CASE REPORT

Infantile generalized pustular psoriasis: a case report and review of the literature

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ABSTRACT

Infantile generalized pustular psoriasis (von Zumbusch) is a rare form of psoriasis. Here, we report a 3-month-old Taiwanese girl who was treated with Chinese herbal medicine for abdominal fullness starting at 1 month of age. Around 1 month into the treatment, sudden onset of generalized erythematous plaques with scales and pustules was noted which lasted for 1 month. Histopathology of the skin lesion revealed accumulation of polymorphs within the parakeratotic stratum corneum and subcorneal neutrophilic pustules. Based on clinical presentation, histopathology, and negative bacterial cultures, our final diagnosis was infantile generalized pustular psoriasis. Both drug use and physical stress (abdominal fullness for 1 month) may play a role in inducing generalized pustular psoriasis in this patient. The patient responded well to topical treatment, and the disease is under control with only occasional flares.

In summary, because infants are at a stage of rapid growth, timely diagnosis and tailored therapy according to response are essential to ensure that the disease process does not interfere with normal development.

Introduction

Psoriasis is a common T-lymphocyte mediated inflammatory dermatosis in children. Infantile psoriasis is rare and accounts for only 3.5%–16% of childhood psoriasis.1–4 Various provoking agents, including infections, irritating topical treatment (Koebner phenomenon), drugs,5,6 and stress,7,8 have been suggested. Early diagnosis with appropriate treatment is necessary to prevent the affected infants from severe complications, such as bacterial superinfection, dehydration, and sepsis.9

Case report

A 3-month-old Taiwanese girl born by normal spontaneous vaginal delivery at 39 weeks presented at our pediatric emergency department with generalized erythematous plaques (Figure 1) covered with scales and pustules (Figure 2) over scalp, trunk, and face for about 1 month.

She was fed with unknown Chinese herbal medicine for treatment of abdominal fullness starting at 1 month of age. Around 1 month into the treatment, the baby began to develop erythematous changes of the skin starting on the scalp and spread to the trunk, four extremities, and face. Multiple pustules and excoriations were conspicuous within the erythrodermic and scaly skin. The baby was febrile during the course of skin changes.

Because of fever and appearance of skin eruption, herbal medication was discontinued; however, skin lesions continue to progress for additional 1 month. She was then admitted to pediatric ward under the impression of generalized eczema with severe secondary infection.

Tracing back her illness, the infant had no preceding history of infection. Her personal and family history were unremarkable. There were no mucosa involvements or nail changes. Routine laboratory studies revealed elevated C-reactive protein, whereas complete blood count, serum liver, kidney, and lipid parameters and electrolytes were all within the normal limits. Analysis of her residual herbal medication failed to identify any toxin or drug, including corticosteroids, nonsteroidal anti-inflammatory drugs, and benzodiazepines. Gram's stain and potassium hydroxide examination revealed no bacteria or fungi. Bacterial cultures from pustules and blood were all negative. Herpes simplex virus and varicella zoster virus isolation from pustules were unremarkable.

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Dermatologists were then consulted for evaluation of the infant's condition. Initial differential diagnosis included infantile generalized pustular psoriasis, seborrheic dermatitis with secondary infection, and acute generalized exanthematous pustulosis. Skin biopsy was done, which revealed hypogranulosis, accumulation of polymorphs within the parakeratotic stratum corneum, subcorneal neutrophilic pustules of varying sizes, and lymphohistiocytic perivascular infiltrate in the superficial dermis (Figure 3). Periodic acid-Schiff studies and direct immunofluorescence staining were unremarkable. Combining the course of illness, histopathology, and laboratory findings, the diagnosis of infantile pustular psoriasis was established.

Oxacillin was prescribed to prevent infection. Twice daily application of emollients and hydrocortisol lotion was administered. Oxacillin was discontinued after 4 days. Fever with widespread erythema and pustules relapsed again after 1 week of treatment. Leukocytosis, neutrophilia, and elevated C-reactive protein were noted; bacterial culture from pus and blood were again negative. After applying mometasone furoate cream 0.1% daily for 1 month, pustular lesions disappeared. Twelve months after the initial episode, the patient still experiences scattered flares of disease, which respond favorably to topical steroid.

**Discussion**

Our patient was diagnosed to have infantile generalized pustular psoriasis. On withdrawal of Chinese herbal medicine, chronic, relapsing skin manifestation for more than 1 month excluded acute generalized exanthematous pustulosis. Seborrheic dermatitis with secondary infection appeared unlikely because repeated culture from pustules were negative.

Generalized pustular psoriasis is a rare type of psoriasis first described in 1910 by von Zumbusch. Attacks are characterized by fever that lasts for several days and a sudden generalized eruption of sterile pustules. The pustules are disseminated over the trunk and extremities, including the nail beds, palms, and soles. Severe pustular psoriasis can be difficult to control and requires a potent treatment regimen to avoid life-threatening complications. In a case series reviewed by Shih and Liu, complete remission never occurred in 11 patients (58%). Of those cleared or much improved after therapy, three relapsed within 6 months, and only five maintained remission for more than 6 months.

Unlike psoriasis in adulthood, psoriatic diaper rash with dissemination was the most common type of psoriasis (45%) in the children less than 2 years of age.

Various triggers, including stress, infections, trauma, irritating topical treatment (Koebner phenomenon), and drugs, have been proposed to provoke generalized pustular psoriasis previously. Flexors and diaper regions, which are friction/trauma-prone sites, have been found to be more commonly involved in infants and young children. However, our patient's skin lesion started on the scalp and subsequently spread to the trunk and four extremities, with only minimal involvement of the diaper area. Therefore, the increased bacterial and fungal growth because of occlusion by diaper is not likely to be the trigger factor in this patient. In our patient, abdominal fullness lasting for 1 month and treatment with Chinese herbal medication suggest that the infant has undergone a great physical stress. Withdrawal of oral corticosteroids and physical stress-induced infantile generalized pustular psoriasis was highly suspected on the dermatologist's consultation. Although analysis of Chinese herbal medicine failed to identify presence of toxin or drug, the induction of psoriasis by other potential incipient drug cannot be ruled out. We believe that the exact trigger of generalized pustular psoriasis in our patient involves physical stress and possibly drug use.

The choice of treatment in infantile psoriasis should be determined by age, gender, weight, distribution of the lesions, associated symptoms and signs, and comorbidities. Because of young age, topical emollients and corticosteroids are usually the first option. Other agents are also found to be effective in infantile psoriasis, including topical captopril (cilcaprolene) and topical pimecolimus. For generalized pustular psoriasis, however, the disease course is usually fulminant and prolonged. Once diagnosis is established,
is established, systemic acitretin has proved to be effective and is usually administered immediately to prevent severe complications, such as dehydration and sepsis.\textsuperscript{17,18} However, the major concern is the risk of skeletal toxicity.\textsuperscript{16} Interestingly, in our case, although more than 70% of body surface area was involved, the patient showed good response to topical steroid of medium potency. Our experience indicated that even in infantile generalized pustular psoriasis, stepwise regimen from topical to systemic treatment is warranted.

In summary, infantile generalized pustular psoriasis is uncommon and may pose a diagnostic and therapeutic challenge. We reported an infant with pustular psoriasis associated with physical stress and herbal drug use, whose disease although extensive, responded favorably to topical mometasone furoate cream. As infants are at a stage of rapid growth, timely diagnosis and appropriate treatment is essential to ensure that the disease process does not interfere with their normal development.

References