

Acute and recurrent pustulosis: consolidating uncommon cases of follicular pustulosis induced by UV light and other triggers

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ABSTRACT

Background: There are a growing number of patients with acute and recurrent pustular reactive dermatitis reported without clear parameters to define the entities. Consolidation of cases under the term acute and recurrent pustulosis (ARP) will aid dermatologists in diagnosing such patients in the future.

Objective: Describe the parameters which define acute and recurrent pustulosis and communicate the high predominance for onset in young women based on reported cases.

Methods: PubMed literature search for reports of recurrent follicularly centered neutrophilic eruptions.

Results: According to the clinical characteristics of ARP, 23 patients were identified from prior reports. Interestingly, 20 out of 23 patients were women with a high predominance in early adulthood.

Limitations: This is an understudied and underreported clinical entity. Therefore, limitations include availability of case reports and lack of prior research available on PubMed.

Conclusion: ARP is defined as follicular pustules that occur and remit without treatment and within a week of an identifiable trigger, predominantly affecting women. Consolidating reports of ARP under clear criteria will aid clinical dermatologists in diagnosing this unreported dermatitis.

Keywords: acute and recurrent pustulosis, follicular pustulosis, photo-induced eruption, superficial actinic folliculitis

Background

Acute and recurrent pustulosis (ARP) is an episodic eruption of pustules overlying an erythematous base with neutrophilic infiltrate that occurs and remits without treatment usually within a week. This pustular reaction can be triggered by ultraviolet (UV) light, heat, viral infection, or occur idiopathically. In a previous report by Guzman et al.¹, 3 healthy women who experienced ARP of the face were highlighted (Fig. 1). These patients experienced the rapid onset of follicular pustules localized to the face and neck. These eruptions were noted to be overlying a base of erythema and, at times, associated with mild pruritus. They remitted without treatment or residual scarring rapidly, within the same week. Triggers for recurrence were either sun exposure or viral infection; however, in 1 case, there was no known trigger identified. A small but growing number of cases that share these attributes are being reported, such as the case in a recent Journal of the American Medical Association Dermatology clinicopathological challenge.² As

they accumulate, we feel it is imperative that the parameters of this entity be identified and a designation developed that consolidates the prior reports and allows for better identification and diagnosis of future cases.

Methods

Literature searches for patients with acute pustular dermatitis previously reported as actinic folliculitis, actinic superficial folliculitis, acne aestivalis (AA), and ARP were conducted without a filter for date and in English only. Based on these searches, 21 publications were identified as being of interest. These articles were reviewed in their entirety looking for those cases that fit the following criteria: (a) acute onset of (b) follicularly centered pustular eruption, which (c) spontaneously resolved within days to 3 weeks without scarring, and (d) recurred over time. Of note,

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What is known about this subject in regard to women and their families?

- Acute and recurrent pustulosis is a recurrent eruption of follicularly-based pustules which can be triggered by ultraviolet (UV) light.
- Young women in particular have a very high predominance for developing acute and recurrent pustulosis (ARP).

What is new from this article as messages for women and their families?

- ARP is a rare and underreported pathology with a strong predilection for women.
- Although ARP remits without treatment, some studies have shown prophylactic topical retinoids to be affective.

Table 1
Collection of 23 cases of ARP

| Reference | Age | Sex | Trigger | Onset after trigger | Time to resolution | Site of involvement | Biopsy |
|--------------------------------|-----|-----|------------------|---------------------|--------------------|---|--|
| Guzman et al. ¹ | 29 | F | Sun | 12 h | 5 days | Chin, forehead, cheek, neck | Follicularly centered pustules composed of neutrophils |
| | 30 | F | Viral | 12 h | 5 days | Chin, forehead, cheek, neck | Follicularly centered pustules composed of neutrophils |
| | 61 | F | Idiopathic | N/A | 4 days | Chin, forehead, cheek, neck | Follicularly centered pustules composed of neutrophils |
| Veysey and George ⁹ | 29 | F | Sun | 4–6 h | 7 days | Nasolabial folds, chin, glabella, upper chest | Mixed inflammatory cells around follicle* |
| Rahman et al. ⁸ | 35 | F | Sun | 48 h | 2 days | Upper chest, forehead, neck, behind ears | Not performed |
| | 27 | F | Sun | 12–24 h | 5 days | Forehead, nasolabial folds, chin | Not performed |
| Jaeger et al. ¹⁰ | 30 | M | Heat | 24–36 h | 10 days | Back, shoulders, upper chest | Follicularly centered pustules composed of neutrophils |
| | 20 | F | Sun | <24 h | 7 days | Cheeks, chin, nose | Follicularly centered pustules composed of neutrophils |
| Brokamp and Chung ² | 42 | F | Sun | 6–24 h | 3 days | Chin | Not performed |
| Norris and Hawk ¹² | 30 | F | Sun | 12–24 h | 3–4 days | Chin, upper lip | Not performed |
| | 19 | F | Sun | 24 h | 10 days | Chest, neck, shoulders, upper back, extensor surface of arms | Follicularly centered pustules composed of neutrophils |
| Nieboer ³ | 26 | M | Sun | 25 h | 10 days | Chest, neck, shoulders, upper back, extensor surface of arms | Follicularly centered pustules composed of neutrophils |
| | 31 | M | Sun | 48–72 h | 5–7 days | Upper trunk, shoulders, arms, neck | Follicularly centered pustules composed of neutrophils |
| | 31 | M | Sun | 48–72 h | 5–7 days | Upper trunk, shoulders, arms, neck | Follicularly centered pustules composed of neutrophils |
| Butt et al. ⁴ | 28 | F | UV-A irradiation | 2 h | 6 days | Chin, nose, cheeks, upper lip | Not performed |
| | 34 | F | UV-A irradiation | 15 min | 5 days | Face, neck, upper trunk, scalp | Not performed |
| | 38 | F | UV-A irradiation | Unknown | 4 days | Chin, cheeks, forehead, neck, chest | Not performed |
| | 25 | F | UV-A irradiation | 72 h | 2–3 weeks | Forehead, cheek, chin, neck | Not performed |
| | 44 | F | UV-A irradiation | 2–3 h | 1–2 weeks | Forehead, chin, neck, upper trunk | Not performed |
| | 25 | F | UV-A irradiation | 30 min | 5 days | Face, neck, upper trunk, occipital hairline, extensor surface of arms | Not performed |
| | 30 | F | UV-A irradiation | 24 h | 7 days | Forehead, cheek, neck | Not performed |
| | 36 | F | UV-A irradiation | 12 h | 2–3 days | Forehead, cheeks, nasolabial folds, chin | Not performed |
| | 27 | F | UV-A irradiation | 2 days | 4 days | Face, upper chest | Not performed |
| | 37 | F | UV-A irradiation | 1 week | 3 weeks | Chin, shoulders, thighs | Not performed |

