A dermatomal distribution of acute generalised exanthematous pustulosis

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Summary

We report an unique case of acute generalised exanthematous pustulosis in a 71-year-old Caucasian male who presented in a symmetrical and almost dermatomal distribution. As the affected areas resolved, previously unaffected became erythematous, in an inverse pattern. We suggest a dermatomal/linear distribution of AGEP should be considered in an atypical presentation.

KEY WORDS: AGEP; acute generalised exanthematous pustulosis; drug eruption; phenotype.

Introduction

We report an interesting case of acute generalised exanthematous pustulosis (AGEP), which presented in a stripe like, almost dermatomal distribution.

Case Report

A 71-year-old Caucasian male presented with increasing dyspnoea, pleuritic chest pain and a positive troponin on the background of chronic obstructive pulmonary disease (COPD) and severe congestive cardiac failure secondary to ischaemic cardiomyopathy. Upon arrival, he was found to be febrile (37.8°C) and in respiratory distress. Clinical examination was consistent with lower respiratory tract infection and this was supported by investigations (chest X-ray, blood tests, arterial blood gas, CTPA). He was admitted and treated for community acquired pneumonia complicating COPD. He was given a stat dose of IV 500mg azithromycin and commenced on IV 1.2g of benzylpenicillin QID and 37.5mg of prednisolone. On the following morning, he was given 100mg of oral Doxycycline.

On day 3 of admission, he was noted to have a pruritic confluent blanching erythematous eruption affecting the lower limbs (Figures 1, 2, 3a, b) which subsequently spread to involve upper limbs, buttocks, flexural groin; sparing abdomen, chest, back, genitals and an oblique line over the mid thighs (Figure 1). The eruption was strikingly symmetrical over the flexural surfaces of the groin and anterior thighs extending to the buttocks. Multiple pinpoint pustules were present over the hips. Nikolsky sign was negative and there was no evidence of mucous membrane involvement or angioedema.

At this point, further history revealed that he had received doxycycline and IV benzylpenicillin in 2012 for an infective exacerbation of COPD. He had developed...
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A fine, widespread, pruritic macular eruption, which had been attributed to doxycycline. He had been discharged on amoxicillin/clavulanic acid.

Our patient described desquamation and resolution after 1-2 weeks.

A biopsy taken from a pustular area on the right buttocock revealed pustular dermatitis featuring subcorneal collections of neutrophils with negative direct immunofluorescence, in keeping with AGEP. Doxycycline was ceased and our patient was commenced on topical steroids and emollients for symptomatic control. He received 2g of IV vancomycin on Day 7 of admission. His skin and chest symptoms continued to improve over the subsequent 9 days and he was discharged home on a tapering dose of prednisolone.

He represented two days later with recurrent respiratory compromise. Two weeks after the initial eruption, there was clearance of previously erythematous areas and involvement of previously unaffected areas (in an inverse pattern).

Discussion

AGEP is a rare drug related eruption. It is estimated to affect 1-5 cases per million population and commonly presents as erythematous plaques and papules studded with numerous pin point, sterile, non follicular pustules 2-7 days after exposure to a medication (1). It is associated with multiple medications including penicillins, macrolides, sulphur antibiotics, anti-epileptic medications and anti-hypertensives (2). Its onset is often marked by fever and neutrophilia; and in 1/3 of patients, eosinophilia (1). Histopathologically, AGEP typically demonstrates spongioform subcorneal/ intraepidermal pustules, marked oedema of the papillary dermis and perivascular infiltrates with neutrophils and some eosinophils (3). Its distribution characteristically begins in the intertriginous areas or in the face.

The literature has described atypical presentations of AGEP including Toxic Epidermal Necrolysis-like AGEP, AGEP- drug induced hypersensitivity syndrome (DIHS) overlap and localised presentations (2).

Figure 2 - Symmetrical confluent blanching erythematous rash (posterior thighs) with multiple pustules.

Figure 3a,b - a) Confluent blanching erythematous rash affecting right foot; b) Confluent blanching erythematous rash affecting left foot.
To our knowledge, no reports of a similar phenotype of AGEP have been published.

**Conclusion**

AGEP is a rare drug related eruption that can present with varying phenotypes and clinicians should be guided by a thorough history, clinical examination and histological findings in order to make an accurate diagnosis. We suggest a dermatomal/linear distribution of AGEP should be considered in an atypical presentation and patients should be monitored closely for a delayed reaction in previously unaffected areas.

**References**