Annular Pattern of Pustular Vasculitis: A Case Report of Unusual Morphological Presentation

Abstract

Pustular cutaneous vasculitis is a rare entity. Only handful cases are reported from all over the world. They typically present with painful pustules mainly over dorsum of hands and rarely on lower legs. We report a 50-year female who presented with painful pustules arranged in annular pattern predominantly distributed over lower limbs. Histopathology was suggestive of pustular vasculitis. The patient was successfully treated with oral prednisolone and dapsone without any recurrence. We report this case because of unusual morphology with uncommon site of presentation.

Keywords: Annular pattern, lower limbs, NDDH, neutrophilic dermatosis, pustular vasculitis

Introduction

Pustular vasculitis/neutrophilic dermatosis of dorsal hands (NDDH) is a variant of sweet syndrome. It is an extremely rare condition with very few cases reported worldwide. Described lesions include painful pustules on purpuric bases.[1] Majority of the reported cases have involvement of dorsum of hands.[2,3] We report a 50-year-old female with painful pustular lesions arranged in annular pattern and lesions were predominantly distributed over lower limbs. Histopathology was suggestive of pustular vasculitis. The patient was successfully treated with oral prednisolone and dapsone without any recurrence. We report this case because of unusual morphology with uncommon site of presentation.

Case Report

A 50-year-old female presented with sudden onset of painful erythematous and purpuric plaques on medial aspect of bilateral feet, within 5–6 days it involved bilateral legs, thighs and arms. She had severe burning pain on limbs, without any constitutional features or joint pain. There was no history suggestive of infection, connective tissue disorder, Bechet’s syndrome or prior drug intake. On clinical examination, there were multiple erythematous macules and plaques studded with discrete pustules of 0.5 × 0.5 cm² size. Majority of pustules were arranged either at the periphery of a central pustule or collapsed bulla in annular pattern [Figure 1a and b]. Lesions on medial aspect of bilateral foot had undergone ulceration and crusting. In addition, there were multiple haemorrhagic spots on bilateral foot and splinter haemorrhages on tips of fingers, toes and nail beds [Figure 2a and b]. Examinations of all other systems were within normal limit. Patient denied any prior heart disease or surgery. Based on these finding differential diagnosis of leucocytoclastic vasculitis, pustular vasculitis, subacute bacterial endocarditis and erythema multiforme were considered and patient was investigated.

Laboratory investigations revealed a markedly elevated white cell count (19.6 × 10⁻⁹/l), neutrophilia (18 × 10⁻⁹/dl), raised C reactive protein (26 mg/dl) and raised ESR (56 mm in first hour). Other investigations such as renal function test, liver function test, HIV, Hep B and HCV, anti-streptolysin ‘O’ titre, throat swab culture, rheumatoid factor, antinuclear antibody, herpes simplex virus antibodies to Ig M, Ig G, and serology for mycoplasma were within the normal limit. Chest x-ray showed no abnormality.

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Access this article online
Website: www.idoj.in
DOI: 10.4103/idoj.IDOJ_100_19

How to cite this article: Sahu K, Sirka CS, Pradhan S, Rout AN, Nayak S. Annular pattern of pustular vasculitis: A case report of unusual morphological presentation. Indian Dermatol Online J 2020;11:232-4.

Received: March, 2019. Revised: March, 2019. Accepted: June, 2019. Published: March, 2020.
radiography was normal and transthoracic echocardiogram showed no evidence of valvular vegetations. Culture sensitivity of blood, urine and pus taken from skin did not grow any organism. Ultrasound abdomen and pelvis did not reveal any abnormality. Histopathology from skin lesions revealed intra-epidermal abscess, dermal endothelial swelling, fibrin deposit in vessel wall with neutrophilic infiltrate causing karyorrhexis and exocytosis of erythrocytes [Figures 3 and 4a, b]. Predominant finding of neutrophilic abscess and vasculitis favours histological diagnosis of pustular vasculitis over leucocytoclastic vasculitis. Hence, a diagnosis of primary idiopathic cutaneous pustular vasculitis was made. Patient was started on prednisolone 20 mg daily and dapsone 100 mg twice daily. The lesions improved completely after 15 days of therapy. The lesions improved completely after 15 days of therapy. She was maintained with 100 mg dapsone and there was no recurrence even after six months of follow up [Figure 5].

