Violaceous Papules and Plaques over Forearms

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Question

A 65-year-old Asian woman with a history of rheumatoid arthritis and type 2 diabetes mellitus for the past 4 years presented to the dermatology out-patient department with multiple erythematous to violaceous, slightly scaly papules and plaques over extensors of forearms [Figure 1a]. There were interspersed small ulcers with a hemorrhagic crust. The lesions were mildly itchy and coincided with the exacerbation of her arthritis. Dermoscopy revealed a reddish-brown background, follicular plugs, orange-brown structureless areas, shiny white lines, white to brown scales, prominent irregular dot vessels, and linear vessels with and without bends [Figure 1b]. Histopathology revealed epidermal acanthosis, increased dermal vessels, palisaded and interstitial granulomas composed of lymphocytes, histiocytes, giant cells (Foreign body and Langhans type) surrounding degenerated collagen, many neutrophils, and abundant nuclear dust with leukocytoclastic vasculitis of a few vessels [Figure 2a-c]. Mucin was inconspicuous on Alcian blue staining.

What is the diagnosis?

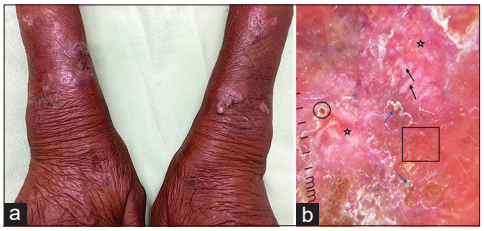

Figure 1: (a) Multiple erythematous to violaceous, slightly scaly papules and plaques over extensors of forearms. (b) Dermoscopy shows a reddish-brown background, follicular plugs (black circle), orange-brown structureless areas (black square), white streaks (black arrow), white to brown scales (blue arrow) prominent irregular dot vessels, and linear vessels with and without bends (star). (Polarized mode, Dermlite DL200 hybrid, 10 × magnification)

Diagnosis

Palisaded neutrophilic granulomatous dermatitis.

Discussion

Palisaded neutrophilic granulomatous dermatitis (PNGD) is now considered an umbrella term to include interstitial granulomatous dermatitis, granulomatous drug reaction, Churg Strauss granuloma, rheumatoid granuloma, and cutaneous necrotizing extravascular granuloma.[1] It represents a reaction pattern to underlying systemic inflammation and has been reported in association with a myriad of diseases, including rheumatoid arthritis, systemic lupus erythematosus, anti-neutrophil cytoplasmic autoantibody-associated vasculitis, systemic scleroderma, Sjogren's syndrome, Hodgkin's lymphoma, chronic myeloid leukemia, inflammatory bowel disease, and various infections (streptococcal, Epstein Barr virus, human immunodeficiency virus, Hepatitis C, mycoplasma, and parvovirus).[2] Histiocytoid cells in PNGD lesions of systemic lupus erythematosus patients show diffuse positivity for cluster of differentiation 68 (CD68) and CD163, suggesting the role of M2 macrophages in pathogenesis.[3] There are also anecdotal reports of its association with certain drugs, namely, allopurinol, ledipasvir/sofosbuvir, and adalimumab. Dykman *et al*.[4] in 1965 first described it in a patient with rheumatoid arthritis presenting with linear bands over the trunk, known as the rope sign, and labeled it as a rheumatoid granuloma. Later, Chu *et al*.[1] in 1994 coined the term PNGD, which is most accepted now. It is an uncommonly reported entity, and a PubMed search revealed 60 cases until December 31, 2017. PNGD could be under-diagnosed or mis-diagnosed as the clinical presentation and histopathology vary with the stage of the disease. Early lesions clinically present

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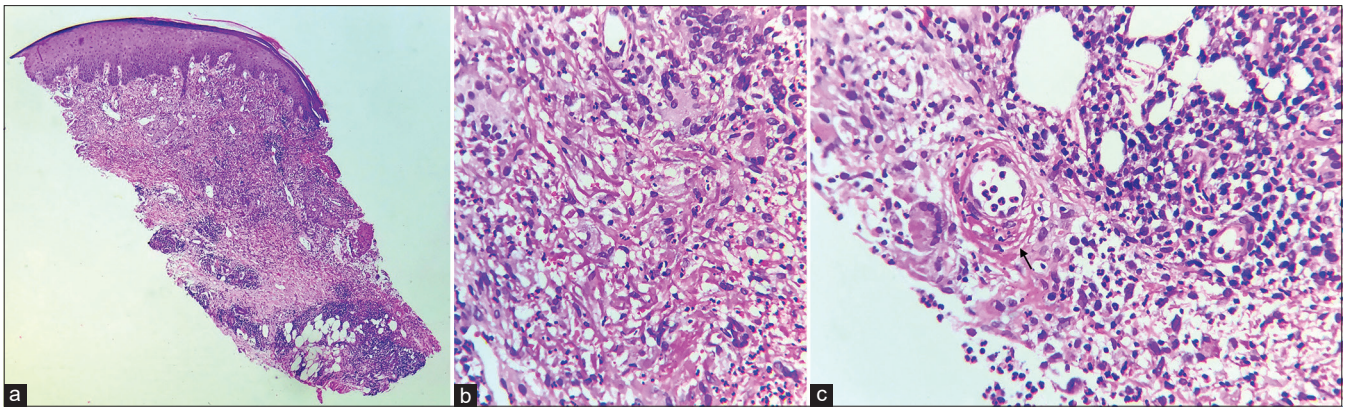