ARP, acute and recurrent pustulosis; F, female; M, male.

Illustration of the characteristics of ARP within 23 patients described in the literature, with 20 out of 23 being women.

*Veysey and George describe their patients to have pustule formation; therefore, we consider the histologic report to be from an older lesion.

articles were excluded if treatment was required for resolution, pustules were not of neutrophilic origin or were associated with infection or the episodes lasted for extended periods of time (>3 weeks). After exclusions, we were left with 9 articles describing 23 patients with the characteristics of ARP.



Fig. 1. Visual representation of ARP of a patient previously described in Guzman et al.¹ ARP, acute and recurrent pustulosis.

Results

Of the 23 patients we identified, 20 were women (Table 1). The average age at report was 31 years, with 18 out of 23 patients' age of onset in their twenties. Triggers of ARP were found to be sun exposure, heat, viral infections, and none in 1 case, as shown in Table 1. In all patients, a localized area of involvement occurred. The areas involved had overlap, so it was found that 19 had facial lesions, 13 neck lesions, and 11 truncal lesions. Table 2 summarizes this relatively homogenous group of cases.

Discussion

Most prior cases reported were of the sun-induced variety and were designated actinic superficial folliculitis. This was first described by Nieboer et al.³ in 1985, who reported 2 patients as having the acute onset of fine pustules of the trunk and arms after sun exposure with spontaneous rapid resolution. Biopsies revealed follicularly based neutrophilic infiltrates. Butt et al.⁴ found that UV-A induced the lesions both experimentally and clinically as it could be initiated by exposure to the sun through window glass. Although the pathogenesis was not clearly defined, it was hypothesized that UV-A incites epidermal thickening and resultant occlusion of follicles.⁴

While some diseases may appear similar to ARP, including, acne aestivalis, acute localized exanthematous pustulosis, and

Table 2**Summary of previously reported cases of ARP**

| Sex ratio (male:female) | 1–6.67 |
|--------------------------------------|--|
| Age of onset of first eruption | 78% of patients' onset in their twenties |
| Time to onset after trigger exposure | 29.5 h (range 15 min–7 days) |
| Site of involvement | 82.6% head, 56.5% neck, and 47.8% chest |
| Time to clearance | Average of 7 days (range 2–21 days) |

ARP, acute and recurrent pustulosis.

ARP has a strong predilection for women beginning in early adulthood with lesions primarily on the head and neck.

polymorphic light eruption, several key distinctions exclude them. acne aestivalis differs from ARP in that it presents as papules instead of pustules, and the lesions can last weeks to months with potential scarring. Under histologic examination, early lesions show necrosis within the hair follicle, that eventually reencapsulates and causes epithelial thickening. Additionally, it is unclear what factors trigger the onset.⁵ Acute localized exanthematous pustulosis is an acute pustular eruption with quick resolution. However, the inciting factor is a drug that when identified and avoided does not recur. The histology is nonfollicular, and patients may be quite symptomatic with fever.⁶ Polymorphic light eruption can also be excluded from ARP based on the severity of pruritis, polymorphic clinical findings, and different histologic findings, showing lymphoid infiltrate around vasculature within the dermis.⁷

Conclusions

We propose the criteria to be used to diagnose ARP to include an acute eruption of monomorphic pustules overlying an erythematous base in a localized distribution. Usually, it involves the head and neck or the trunk. On histology, perifollicular neutrophilic infiltrate is present. Finally, the lesions typically self-resolve within the same week of onset without treatment, and there is no evidence of residual scarring. Interestingly, ARP disproportionately affects women, as shown in Tables 1 and 2, with age of onset usually in early adulthood. Although the exact pathogenesis is unknown, UV-A, viral illness, and heat are known environmental triggers. Treatment is not needed for ARP; however, Rahman et al.⁸ reported that prophylactic topical retinoids kept their 2 patients in remission throughout the year regardless of sun exposure.

In the current literature, instances of ARP have been described, but based on slightly differing clinical presentations they have been identified by different names. Creating new titles for this underreported pustular dermatitis may not change the clinical course for the patient; however, it can cause confusion for dermatologists by having multiple names for the same pathology. Rather than generating new categories for subtle variability in

these findings, grouping them into one category, ARP, will aid dermatologists in its recognition and diagnosis. Along with this consolidation of terminology, future research may be aimed at identifying the exact pathophysiology underlying ARP.

Conflicts of interest

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Author contributions

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