An unusual presentation of acute generalized exanthematous pustulosis

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Sri Lanka Journal of Dermatology, 2019/2020, **21**: 51-53

Abstract

Introduction: Acute generalized exanthematous pustulosis (AGEP) is a type of cutaneous adverse reaction mostly due to drugs. Typically it presents as sterile pustules over the face or in the major flexures with fever. Herein we report a case of AGEP with unusual clinical presentation.

Case report: A 60 years old lady with diabetes and multiple drug allergies, presented with sudden onset multiple pustules over extensor aspects of upper and lower limbs with lakes of pus, vesicles and bullous lesions.

After establishing the clinical diagnosis of AGEP, patient was started on oral prednisolone to which patient responded well. Histo-pathological features of skin biopsy are compatible with AGEP.

Discussion: The 3 differential diagnosis which were made accordingly, include AGEP, bullous drug reaction, acute generalized pustular psoriasis. However careful analysis of past medical history and the evolution of skin lesions, aided to make the clinical diagnosis AGEP which was later confirmed by histo-pathology.

Introduction

Acute generalized exanthematous pustulosis (AGEP) is being considered as a type of severe cutaneous adverse reaction. This T cell based, neutrophil mediated type IV d hypersensitivity reaction is mostly due to drugs including antibiotics (eg: amino-penicillins, cephalosporins, sulphanomides), anti fungals (eg: terbinafine) and calcium channel blockers (eg: diltiazem).

Typical clinical manifestations of AGEP include numerous small (<5 mm), non-follicular, sterile pustules which arise in a background of oedema and erythema. These lesions begin on the face or in the major inter-triginous zones with rapid dissemination and associated with fever.

Herein we report a case of acute generalized exanthematous pustulosis with unusual clinical presentation.

Case report

A 60 years old lady with diabetes presented with sudden onset vesico-pustular skin eruption over both arms, forearms, thighs and upper part of legs and fever for 3 day duration. She has consumed several drugs other than her routine drugs, which were prescribed for management of lower respiratory tract infection, prior to the onset of skin eruption. On admission she was unable to produce prescription notes given by the general practitioner. She is known to be allergic for several classes of drugs including penicillins and macrolides. History also revealed a single episode of annular pustular eruption over erythematous skin, developed one year ago in which the cause was not identified or determined.

Examination revealed multiple pustules over erythematous background involving extensor aspect of distal arms, proximal forearms, thighs and upper part of legs bilaterally (Figure 1 & 2). Some of these pustules have coalesced to form large lakes of pus. Multiple different sized vesicles and bullous lesions filled with clear, straw coloured or purulent fluid were also observed over the involved areas. Further multiple erosions with evidence of possible secondary bacterial infections were also noted.

After establishing the clinical diagnosis of acute generalized exanthematous pustulosis, patient was started on oral prednisolone. Routine haematological investigations later revealed neutrophilic leukocytosis.

Histology of skin biopsy revealed a superficial intraepithelial pustule with mild spongiform pustulation at the margin with exocytosis of neutrophils adjacent to the pustule and an oedematous papillary dermis with heavy mixed inflammatory cell infiltration. These histological features were compatible with the diagnosis of acute generalized exanthematous pustulosis (Figure 3 & 4).

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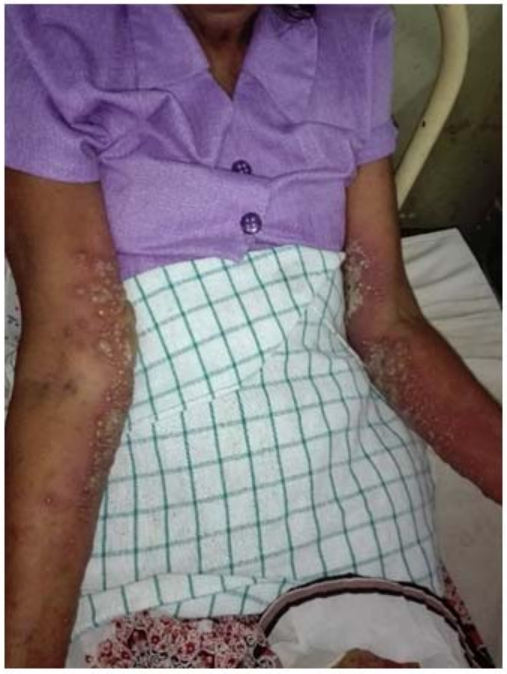

Figure 1.

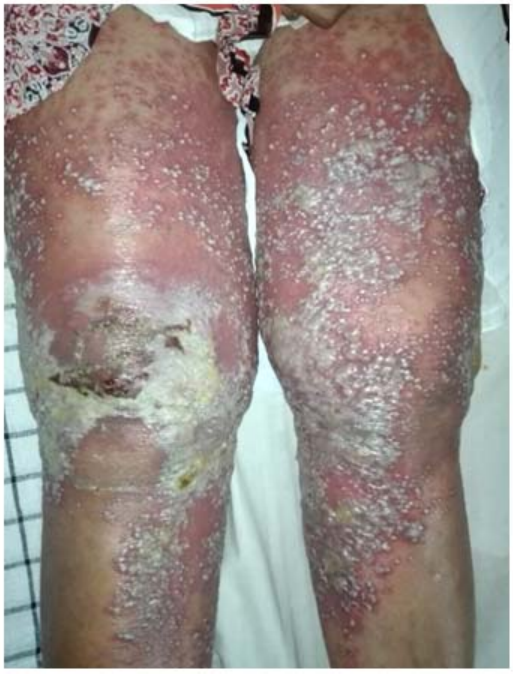

Figure 2.

