CASE REPORT

Pustular generalized perforating granuloma annulare

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Summary

We report a 84-year-old man with a 13-year history of recurrent generalized asymptomatic pustular lesions. Histology revealed areas of necrobiosis surrounded by palisading granulomas and transepidermal elimination of the necrobiotic material. A dense infiltrate of neutrophils was also found. Although 26% of patients with generalized perforating granuloma annulare have some yellow pustule-like papules, these correspond histologically to the yellow viscous necrobiotic material extruding through the epidermis and not to a real neutrophilic infiltrate. This is the first case report of perforating granuloma annulare with recurrent generalized pustular lesions with a dense infiltrate of neutrophils.

Key words: generalized perforating granuloma annulare, neutrophils, pustules

Case report

An 84-year-old man presented with a 13-year history of crops of recurrent pustular lesions with an erythematosus halo, 1–2 mm in diameter, symmetrically located in the abdominal area, trunk, extensor surfaces of upper and lower limbs, dorsum of the hands, and the palms (Figs 1–3). Lesions were asymptomatic. He also exhibited a few scaly, umbilicated papules and plaques made by the confluence of pustular lesions. The lesions resolved without scarring.

Histology of the pustular lesions showed transepidermal and follicular elimination of degenerated mucinous collagen bundles, and granulomas with palisading lymphoid cells and histiocytes and a dense infiltrate of neutrophils. Granulomas were located in the upper dermis. Microbiological culture of the pustules was performed four times and was negative. Full blood count, blood chemistry and urinalysis showed no abnormalities.

The patient received treatment with pentoxifylline 500 mg three times daily for 7 months and then acitretin 35 mg daily for 8 months. He showed an initial improvement but after 1 month crops of new lesions appeared. In January 2001 he received treatment with prednisone, tapered from 30 mg daily to 10 mg on alternate days, with improvement of lesions that only reappeared after withdrawal of treatment.
Since January 2002 we started treatment with psoralen plus ultraviolet A (PUVA) and low-dose prednisone (10 mg on alternate days), with no crops of new lesions.

Discussion

PGA is a rare subtype of GA first named by Owens and Freeman.\(^1\) It can be localized or generalized, with generalized being the most common form of presentation. It affects children and young adults, and females are affected twice as often than males. One familial case has been reported.\(^2\)

Some cases are associated to diabetes.\(^3-7\) Both types of PGA (localized and generalized) are related to diabetes mellitus (17% of cases). The mean age at onset of the disease is higher in the diabetic group.\(^7\) Our patient started with PGA lesions at a very late age (71 years) compared with the mean age at onset of 31 years.

Generalized PGA is clinically characterized by crops of erythematous, umbilicated or scaly papules. Pustule-like lesions, scars and plaques may also be seen, especially in the generalized form.\(^7\) Lesions are localized in the abdominal area, trunk, extremities, dorsum of hands, and palms. Transepithelial elimination may occur by the transepidermal and/or the transfollicular route.\(^8\)

Pustular lesions are present in 26% of cases of PGA (localized and generalized) but in 58% of cases of generalized GA.\(^7\) Pustular lesions are usually present with red, scaly and crusted papules. These do not

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**Figure 1.** Pustules located on the trunk and abdomen.

**Figure 2.** Pustules surrounded by an erythematous halo.

**Figure 3.** Pustules on the abdomen surrounded by a red halo, and pustular and scaly lesions on the palm.

**Figure 4.** Histological examination of a pustular lesion on the palm shows necrobiotic material surrounded by granulomas with transepidermal perforation and some neutrophils (haematoxylin and eosin, original magnification \(\times 40\)).
represent true pustular lesions and are filled with a yellow, viscous necrobiotic material, not with a neutrophilic infiltrate. The clinical presentation of PGA as a generalized pustular eruption is rare. The differential diagnosis includes psoriasis, pustular drug eruptions, folliculitis and Sneddon–Wilkinson disease.

There is one reported case of PGA with the clinical presentation of mainly pustular lesions; our patient represents the second case. Both cases had necrobiotic areas surrounded by lymphocytes, epithelioid cells and Langerhans cells, but the previous reported case had only some neutrophils located close to the transepidermal area, whereas our patient had a dense infiltrate of neutrophils.

There is also a reported case of generalized GA presenting with recurrent eruptions of generalized pustules that histologically showed well-circumscribed palisading granulomas and a dense neutrophilic infiltrate. The age at onset, the time-course of disease evolution and the good response to oral corticosteroid treatment were similar to our case, the main differences being that repeated biopsies of the pustular lesions did not show perforation and that all lesions had a follicular distribution. Only on one occasion did the patient present some lesions on the palms that histologically demonstrated PGA.

The presence of neutrophils in GA lesions is rare, but some authors have occasionally found polymorphonuclear leucocytes and nuclear dust in the centre of necrobiotic areas in early lesions of GA, suggesting that they may have a primary or secondary role in the initial events of this disease, either inducing the necrobiotic changes or acting as phagocytes. We have found some cases of PGA in which some biopsies showed some neutrophils.

In summary, we present an 84-year-old man with a 13-year history of crops of asymptomatic pustular lesions that histologically showed necrobiotic material, surrounded by granulomas, epidermal and follicular perforation and a dense infiltrate of neutrophils. Lesions improved with oral corticosteroids and with PUVA. No other associated abnormalities were found.

References
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