ACUTE GENERALIZED EXANTHEMATOUS PUSTULOSIS PRESENTING AS DISTRIBUTIVE SHOCK WITH MULTI-ORGAN FAILURE

GEORGE ELKOMOS-BOTROS MD* NIRVI SHAH AND MINA MAKARYUS

INTRODUCTION: Acute Generalized Exanthematous Pustulosis (AGEP), or Pustular Drug Eruption, is a rare cutaneous condition most commonly induced by drug or toxin exposure. AGEP is manifested pathologically by acute sterile intraepidermal pustular eruptions and is a rare presentation of febrile drug reaction. Prior cases have described this pathology as similar in presentation to septic shock or toxic epidermal necrolysis. Vancomycin, antiepileptic agents, non-steroidal anti-inflammatory medications, antibiotics, and diltiazem have all been identified as possible etiologies of AGEP. Here we present a rare case of AGEP in a patient with no prior medication exposures presenting with shock and multisystem organ failure.

CASE PRESENTATION: An 86 year-old woman presented to the Emergency Department (ED) with report of recurrent high grade fevers and progressive maculopapular rash for one week. The patient had previously been evaluated by outpatient dermatology. Biopsy was performed and results were pending at the time of initial ED evaluation. Symptoms were additionally refractory to topical steroid therapy. ED course was significant for tachycardia (110 beats per minute), hypotension (84/48 mmHg), fever (103°F), bandemia (15%) and acute kidney injury (AKI). The patient was admitted to the Intensive Care Unit with distributive shock of unclear etiology. The patient received intravenous fluid resuscitation, norepinephrine infusion, vancomycin, and piperacillin/tazobactam. She subsequently developed multiorgan failure involving the renal, hepatic, cardiac and respiratory systems. Prior skin biopsy results were obtained and revealed intraepidermal neutrophilic pustules with no evidence of infection. Findings were consistent with AGEP. An extensive evaluation for an infectious etiology was unremarkable. Antibiotic therapy was discontinued and the patient was treated with pulse methylprednisolone therapy with a prolonged prednisone taper. The patient improved on steroid therapy and resolution of multiorgan failure was noted. The patient was subsequently discharged to a rehabilitation facility.

DISCUSSION: AGEP is a rare cutaneous condition typically associated with medication or toxin exposure. We present a unique case of idiopathic AGEP in a patient with no prior infectious, toxic or medication exposures. This case is likewise unique with respect to disease severity and only four prior case reports describe AGEP progressing to multiorgan failure and shock.

CONCLUSIONS: This is the first case, to our knowledge, describing the use of pulse dose steroids in the treatment of severe AGEP. Pulse therapy, in this setting, led to complete resolution of multiorgan dysfunction. Determination of the etiology of distributive shock is essential for the prompt and appropriate treatment of patients in the critical care setting.


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