Figure 2: (a) Acanthotic epidermis, increased dermal blood vessels with interstitial and palisaded granulomas surrounding degenerated collagen and interspersed neutrophils with nuclear dust (H and E ×10). (b) Interstitial and palisaded granulomas surrounding degenerated collagen and interspersed neutrophils (H and E ×40). (c) Deep dermal vessel exhibiting leukocytoclastic vasculitis (black arrow, H and E ×40)

as erythematous macules or urticarial plaques, whereas mature lesions are violaceous papules, plaques, or nodules distributed symmetrically over extremities and lateral sides of the trunk. Umbilicated papules over bony prominences have also been reported. Histopathologically early lesions are characterized by leukocytoclastic vasculitis, possibly because of immune complex deposition with dense neutrophils that can be mis-interpreted as Sweet's syndrome or cutaneous small-vessel vasculitis. Evolved lesions predominantly show palisaded granulomas surrounding degenerated collagen with a variable degree of interstitial mucin, fibrin, and neutrophilic debris. These changes involve the whole dermis, primarily the lower dermis. Old lesions show, besides, ample mucin and fibrosis. These can be confused histopathologically by the unwary with granuloma annulare or suppurative granulomas if neutrophilic infiltrate is prominent. Histological resemblance with cutaneous extravascular necrotizing granulomas led to it being labeled as Churg Strauss granuloma previously. Thus, one biopsy may not be diagnostic of PNGD. However, the most characteristic hallmark appears to be palisaded granulomas surrounding degenerated collagen with neutrophilic debris. A high index of suspicion can help in diagnostic confirmation. Some consider PNGD, granuloma annulare, and interstitial granulomatous dermatitis as being on the same spectrum. Interstitial granulomatous dermatitis presents as annular plaques over flexures exhibiting interstitial infiltrate of histiocytes, including giant cells showing elastophagocytosis, lymphocytes, eosinophils, and plasma cells. It is seen as a reaction to drugs such as calcium channel blockers, diuretics, and angiotensin-converting enzyme inhibitors in addition to underlying systemic diseases such as rheumatoid arthritis, diabetes, and infections. However, considering the clinicopathological overlap, it is most prudent to categorize it under PNGD. Treatment of underlying diseases results in resolution of lesions, strengthening the belief that these represent a reaction pattern. Other available treatment options include topical and oral corticosteroids, non-steroidal anti-inflammatory drugs, infliximab, colchicine, and dapsone. In the present case, a classical clinical setting with palisaded and interstitial granulomas along with nuclear dust clinched the diagnosis. Dermoscopy of PNGD has been reported previously to reveal a reddish background, keratin plugs, variable vessels (dot, loop, and linear), gray-white scales, orange-brown structureless areas, and white dots and streaks.[5] All these were seen in the reported case. Orange-brown structureless areas represent underlying granulomas, and white streaks and dots represent degenerated collagen. In the present case, thick white streaks were seen, suggesting abundant degenerated collagen confirmed on histopathology. The patient was treated with oral methylprednisolone in a dose of 32 mg per day for rheumatoid arthritis, and her cutaneous lesions reduced substantially in 2 weeks.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Chu P, Connolly MK, LeBoit PE. The histopathologic spectrum of palisaded neutrophilic and granulomatous dermatitis in patients with collagen vascular disease. Arch Dermatol 1994;130:1278-83.
2. Kalen JE, Shokeen D, Ramos-Caro F, Motaparthi K. Palisaded neutrophilic granulomatous dermatitis: Spectrum of histologic findings in a single patient. JAAD Case Rep 2017;3:425-8.
3. Terai S, Ueda-Hayakawa I, Nguyen CTH, Ly NTM, Yamazaki F, Kambe N, *et al*. Palisaded neutrophilic and granulomatous dermatitis associated with systemic lupus erythematosus: Possible involvement of $CD163^{+}$M2 macrophages in two cases, and a review of published works. Lupus 2018;27:2220-7.
4. Dykman CJ, Galens GJ, Good AE. Linear subcutaneous bands in rheumatoid arthritis. An unusual form of rheumatoid granuloma. Ann Intern Med 1965;63:134-40.
5. Razmi TM, Sawatkar GU, Sekar A, Vinay K, De D, Saikia UN, *et al*. Dermoscopy of palisaded neutrophilic and granulomatous dermatitis. Clin Exp Dermatol 2019;44:e34-8.