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CASE REPORT

A Case of Pustular Psoriasis in a Young Boy

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ABSTRACT

We report a case of a 13-year-old boy who presented with acute onset of generalised erythematous skin and patchy areas of pustules for one week duration. He was well until one month ago when he started having small scaly plaques on his scalp and extensors of his legs. During the acute episode, he also had joint pain and bilateral conjunctivitis. Skin biopsy confirmed pustular psoriasis. He developed leucocytosis and transaminitis during the acute phase of the pustular eruption while on acitretin, which was then withheld. Subsequently, treatment with oral cyclosporine induced remission of his skin and joint disease. The case is hereby reported because of rarity of presentation and clinical features. Oral cyclosporine should be considered in patients with generalised pustular psoriasis complicated with transaminitis.

KEYWORDS: Psoriasis, pustular psoriasis, cyclosporine, treatment effectiveness

INTRODUCTION

Pustular psoriasis is a rather uncommon form of psoriasis in Malaysia, representing less than 1.6% of psoriasis cases [1]. We report a case of pustular psoriasis as a first presentation of severe psoriasis, in a young school boy complicated with arthritis and liver impairment. Delay in inducing pustular remission was due to the difficulties in initiating systemic treatment in the presence of acute hepatitis.

CASE PRESENTATION

A 13-year-old boy presented with generalised erythematous skin lesions involving 90% of his body surface area and appearance of pustules over past one week. One month prior to the current illness he had developed small scaly plaques on his scalp and legs. He gave a history of receiving oral diclofenac for a sports injury one week prior to the development of scaly plaques. During the course of the development of scaly lesions, he was treated with oral erythromycin for upper respiratory tract infection and the skin lesions became diffusely erythematous and studded with pustules two weeks later. The lesions were itchy and painful. They were associated with intermittent fever, myalgia, conjunctivitis and joint pain over his hips, right knee, left ankle and lower back. Of note, his mother has systemic lupus erythematosus and is on treatment. He had no family history of psoriasis or atopy.

On examination, his temperature was 37.8˚C, blood pressure was 130/80 mmHg, heart rate was 110 beats per minute and Spo_2 was 100%. There was generalized erythema on the face, limbs and trunk (Figure 1). Thick scaly plaques covered almost the entire scalp. Skin was generally erythematous with total body surface area involvement up to 90%. There were pustules studded on the erythematous patches on his trunk and limbs, with areas of desquamation. There were thick scales on his soles (Figure 2). His nails showed onycholysis, pitting and subungual hyperkeratosis (Figures 3). He had swelling and tenderness over his left ankle. Cardiovascular, respiratory, abdominal examinations were normal.

