CASE REPORT

Pemphigus foliaceus in a child responded to flucloxacillin therapy: Case report

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Abstract  Pemphigus foliaceus is a rare autoimmune blistering disease comprising two major types: endemic and sporadic. The endemic form, also known as fogo selvagem, affects children and young adults in rural Brazil while the sporadic form generally affects the middle-aged and the elderly. Here we report a sporadic form of pemphigus foliaceus in a child in which the skin eruption responded to flucloxacillin. Upon discontinuation of flucloxacillin, lesions reappeared and then cleared upon resuming the antibiotic. Dapsone was thereafter administered with good control.

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1. Introduction

Pemphigus foliaceus is a rare autoimmune blistering disease mediated by IgG4, anti-desmoglein 1 autoantibodies (Qaqish et al., 2009). Patients with PF will have direct immunofluorescence deposits of IgG and C3 within the upper part of the epidermis. There are two major subtypes of PF: endemic and sporadic. The endemic form, mainly seen in rural areas of Brazil, is known as fogo selvagem. Black fly, Simulium nigrimanum, is thought to play a major factor in the disease transmission. The endemic type affects children and young adults while the sporadic type affects the middle-aged individuals and the elderly. The sporadic pemphigus foliaceus is rare among children; thus clinical data and therapy to this group of patients are lacking. We describe a 5-year-old girl with the disease who interestingly responded to flucloxacillin, which has not been described before.

2. Case report

A 5-year-old Saudi girl presented with 1 year history of a persistent, mildly itchy, generalized skin eruption involving face, neck and upper trunk. The skin eruption began at the perioral skin spreading gradually over face and neck for which she was given multiple topical remedies (unknown) by a pediatrician without any improvement. She was then started on oral prednisolone (dose was not specified) with good clearance of the lesions but with recurrence upon tapering the dose. She was kept on prednisolone for several months with significant increase in her weight; thus prednisolone was stopped and she was referred to our dermatology clinic. She presented to us
with similar initial skin rash in the form of multiple polycyclic, serpiginous, crusted, erythematous superficial plaques over her face, neck and upper trunk. Fig. 1. Mucous membranes were not involved. The initial clinical impression was linear IgA bullous dermatosis of childhood. Two skin biopsies were taken for H&E and direct immunofluorescence (DIF). Blood tests were done including complete blood count, blood sugar liver enzymes, urea, electrolytes and antinuclear antibodies. All came within normal values. The child’s family lived far away from the city and while waiting for the biopsy results, the child was started on oral flucloxacillin 250 mg three times a day based on a previous experience with linear IgA bullous dermatosis of childhood (Alajlan et al., 2006). Skin biopsy showed upper epidermal acantholysis with neutrophilic infiltrates. DIF showed epidermal intercellular IgG and C3, confirming the diagnosis of pemphigus foliaceus (PF). Although clinical, histopathological and DIF were consistent with diagnosis of PF, the child showed good improvement of skin lesions within the first week of flucloxacillin therapy. After few weeks while on flucloxacillin, the skin eruption mostly cleared, Fig. 2. After 8 weeks, flucloxacillin was tapered over 2 weeks. One month later the child started to develop similar lesions over face and upper trunk that cleared again upon resuming the antibiotic. The child was investigated for glucose-6-phosphate dehydrogenase serum level (which came to normal) prior to dapsone administration. Dapsone at a dose of 25 mg was started and the lesions started to improve and further cleared after 50 mg dose administration within 2 weeks. Hemoglobin dropped 1.2 g (from 13.8 to 12.6 g/dl) in 1 week but it stabilized thereafter.

3. Discussion

Childhood PF is commonly misdiagnosed as bacterial or fungal infection, seborrheic dermatitis, atopic dermatitis or less commonly psoriasis. Thus, it is not uncommon to have lag time of several months before correct diagnosis can be made. It seems that there is no relation between the prognosis and such delay in the diagnosis. In histopathology, majority of previous cases in the literature were consistent with a diagnosis of PF with the presence of upper epidermal acantholysis and neutrophilic and/or eosinophilic infiltrate. DIF usually confirms the diagnosis with IgG and C3 deposition at upper intracellular epidermis. Although rarely available, enzyme-linked immunosorbent assay (ELISA) for the desmoglein 1 autoantibody can successfully confirm the diagnosis.

In this case, we have shown that flucloxacillin was able to completely control the skin eruption during its administration which is similar to all the existing treatments for PF. Until now, there is no curing therapy for PF till the spontaneous remission occurs. To have a new effective medicine in the dermatologists’ armamentarium, particularly for chronic illness, is very helpful when other treatments cannot be used. The proposed theory for flucloxacillin effect on such immunobullous disease is thought to be through its anti-inflammatory effects. It has been shown that flucloxacillin can control other immunobullous disease; linear IgA bullous dermatosis of childhood (Alajlan et al., 2006). There are not many examples in the literature of such anti-inflammatory effect of flucloxacillin; however, other antibiotics such tetracycline group have been shown to have an anti-inflammatory property both clinically.
and in the laboratory (Aho and Mannisto, 1988; Thornfeldt and Menkes, 1987; Amato et al., 2002). Flucloxacillin is relatively safe for short courses as it is evident by its wide use in bacterial infections both in children and adults (Janknegt, 1997). However, a major adverse effect of flucloxacillin is cholestatic hepatitis that might rarely be fatal (Derby et al., 1993; Devereaux et al., 1995; Koek et al., 1994). Other very rare but significant adverse effects of flucloxacillin include aplastic anemia, hemolytic anemia, agranulocytosis, acute interstitial nephritis and acute renal failure (Xu and Murray, 2008; Tuffs and Manoharan, 1986; Burton et al., 1995; Bakker et al., 1995; Dobson et al., 2005).

In PF, topical and systemic corticosteroids were used in majority of the cases (Kahn and Lewis, 1971; Qureshi et al., 1997). Dapsone, erythromycin (Perry, 1961), sulfapyridine (Goodyear et al., 1991), chloroquine (Petratos and Andrade, 1967), corticotropin (ACTH) (Siregar et al., 1971) as well as immunosuppressive therapy like azathioprine (Larregue et al., 1980) and methotrexate (Wananukul and Pongprasit, 1999) were also used. Relative effectiveness among the various agents used to treat PF was difficult to assess because of multiple factors: frequent use of overlapping agents, differences in disease severity, and lack of clear or complete information regarding time to disease resolution, total duration of therapy and clinical outcome. The majority of those children reviewed in the literatures were clear of disease, either off medication or on low maintenance doses of medication, within 1 year, regardless of the type of therapeutic intervention. However, the longest follow up period available from the literature was only 4 years (Ahmed and Salm, 1983). One death attributed to the skin disease was reported (Perry, 1961).

In conclusion, the response of PF to flucloxacillin was dramatic in 2 weeks and consistent over the 10 weeks period and upon resuming the treatment in the second round. From our experience, flucloxacillin should be tried for further use in immunobullous diseases.

References