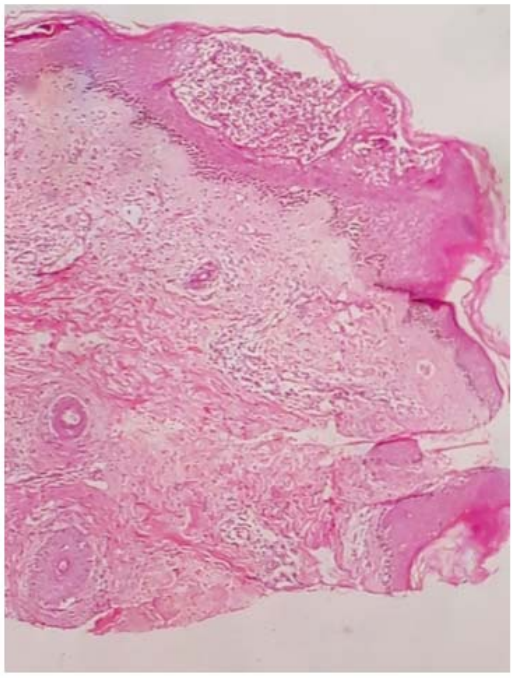

Figure 3.

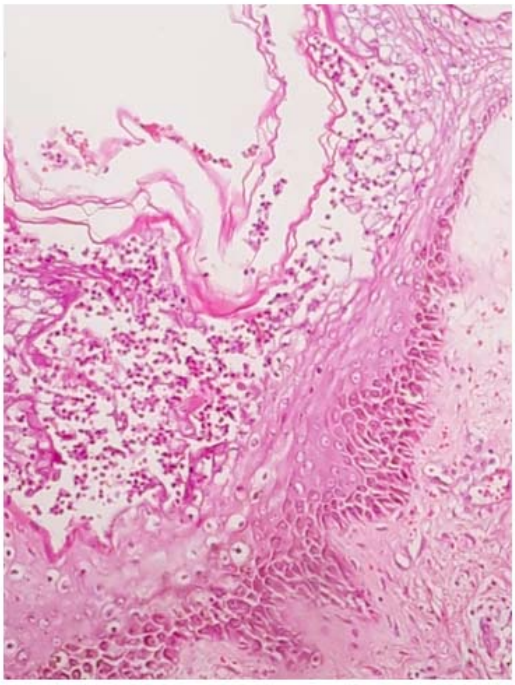

Figure 4.

Patient responded tremendously for oral prednisolone and the steroid was tapered accordingly in parallel with patient's clinical improvement, in a period of 4 weeks (Figure 5 & 6).

Retrospective analysis later revealed co-amoxyclav, a drug which was known to be allergic to her, was included in drugs which were consumed by the patient prior to the skin eruption.

Figure 5.

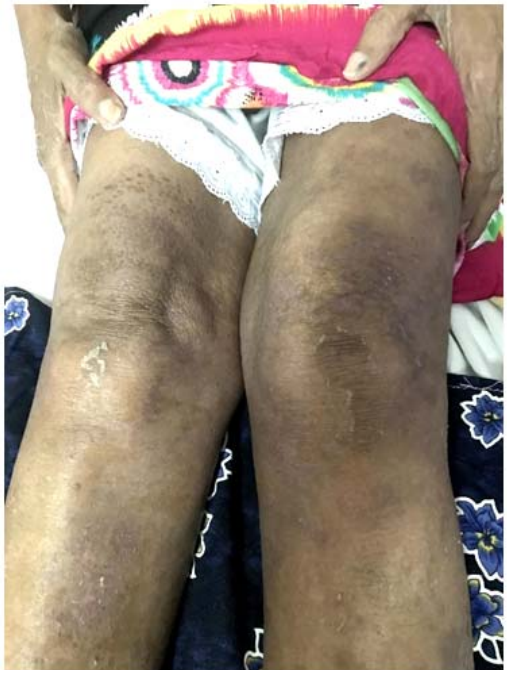

Figure 6.

Discussion

This patient's initial clinical presentation was abrupt onset vesico-pustular skin eruption over extensor aspect of all limbs. With the background of multiple drug allergies and prior consumption of several drugs probably including antibiotics, the first consideration should be given to a cutaneous adverse drug reaction. Since the eruption composed of mixture of pustules, vesicles and erosions, differentiation of acute generalized exanthematous pustulosis from bullous drug reaction is quiet challenging. Matters were further complicated by the distribution of skin lesions which was entirely different from the usual pattern seen in acute generalized exanthematous pustulosis.

Further the previous episode of annular shaped pustular eruption and the presence of lakes of pus in this episode pointed towards acute generalized pustular psoriasis which should be considered as a differential diagnosis.

However in the absence of a past history of chronic plaque psoriasis, careful analysis of the evolution of skin lesions from the onset and adverse effects profiles of probable drugs she may have ingested, made the clinical diagnosis of acute generalized exanthematous pustulosis most likely.

This clinical diagnosis was well confirmed by the skin biopsy which showed typical histopathological findings of acute generalized exanthematous pustulosis without any evidence of psoriasiform hyperplasia.

It was also well supported by the tremendous response shown by the patient for oral steroids.

Therefore in conclusion, this can be considered as a case of acute generalized exanthematous pustulosis with unusual presentation.

References

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