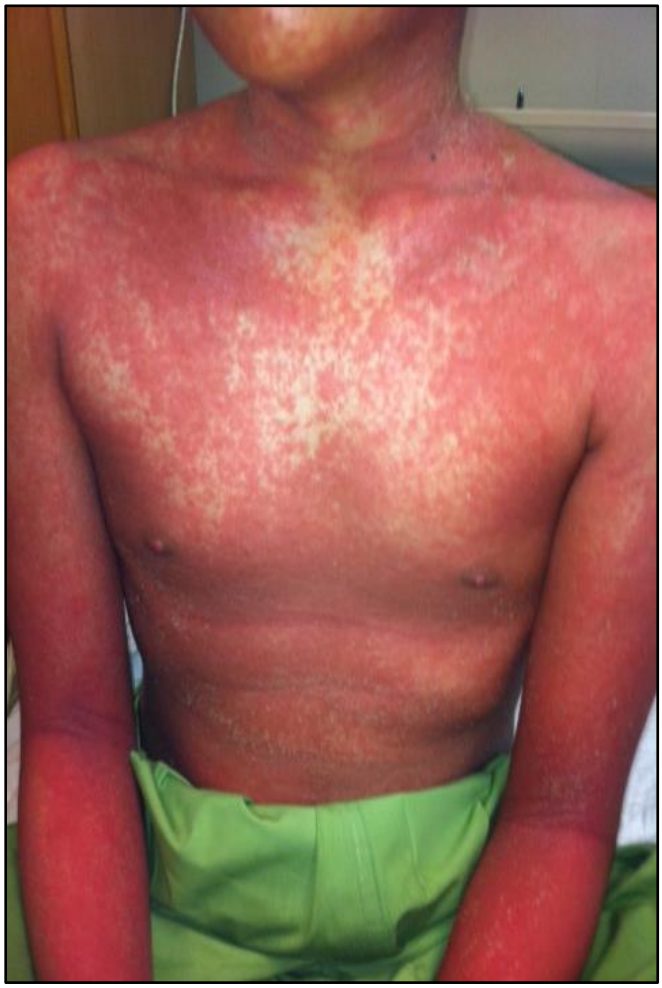

Figure 1 Pustules on erythematous base on the trunk

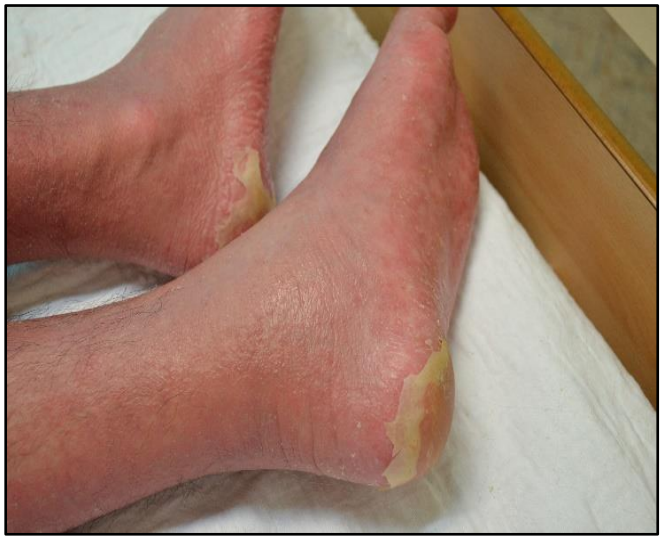

Figure 2 Thickened hyperkeratotic soles

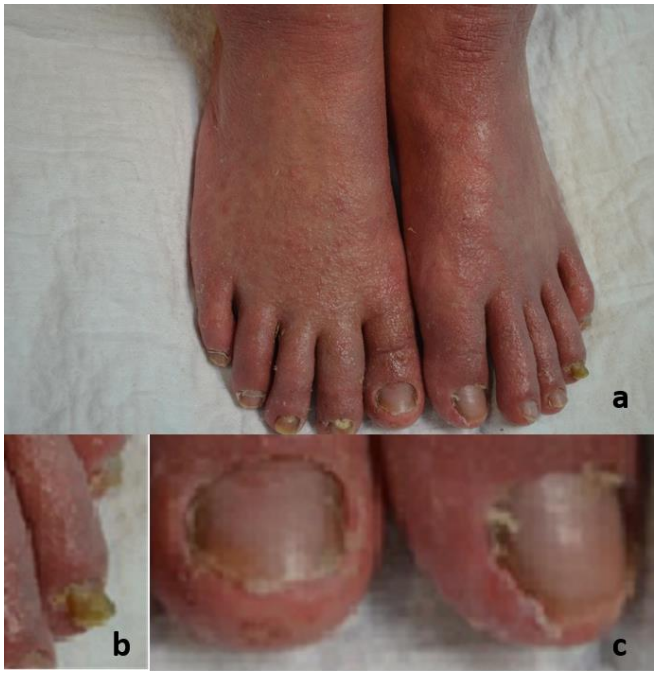

Figure 3 Clinical findings on both feet. a) Pustules on the dorsum of feet b) Subungual hyperkeratosis c) Onycholysis

Baseline blood investigation showed hemoglobin 12.6 g/dL, platelet count 208 X 10^9/L, total white count 10.32 X 10^9/L, alanine transaminase (ALT) 45 U/L, aspartate transaminase (AST) 30 U/L, rheumatoid factor negative, normal renal profile, corrected serum calcium 2.25 mmol/L, ASOT negative, erythrocyte sedimentation rate 20 and C-reactive protein (CRP) of 3.88. His viral screen was negative. ANA and rheumatoid factor were negative. Pelvic radiograph showed left sacroillitis. Chest radiograph was clear. Skin biopsy done 16 days after admission showed parakeratosis, mild acanthosis with elongation and thickening of the rete ridges. There was scattered spongiosis with early pustule formation (spongiform pustule of Kogoj). The papillary dermis was oedematous with mild infiltrate of lymphocytes and some neutrophils around the superficial dermal blood vessels. These results were compatible with the clinical diagnosis of pustular psoriasis.

Treatment proved to be challenging and systemic steroid was started for arthritis and was tapered down after introduction of steroid sparing agents. This patient was started on acitretin and continued a prednisone taper. Liver enzymes became elevated by the fifth day of acitretin therapy. Peak liver enzyme values on day 6 were 368 U/L and 97 U/L for ALT and AST respectively. Elevated liver enzyme levels were accompanied with a rise in CRP to 14.

