Subcorneal pustular dermatosis as a cause of pityriasis amiantacea in a young child

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INTRODUCTION

Subcorneal pustular dermatosis (SPD), also known as Sneddon–Wilkinson disease, is a rare neutrophilic dermatosis characterized by recurrent sterile pustules.[1] SPD usually occurs in middle-aged or elderly women; however, cases of children with atypical features have also been reported in the literature.[2,3] Herein, we report the case of a 6-year-old boy diagnosed with SPD, which caused pityriasis amiantacea (PA).

Abbreviations used:

GPP: generalized pustular psoriasis
IL: interleukin
PA: pityriasis amiantacea
SPD: subcorneal pustular dermatosis

CASE REPORT

A 6-year-old boy presented with 2-month-old itchy pustular lesions on the trunk and extremities. One month before he presented to our hospital, he was initially diagnosed with generalized pustular psoriasis (GPP) on the basis of clinical and histopathologic examination. He had been treated with oral steroid and topical calcipotriol, but it had been ineffective. Fever and other constitutional symptoms were not present. He had no other medical disease and no family history of any related diseases.

Physical examination revealed multiple scaly pustular eruptions forming annular or serpiginous patterns on the face, neck, trunk, and extremities. The palms, soles, and mucous membranes were spared (Fig 1).

A complete blood cell count and the studies of serum biochemistry showed normal results. Serum protein electrophoresis revealed normal findings.

Outside pathology slides were reviewed and demonstrated subcorneal pustules composed of predominantly neutrophils with occasional eosinophils. They also revealed hyperkeratosis and parakeratosis with irregular acanthosis, papillary dermal edema, and superficial perivascular infiltration of neutrophils (Fig 2).

Based on the clinical and histopathologic findings, a diagnosis of SPD was made. He was given dapsone 25 mg/d orally and achieved significant improvement within 2 weeks.

After 18 months of complete remission, pustular lesions recurred shortly after the consumption of a red ginseng health supplement. He denied any constitutional symptoms, and all laboratory findings were normal. One week of treatment with topical calcipotriol/betamethasone was ineffective and his pustular eruptions worsened. Moreover, he presented with sticky, silvery scales adhering to the scalp and hair shafts (Fig 3). We diagnosed the characteristic clinical feature as PA caused by SPD. Dapsone 25 mg/d was reinstituted, and eruptions almost resolved after 4 weeks. No side effects were observed during the 19-month follow-up period.

DISCUSSION

SPD is an uncommon chronic, relapsing, pustular eruption that was first described by Sneddon and Wilkinson.[1] SPD is more common in middle-aged and elderly women.[2] Few cases have been reported in children, without differences described in clinical

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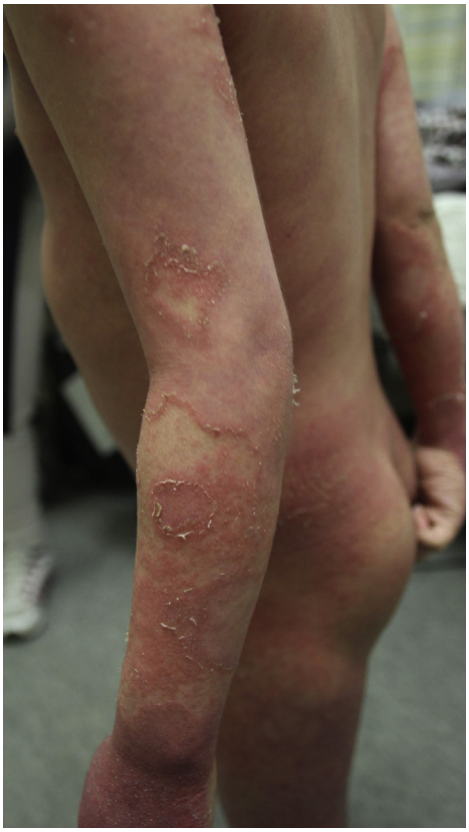

Fig 1. Multiple scaly eruptions forming annular or serpiginous patterns distributed throughout the trunk and extremities.

features and prognosis between children and adults, but some cases tend to have atypical features (Table I).

The differential diagnosis of SPD includes other disorders that may present with widespread pustules or annular, circinate, or serpiginous plaques. Examples include pustular psoriasis, acute generalized exanthematous pustulosis, impetigo, dermatophytosis, eosinophilic pustular folliculitis, dermatitis herpetiformis, necrolytic migratory erythema, pemphigus foliaceus, and IgA pemphigus.[3]

GPP is probably the most difficult entity to distinguish from SPD due to their clinical and histologic resemblances. Our patient was also initially mistaken to be suffering from GPP. A careful clinical and histopathologic examination is necessary to accurately diagnose SPD and provide effective treatment.

Clinically, pustules in SPD manifest as half pustular and half vesicular blisters, as opposed to purely pinpoint pustules commonly seen in GPP. Patients with GPP usually have systemic symptoms, including fever and malaise, and GPP should be suspected in a patient with a patient or family history of psoriasis or nail or joint findings of psoriasis. SPD has a benign clinical course. In contrast, GPP may develop life-threatening complications without supportive treatment.

