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CASE REPORT

Acute generalized exanthematous pustulosis in a 9-year-old child

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Abstract

Acute generalized exanthematous pustulosis (AGEP) is a rare disease characterized by the onset of non-follicular sterile pustules on a background of edema and erythema, fever, and leukocytosis. **Objectives:** This paper aimed to report the case of a child with AGEP admitted to a pediatric ward for fever and disseminated sterile pustules and diffuse erythema and to review current literature on the subject. **Method:** This case report describes the information collected from the patient's chart, contains a photographic record showing the progression of the disease, and offers a review of current literature on the subject. **Conclusion:** This case report and current literature on the subject stress the relevance of considering AGEP in children suspected for the condition and the importance of interviewing patients and conducting physical examination in order to consider all relevant diagnostic, therapeutic, and prognostic factors.

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INTRODUCTION

Acute generalized exanthematous pustulosis (AGEP) is a rare disease characterized by the onset of non-follicular sterile pustules on a background of edema and erythema preferentially affecting the face and intertriginous areas, fever, and leukocytosis. AGEP is part of a group of diseases categorized as severe cutaneous adverse reactions, with an incidence of 1-5 patients per million/year. The condition progresses favorably in most cases, with a death rate below 5%. The most common causes of the disease are drug hypersensitivity and viral infection[1-3].

This paper reports the case of a child diagnosed with AGEP based on lesion histopathology and findings of fever and generalized maculopapular erythematous rash. Since fever and rash are cited in the diagnostic criteria of numerous conditions, knowledge of AGEP is required in the rendering of accurate diagnosis and prescription of adequate therapy[4].

CASE REPORT

A 9-year-old girl was admitted to the pediatric ward of our center with fever and pruritic generalized maculopapular rash of cephalocaudal progression initiated three days before hospitalization. She was on steroids and antihistamines, but showed no signs of improvement. The patient was in good health before the onset of symptoms. Her parents said she did not have allergies and that she had not been on medication. She was afebrile upon admission and had maculopapular erythematous rashes predominantly on her trunk and proximal portions of her limbs, with multiple non-follicular sterile pustules each with a diameter of approximately three millimeters (Figure 1), mild oropharyngeal hyperemia, and palatal petechiae.

Laboratory tests showed she had leukocytosis with a predominance of segmented neutrophils, elevated C-reactive protein, and altered liver function. The patient was prescribed an immunosuppressive dose of corticosteroids and was kept on antihistamines. A dermatologist from the hospital saw the patient and considered she might have AGEP, infection by staphylococcus, or pustular psoriasis. The patient was prescribed oxacillin and moisturizing cream. She underwent a skin biopsy.

One week into hospitalization her condition deteriorated with confluence of pustular lesions and lesion desquamation (Figure 2). The patient improved and was discharged a week later on cephalexin and antihistamines. Her skin biopsy showed dermatitis with lymphocytes and occasional eosinophils and an epidermis with foci of erosion.

DISCUSSION

AGEP is characterized by an abrupt onset of non-follicular pustules on a background of erythema associated with fever and leukocytosis. Estimates indicate that 90% of the cases are connected to medication, with reports in the literature also citing cases triggered by viral, bacterial, and parasitic infection, dietary supplements, mercury hypersensitivity, radiation, and spider bites, with viral infection and vaccines ranking as the most frequent causal agents in pediatrics[5].



Figure 1. Aspect of the lesions present at the patient's admission.

Manifestations usually start within ten days of exposure to the triggering factor and resolve spontaneously 7-14 days after the removal of the causal agent[6]. Britschgi et al. (2001) reported elevated levels of IL-8, a chemokine associated with neutrophil recruitment produced by keratinocytes and mononuclear cells in cutaneous inflammatory infiltrate, in individuals with the disease. The authors concluded that AGEP appears to express a reaction in which drug-specific T-cells trigger a positive CD4 and CD8 immune response with IL-8 overexpression.

Classic clinical presentation consists of generalized rash of abrupt onset, with hundreds of non-follicular sterile pustules on the face or intertriginous areas showing a cephalocaudal dissemination pattern, preceded or accompanied by fever and pruritus and followed by lesion desquamation. In some cases the confluence of pustules may mimic Nikolsky's sign and mislead physicians into thinking their patients have toxic epidermal necrolysis (TEN). Although systemic and mucosal



Figure 2. Aspect of lesions after one week of evolution.

involvement is uncommon, mild self-limiting mucosal involvement has been reported in 20% of the cases[8].

Complementary tests indicate leukocytosis with a predominance of neutrophils or eosinophils, with possible transient renal impairment and elevated transaminase levels[9]. Histology reveals intracorneal, subcorneal, or intraepidermal pustules with dermal papilla edema of varying degrees, and a perivascular infiltrate with neutrophils or eosinophils. Patients may also present leukocytoclastic vasculitis or focal keratinocyte necrosis. Diagnosis is based on clinical and histology findings. The EuroSCAR group developed a validation score that is particularly useful in situations where it is not possible to correlate episodes to a triggering factor[10].

The patient described in this case did not have a recent track record of medication or immunization, which were thus excluded from the list of possible triggering factors. However, the patient had oropharyngeal hyperemia and palatal petechiae upon admission. Since mucosal involvement is uncommon in AGEP and the alterations described in oral examination were atypical for the condition at hand, the authors suspected that the factor triggering these findings might have been a mild upper airway viral infection unnoticed by the patient or her family.

The main purpose of treatment is the withdrawal of the causal agent. Given the benign progression and self-limiting character of AGEP, support therapy based on topical steroids and disinfecting solutions in the pustular phase and moisturizing cream in the event of desquamation with the addition of antipyretics is usually satisfactory. In cases of extensive disease, empirical administration of systemic steroids may be indicated despite the lack of evidence supporting the alleged decreases in disease duration and the fact that reports have described cases of steroid-induced AGEP[11].

CONCLUSION

This case illustrates the importance of knowing the etiology of AGEP, the associations the condition may have with different drugs, the clinical manifestations inherent to the disease, and the need to manage pediatric patients with the disease in a hospital setting. Since fever and rash are cited in the diagnostic criteria of numerous conditions, recognizing AGEP from other conditions is required in the prescription of adequate therapy.

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