However, the patient did not develop any symptoms of hepatotoxicity. Acitretin was discontinued and liver enzyme levels returned to baseline within 1 week. At this time, ultrasound of the hepatobiliary system and screening for autoimmune hepatitis was done and was negative. Oral methotrexate was not an option at this point of time for the same reason. He was started on oral cyclosporine 50 mg twice a day. The pustules had remitted after 48 - 96 hours of starting cyclosporine. Three weeks later, the transaminases were restored to normal values and his general clinical condition improved. The high dose of systemic steroids could be tapered down without recurrent pustular flare and the joints pain dramatically resolved. His disease remained stable throughout the outpatient follow up in the first year, but had recurrent flares of generalised pustular psoriasis subsequently in the following 4 years. Currently, patient is awaiting biologics treatment.

DISCUSSION

Generalized pustular psoriasis (GPP) is a serious dermatological disease characterized by fever, chills, rigors, and generalized pustule formation on the skin [2, 3]. The occurrence of GPP in childhood is rare, and fewer than 200 cases have been reported in the literature [4]. Several antigenic factors have been shown to elicit GPP, including withdrawal of steroids, emotional stress, infection, drugs such as lithium and hydrochloroquine, irritative topical therapy (e.g. coal tar), dental and upper respiratory infections, pregnancy, and solar irradiation [4, 5]. Recessive mutations of IL36RN have been identified in majority of GPP patients, without previous history of psoriasis [6]. Furthermore, studies done in Japan have shown that the age of onset of the pustular outbreak in psoriasis naïve patients is earlier and occurs more frequently following infections [3].

This was a case of pustular psoriasis in a young boy who presented with erythroderma which is a rare presentation of pustular psoriasis. The occurrence of mucosal involvement is also rare and was found in this patient who had sterile conjunctivitis [7]. The commoner presentation of pustular psoriasis in Malaysia are fever, painful skin, arthritis and leucocytosis [8].

Possible differential diagnoses include acute generalized exanthematous pustulosis, Gram negative sepsis, subcorneal pustular dermatosis and Ofuji syndrome. Clinical presentation and the histopathology confirmed the diagnosis of pustular psoriasis. GPP is an acute form that presents itself by sudden eruption of sterile papules on erythematous base and is accompanied by severe systemic symptoms and is also known as von Zumbusch.

Notably, this patient also had elevated liver enzymes during the acute phase of the disease which subsequently subsided as his condition improved. Studies have shown that the occurrence of liver enzyme abnormalities in patients with GPP was 47% with an increased leucocyte count and male preponderance [8].

With regards to the treatment options, there is little evidence based information to guide the treatment of GPP. Acitretin, cyclosporine, methotrexate, and infliximab are considered to be first-line therapies [9]. Adalimumab, etanercept, and psoralen plus ultraviolet A are second-line modalities [9]. Studies have also shown that acitretin appears to provide better efficacy in pustular psoriasis than in psoriasis vulgaris as monotherapy [10]. There are also reports of successful use of dapsone in the treatment of GPP [11].

As the patient continued to have flares of pustules and a painful hip joint with transaminitis, which persisted despite discontinuing acitretin, the decision to start an alternative immunosuppressive drug proved to be a challenging one. Studies done in children with pustular psoriasis have shown that cyclosporine is effective and safe for children with a median time to total clearance of 4 weeks [12]. There have also been reports of successful use of cyclosporine in pregnancy with good outcome [13]. The recommended starting dose of 5 mg/kg is associated with a higher degree of clearance in patients with GPP [14]. In this case, oral cyclosporine induced remission of both the skin and joint disease.

The nature of the swollen joint, whether psoriatic or reactive was not well defined in this case from clinical appearance alone. Features favouring the diagnosis of reactive arthritis were younger age of onset, sacroillitis and keratoderma of the soles [15]. This differentiation was important to predict long term

prognosis for this patient as psoriatic arthritis is associated with higher morbidity.

In conclusion, the management of patients with pustular psoriasis generally requires systemic agents such as acitretin, methotrexate, cyclosporine and azathioprine. All of these require special precautions. In managing this patient, the choice of systemic agent proved to be a challenge. Although there are guidelines in managing acute pustular psoriasis, the management decision should be individually tailored according to the clinical scenario.

Conflict of Interest

Authors declare none.

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