

Pustular Psoriasis: An Rare Infantil Pustular Eruption

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Abstract

Generalized pustular psoriasis, a severe form of psoriasis, is very rare in children. It is characterized by a fever of several days' duration, together with the sudden appearance of sterile pustules, 2 - 3 mm across, over the trunk and extremities. A safe and consistently effective treatment regimen has not been identified. We aim to report a case of a 14-year-old child with this unusual variant of psoriasis.

Keywords: Pustular Eruption; Psoriasis; Pustular Psoriasis; Child

Introduction

Generalized pustular psoriasis (GPP) in infancy is extremely rare [1]. It is a cyclical process evolving by thrusts and remissions [2]. It manifests as sterile erythematous pustules on a glossy erythematous base that confluence and end in the desquamation phase. The acute rash is painful and preceded by a feeling of fever and discomfort [3]. A safe and consistently effective treatment regimen has not been identified [4]. We report a case of this rare entity occurring in a 14-year-old child.

Case Presentation

A 14-year-old child, with no significant pathological history, presented with 10 days' history of an erythematous pruriginous lesions of the neck, becoming infiltrated and sprinkled with pustules, with rapid extension to the trunk, genital area, folds and lower limbs. The symptomatology was evolving in a context of pyrexia and conservation of the general state. The examination had revealed multiple erythematous plaques, well limited, confluent in large patches, dotted with pinhead's pustules with lactescent content. We have noted a desquamation by location. The lesions occupy 30% of the cutaneous surface (Figure 1). The examination of the mucous membranes, the nails and the scalp was without anomalies. The ganglionic areas were free. The biological examinations were normal except hyperleukocytosis with neutrophil polynuclear predominance. Cutaneous histology was compatible with pustular psoriasis. The child was initially put on magistral preparation with a slight improvement (Figure 2) then oral retinoids were initiated with a good evolution. The current decline is 3 months.

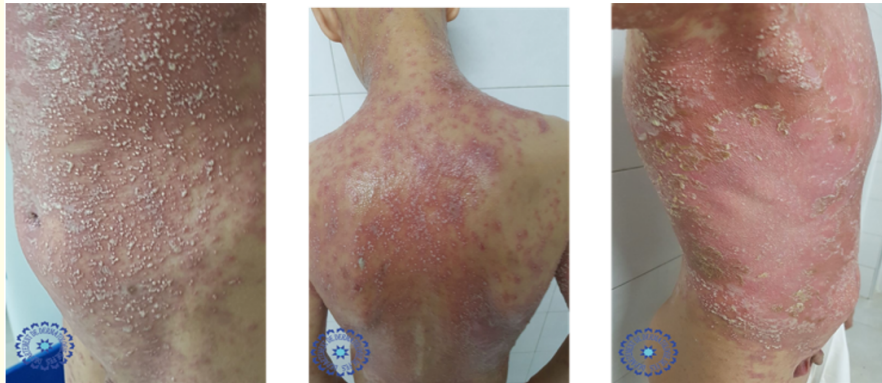


Figure 1: Multiple erythematous plaques confluent in large patches, dotted with pinhead's pustules.



Figure 2: After one week of treatment.

Discussion

GPP or von Zumbusch psoriasis is a rare type of psoriasis [5]. Infantile pustular psoriasis (IPP) is a very rarer form of GPP. Fewer than 200 IPP cases are reported in the literature [3]. Usually it starts between 4 and 6 years but there may be a wide range and each year of the first decade has been represented; 27% started in the first year of life [6]. Boys outnumber girls [1]. The most common subtype of psoriasis in children is generalized plaque psoriasis, with well-demarcated pink plaques with overlying silver scale commonly on the elbows,

knees, and scalp. The second most common pediatric subtype is guttate psoriasis, which presents as an eruption of individual drop-like red, scaly papules and small plaques. The pustular forms of psoriasis are the least common subtype [2]. Various precipitating factors have been reported to trigger or flare acute GPP, including corticosteroid use and withdrawal, pregnancy (also termed impetigo herpetiformis), upper respiratory tract infections, stress, nonsteroidal anti-inflammatory drugs, terbinafine, ustekinumab, tumor necrosis factor (TNF)- α inhibitors, and methotrexate [7]. Clinically, it is an acute variant of pustular psoriasis that initially presents as sterile pustule formations on a bright erythematous base, 2–3 mm across, over the trunk and extremities. that coalesce into lakes, subsequently forming the second phase consisting of desquamation. The acute pustulation is painful and usually preceded or accompanied by pyrexia and malaise [3,5]. Histopathological studies revealed subcorneal pustules filled with neutrophils, acanthosis and superficial perivascular lymphocytic infiltrates [4]. The main complications are bacterial superinfection, dehydration, and sepsis [3]. A limited therapeutic armamentarium exists for children due to different inherent risks [2]. In addition to topical corticosteroids, other products found to be effective in childhood GPP include tar preparations, PUVA, sulfapyridine, systemic steroids, methotrexate, erythromycin and oral etretinate has been reported to be effective [3]. The treatment choice should be based on her age and general condition, and the disease severity [5]. Robinson, *et al.* proposed systemic treatment with acitretin, cyclosporin, methotrexate and etanercept as the first-line therapy for infantile GPP. Adalimumab, infliximab and ultraviolet B phototherapy were proposed as second-line therapy. A small dose of etretinate (<1 mg/kg per day) is considered as an appropriate therapy for infants with severe or recalcitrant GPP. However, this medication has potential toxicity on the skeletal system [8]. In Japan, the GPP treatment guidelines recommend cyclosporin, etretinate, systemic steroid and anti-tumor necrosis factor- α (TNF- α) agents as standard therapy for infantile GPP. Cyclosporin is regarded as the better initial choice, as etretinate has a risk of premature epiphyseal closure, and its administration should be intermittent to prevent side-effects. For infants, the guidelines allow us to use TNF- α inhibitors, such as infliximab, for emergency treatment if no other systemic therapy can be used during the acute aggravation phase accompanied by systemic symptoms. When using TNF- α inhibitors, it is recommended to switch to other non-biological drugs immediately when the symptoms improve [5].

Conclusion

Generalized pustular psoriasis is a rare and unusual disease in children who are recalcitrant and difficult to treat. We present our case to sensitize that it should be considered in the differential diagnosis of pustular eruptions in children.

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