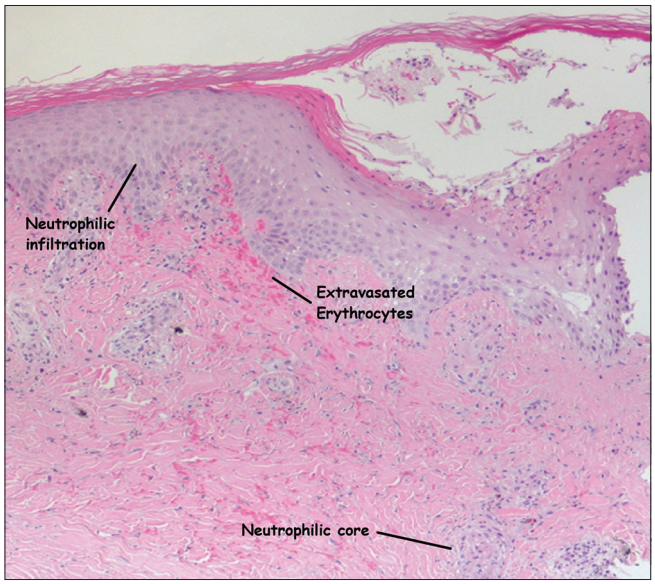

Figure 2: Neutrophilic core and perivascular neutrophilic infiltration with extravasated erythrocytes. [Hematoxylin and eosin stain; ×4 magnification]

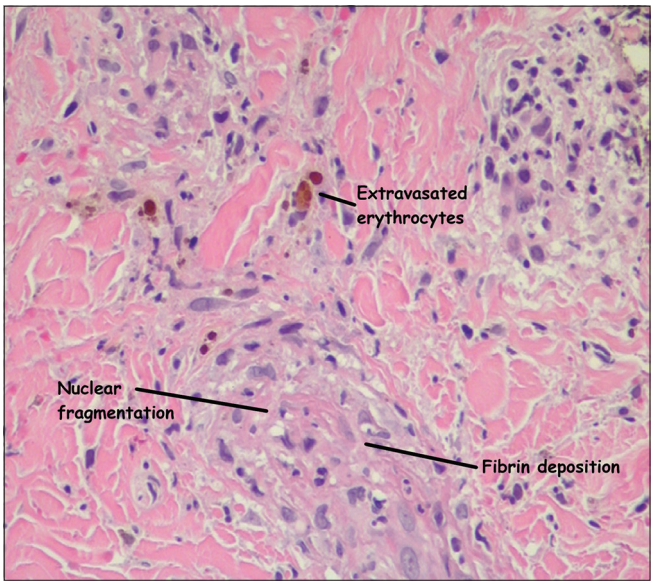

Figure 3: Fibrin deposition; nuclear fragmentation (leukocytoclasia) along the vessel wall and extravasated erythrocytes are seen. [Hematoxylin and eosin stain, ×40 magnification]

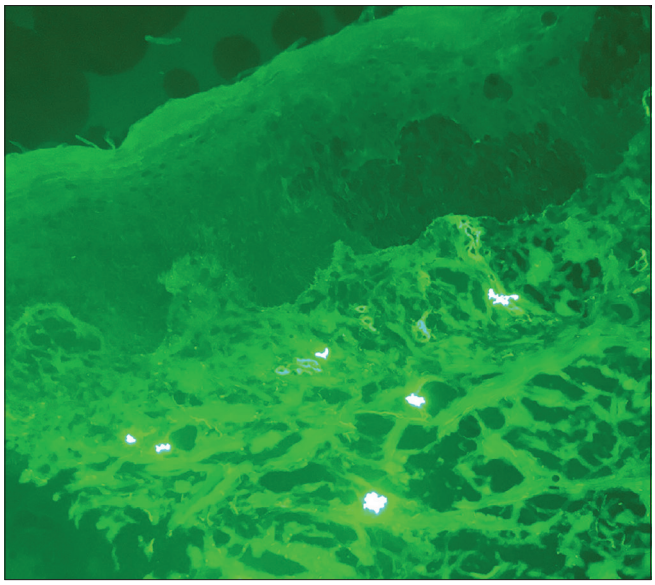

Figure 4: DIF with C3 and fibrinogen deposition

these various designations represent similar conditions. Our case represents idiopathic pustular leukocytoclastic vasculitis, and we think it is important to familiarize readers with this entity and the different terms used to describe it as they can be confusing and often lead to misdiagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Jorizzo JL. Pustular vasculitis: An emerging disease concept. J Am Acad Dermatol 1983;9:160-2.
2. Jorizzo JL, Hudson RD, Schmalstieg FC, Daniels JC, Apisarnthanarax P, Henry JC, *et al.* Behçet's syndrome: Immune regulation, circulating immune complexes, neutrophil migration, and colchicine therapy. J Am Acad Dermatol 1984;10:205-14.
3. Jorizzo JL, Schmalstieg FC, Dinehart SM, Daniels JC, Cavallo T, Apisarnthanarax P, *et al.* Bowel-associated dermatosis-arthritis syndrome: Immune complex-mediated vessel damage and increased neutrophil migration. Arch Intern Med 1984;144:738-40.
4. McNeely MC, Jorizzo JL, Solomon AR, Schmalstieg FC, Cavallo T. Primary idiopathic cutaneous pustular vasculitis. J Am Acad Dermatol 1986;14:939-44.
5. Miyachi Y, Danno K, Yanase K, Imamura S. Acute generalized pustular bacterid and immune complexes. Acta Derm Venereol 1980;60:66-9.
6. Tan RH. Acute generalized pustular bacterid An unusual manifestation of leukocytoclastic vasculitis. Br J Dermatol 1974;91:209-15.
7. Struton G, Weedon D, Robertson I. Pustular vasculitis of the hands. J Am Acad Dermatol 1995;32:192-8.
8. Galaria NA, Junkins-Hopkins JM, Kligman D, James WD. Neutrophilic dermatosis of the dorsal hands: Pustular vasculitis revisited. J Am Acad Dermatol 2000;43:870-4.
9. Hall AP, Goudge RJ, Ireton HJ, Burrell LM. Pustular vasculitis of the hands. Australas J Dermatol 1999;40:204-7.
10. DiCaudo DJ, Connolly SM. Neutrophilic dermatosis (pustular vasculitis) of the dorsal hands: A report of 7 cases and review of the literature. Arch Dermatol 2002;138:361-5.
11. Weenig RH, Bruce AJ, McEvoy MT, Gibson LE, Davis MD. Neutrophilic dermatosis of the hands: Four new cases and review of the literature. Int J Dermatol 2004;43:95-102.
12. Selvan S, Shakir R, Chan A. Case Report: Pustular vasculitis. BMJ Case Rep 2013;2013:bcr2013008806.
13. Boussofara L, Gammoudi R, Ghariani N, Aounallah A, Sriha B, Denguezli M, *et al.* Neutrophilic dermatosis of the hands: Acral sweet syndrome? Rev Med Interne 2015;36:287-90.
14. Tous-Romero F, Prieto-Barrios M, Velasco-Tamariz V, Burillo-Martínez S. RF-neutrophilic dermatoses of the hands: Different names for the same condition? Actas Dermosifiliogr 2017;5:471-2.
15. Del Pozo J, Sacristan F, Martinez W, Paradela S, Fernández-Jorge B, Fonseca E. Neutrophilic dermatosis of the hands: Presentation of eight cases and review of the literature. J Dermatol 2007;34:243-7.