Discussion

Pustular vasculitis is an extremely rare condition with only few cases reported in literature throughout the world.[1] Underlying pathogenesis, is believed to be immune-complex mediated lysosomal product release causing neutrophilic inflammatory infiltrate.[2] Authors believe it to be is interchangeable with NDDH and a variant of neutrophilic dermatosis such as sweet syndrome and Bechet’s disease.[3]

It is characterised by papules, haemorrhagic plaques, bullous lesions[3] and pustules on purpuric bases.[4] Systemic involvement, myalgia and neutrophilia may be present. Other associated findings may be pathergy and neutrophilia. The lesions are often intensely painful “often disproportionate to the size”.[2] Our patient presented with intensely painful pustular lesions and new lesions were appearing around the old lesions giving annular appearance. Authors could not explain the reasons for such a morphology. In addition, there were large ulcers and haemorrhagic spots on toes, which are also not described with pustular vasculitis.

Majority of cases described in the literature predominantly involve the dorsal hands and may subsequently spread to involve the lower limbs.[2-3] In the present case, lesions were predominantly distributed on lower limbs and few on bilateral hands. It was similar to one report by Selvan et al. where lesions were confined to the lower limbs.[5]

Underlying systemic diseases can include Behçet’s syndrome,[2] inflammatory bowel disease, myeloproliferative disorders, visceral malignancies and chronic gonococcaemia.[2,4] Other common associations are thermal injury, exposure to chemical fertiliser, pregnancy, drug reactions and streptococcal infection.[3,7] Additionally, primary idiopathic pustular vasculitis has also been described[9] in absence of predisposing conditions. As investigations ruled out focus of infection or any
other systemic abnormality, our case was of the primary idiopathic type.

Histopathologically, the lesions share common features of neutrophilic dermatosis and leukocytoclastic vasculitis.\cite{3,4} Vascular endothelial damage can range from endothelial swelling to fibrinoid necrosis.\cite{3} Sub epithelial oedema, spongiosis, ulceration and neutrophilic micro-abscesses may also be present.\cite{7} In our case, histopathology had similar neutrophilic micro-abscess along with features of vasculitis suggestive of pustular vasculitis. Malone et al. proposed that the vascular damage was probably a secondary event related to the intensity of the neutrophilic infiltrate and the time of evolution of the lesions and did not represent true vasculitis.\cite{10} The term pustular vasculitis of hands was revised after recognising that vasculitis was an inconsistent finding and of secondary importance in its diagnosis.\cite{1}

The differential diagnosis includes pustular vasculitis, acute febrile neutrophilic dermatosis (Sweet’s syndrome), small vessel vasculitis and pyoderma gangrenosum.\cite{8} But the histopathology can distinguish pustular vasculitis from others.

Management is primarily corticosteroids and/or dapsone.\cite{3} Resolution of lesions takes place over a period of weeks. Recurrence of the lesions is common. Low-dose dapsone has been used to prevent/reduce episodes of recurrence\cite{1}, and cyclosporine and tacrolimus can be used in resistant cases.\cite{3}

Our case is unique as there were extensive pustular lesions with annular morphology along with ulcerations and haemorrhagic spots, which have not been described previously. The lesions were mostly confined to lower limbs, which is again an uncommon finding. We report this case because of its rarity and emphasise that physicians must keep in mind these type of clinical features while dealing with lesions of vasculitis.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. 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.
2. Koulaouzidis A, Campbell S, Bharati A, Leonard N, Azurdia R. Primary biliary cirrhosis associated pustular vasculitis. Ann Hepatol 2006;5:177–8.
3. Del Pozo J, Sacristan F, Martínez W, Paradela S, Fernández-Jorge B, Fonseca E, et al. Neutrophilic dermatosis of the hands: Presentation of eight cases and review of the literature. J Dermatol 2007;34:243–7.
4. McNeely MC, Jorizzo JL, Solomon AR Jr, Schmalstieg FC, Cavallo T. Primary idiopathic cutaneous pustular vasculitis. J Am Acad Dermatol 1986;14:939–44.
5. Selvan S, Shakir R, Chan A. Pustular vasculitis. BMJ Case Rep 2013;2013:bcr2013008806.
6. 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.
7. Curro N, Pagerois X, Tarroch X. Pustular vasculitis of the hands. Report of two men. Dermatology 1998;196:346–7.
8. Yung A, Merchant W, Sheehan-Dare R. Streptococcus induced pustular vasculitis affecting the hands resembling pustular vasculitis of the hands—first reported case. Clin Exp Dermatol 2005;30:366–8.
9. Asano Y, Kaji K, Idezuki T. A case of primary idiopathic cutaneous pustular vasculitis. Acta Derm Venereol 2010;90:420–1.
10. Malone JC, Slone SP, Wills-Frank LA, Fearnehough PK, Lear SC, Goldsmith LJ, et al. Vascular inflammation (vasculitis) in Sweet syndrome: A clinicopathologic study of 28 biopsy specimens from 21 patients. Arch Dermatol 2002;138:345–9.