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Case report

Subcorneal pustular dermatosis in a 7-year old Saudi child: A case report and review of the literature☆

Ali Al Ameer *, Abdullah Al Salman, Ibraheem Al Braheem, Yosif Al Marzoq, Mariam Imran

Dermatology Department in King Fahad Hofuf Hospital, Saudi Arabia

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Abstract

Subcorneal pustular dermatosis (SCPD) also known as Sneddon–Wilkinson disease (Sneddon and Wilkinson, 1956) is a rare, benign, chronic, sterile pustular eruption which usually develops in middle-age or elderly women; it is rarely seen in childhood and adolescence (Johnson and Cripps, 1974). The primary lesions are pea-sized pustules classically described as half-pustular, half-clear flaccid blisters. Histologically the most important feature is a subcorneal accumulation of neutrophils with the absence of spongiosis or acantholysis. In this paper we present the case of a 7-years-old boy diagnosed with SCPD based on the characteristic clinical and histological features. Oral and topical corticosteroid has been successfully used in the treatment of the disease.

Keywords: Subcorneal pustular dermatosis (Sneddon–Wilkinson disease); Histopathology; Immunofluorenscence; Immunoglobulin A; Dapsone; Prednisolone; Clobetasone proprionate

1. Case report

A 7-year-old boy was admitted to our clinic with a recurrent itchy pustular eruption located on the trunk on and off in the last six months (Figs. 1 and 2). The palms, soles, and mucous membrane were spared, and no lymphadenopathy or hepato-splenomegaly was present. There were no abnormalities of the nails and mucous membranes.

A complete blood count and the studies of serum biochemistry showed normal results; moreover serum protein electrophoresis had negative results. Normal Thyroid hormone profile reads Rheumatoid factor as negative.

The dermatologic examination revealed multiple-grouped flaccid pustules varying in size and some of them tended to coalesce to form annular pattern and superficial crusts on the normal or mildly erythematous skin of trunk and upper extremities. Healed lesions presented as residual hyperpigmentation and new lesions in the periphery.

Histopathology demonstrated a subcorneal vesiculobullous dermatitis (Fig. 3); the pustule is located immediately below the stratum corneum and contains mainly neutrophils with few eosinophils. The underlying epidermis to the pustule shows slight intercellular edema. In the dermis, superficial blood vessels are surrounded by a nonspecific mixed inflammatory cell infiltrate consisting of neutrophils and mononuclear cells. Direct immunofluorescence studies

* Corresponding author. Tel.: +966 505922063.
E-mail address: alimalameer@hotmail.com (A. Al Ameer).

☆ We describe a rare case of Subcorneal pustular dermatosis (SNEDDON–WILKINSON DISEASE) in a 7-year old Saudi boy presented to the outpatient clinic in King Fahad Hofuf Hospital. We highlight the clinical features and histopathological findings that distinguish this case from other pustular diseases. We also describe the management and the outcome of this case.

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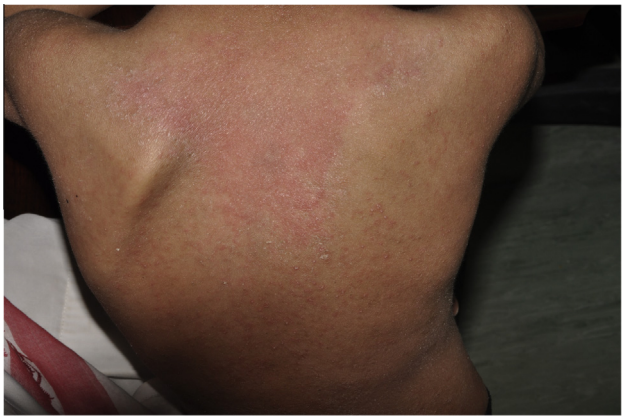

Figure 1. A recurrent itchy pustular eruption located on the trunk.

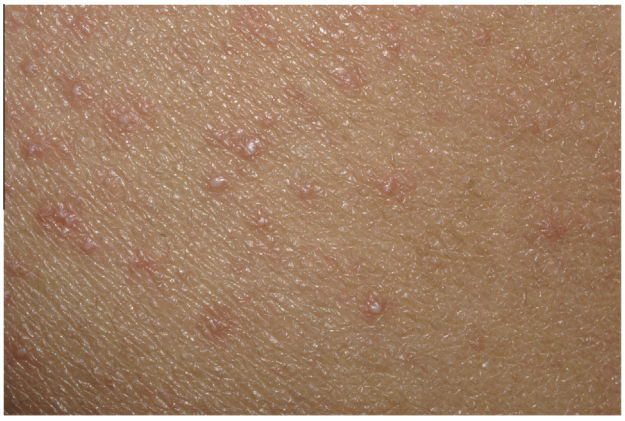

Figure 2. Closer view.

are negative for immunoglobulin A (IgA) intercellular accumulation. On the basis of this finding, associated to histopathological features and the clinical date, a diagnosis of subcorneal pustular dermatitis (SCPD, Sneddon–Wilkinson disease) was made.

With treatment in the form of tapering dose of prednisolone starting with 30 mg over three weeks' time and then topical clobetasone proprionate the patient showed great improvement within eight weeks. The patient was lost to follow-up but presented after 6 months upon relapse. The same course of treatment was repeated with significant improvement within two months. No further follow up of patient could be accomplished.

2. Discussion

Subcorneal pustular dermatosis is a chronic, relapsing, pustular eruption, generally involving the trunk, which affects mainly women over 40 years of age according to Sneddon and Wilkinson's original report (Sneddon and Wilkinson, 1956).

Children can have various bullous and pustular skin diseases like psoriasis, pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid as well as dermatitis herpetiformis; all of these were once thought to be unique to people in the fourth-fifth decade of life. Subcorneal pustular dermatosis appears to be another one of these diseases (Scalvenzi and Palmisano, 2013).

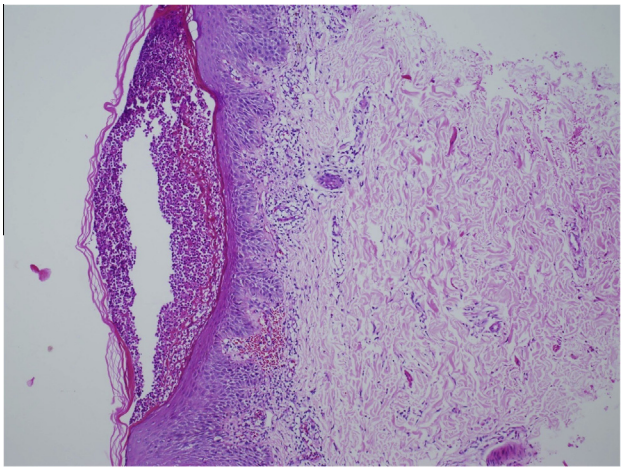

Figure 3. Histopathology demonstrated a subcorneal vesiculo-bullous dermatitis (Fig. 3); the pustule is located immediately below the stratum corneum and contains mainly neutrophils with few eosinophils.

Only 15 cases of pediatric SCPD are described in the literature (Johnson and Cripps, 1974; Yayli et al., 2006).

Even if SCPD is an uncommon condition in childhood, it must be considered as a possible cause of sterile pustular eruptions in a child. An accurate physical examination, a complete blood count, and studies of serum biochemistry are strongly recommended to exclude a pathology in association.

The etiopathogenesis of SCPD is not well known. Culture of the pustules is sterile. A relationship with Pyoderma gangrenosum (Scerri et al., 1994; Marsden and Millard, 1986), benign monoclonal IgA gammopathy (Kasha and Epinette, 1988; Scerri et al., 1994), IgA myeloma (Atukorala et al., 1993; Takata et al., 1994; Vaccaro et al., 1999), SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis) syndrome (Scarpa et al., 1997), Crohn's disease (Delaport et al., 1992), Rheumatoid arthritis (Butt and Burge, 1995), and Hyperthyroidism (Taniguchi et al., 1995) has been documented.

In our case, the history, physical examination, and laboratory results did not reveal any systemic associations. Moreover some cases, which were consistent with SCPD according to the clinical and histologic features, have been reported with the presence of an intercellular IgA deposition within the epidermis (Hashimoto et al., 1987).

This disease involves more frequently the trunk as in this case. Other sites can be involved like the intertriginous areas, and flexor aspects of the limbs; more rarely the face is implicated. Pustules on palms and soles have also been reported (Takematsu and Tagami, 1993), while mucous membranes are almost never affected.

The differential diagnosis of SCPD includes impetigo, pustular psoriasis, dermatophyte infection, immunobullous