Histologically, SPD shows little or no spongiosis, and rarely acantholysis. GPP, on the other hand, reveals the classic psoriasiform changes, parakeratosis, elongation of rete ridges, spongiform pustules of Kogoj, and acantholysis.[4]

The treatment of choice in children is dapsone, similar to what is indicated for adults. However, cases of hematologic toxicity from the drug have been reported in children, mainly hemolytic anemia

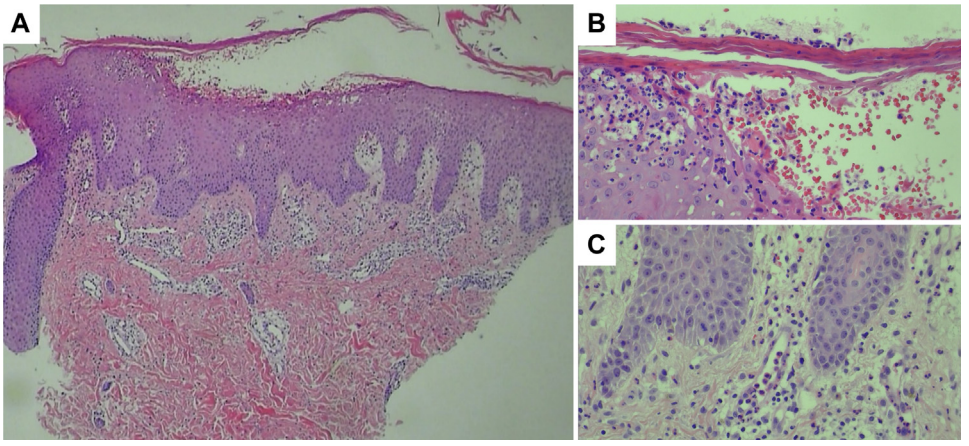

Fig 2. Histopathologic image of punch biopsy specimen obtained from the abdomen at the previous hospital. **A**, Hyperkeratosis and parakeratosis with irregular acanthosis, papillary dermal edema, and perivascular inflammatory infiltrates in the papillary dermis. **B**, Subcorneal pustule composed of predominantly neutrophils with occasional eosinophils. **C**, Neutrophilic infiltration in papillary dermis. (**A**, **B**, and **C**, Hematoxylin-eosin stain; original magnifications: **A**, ×40; **B**, ×400; **C**, ×400.)

Fig 3. Accumulation of thick scale that adheres tightly to the scalp and hair.

and methemoglobinemia. Thus, a comprehensive clinical and laboratory surveillance is needed to detect the earliest signs of its toxicity. Other therapeutic options for children are topical and systemic corticosteroids and acitretin.[2,3] In our case, dapsone showed great response and was well tolerated.

SPD mostly affects the skin; however, it can be associated with a variety of cutaneous and systemic diseases, including multiple myeloma, IgA monoclonal gammopathy, and inflammatory diseases, such as rheumatoid arthritis and Crohn's disease.[2,3] In our case, the history, physical examination, and laboratory results did not reveal any systemic associations.

This case also suggests that SPD can cause PA. PA is a rare scalp condition characterized by the accumulation of thick scale that tightly adheres to the scalp and hair. The etiology is uncertain, but it has been proposed that the disorder may be a

Table I. Summary of pediatric cases of subcorneal pustular dermatosis that have been published in the English-language medical literature

Literature (y)	Age/sex	Presentation	Histopathologic findings	Treatment
Johnson et al (1974)	3 y/female	Pustules forming circinate pattern on face and in the groin, upper portions of thighs, and both axillae	Subcorneal pustules containing mostly neutrophils with a few eosinophils. The dermis showed dilatation of the small blood vessels, surrounded by neutrophils migrating toward the epidermis. There was absence of spongiform pustules, microabscesses, congestion of papillae, or elongation of the rete ridges.	Dapsone 50 mg/d: no response Systemic and topical steroid: good response
	3 y/male	Pustular eruption around neck and chest		Systemic and topical steroid + Dapsone: good response
Koçak et al (2003)	13 y/female	Grouped papules and serpiginous pustules in the axillae and inguinal and gluteal regions	Subcorneal pustule containing neutrophils, dermal edema, and infiltrate composed of mononuclear cells. DIF, IIF: negative for IgA.	Dapsone 50 mg/d: good response within 3 wk Dapsone was stopped due to hemoglobin decrease (12.5 → 9.6) Topical steroid was initiated
Yayli et al (2006)	10 y/female	Pruritic skin eruption located on the trunk and extremities	Subcorneal pustules containing neutrophils accompanied by mixed perivascular infiltration composed of mostly neutrophils in dermis. DIF: negative for Ig or complement within the epidermis.	Acitretin 10 mg/d: good response within 4 wk

Continued

Table I. Cont'd

Literature (y)	Age/sex	Presentation	Histopathologic findings	Treatment
Massimiliano et al (2013)	7 y/male	Multiple grouped flaccid pustular eruption located on the trunk, limbs, and the face	Subcorneal pustules containing mostly neutrophils with a 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. DIF: negative for IgA intercellular accumulation.	Dapsone 30 mg/d: good response within 2 wk
Kundak et al (2017)	5 y/female	Erosions and sterile pustules formed an annular structure on the neck, upper portion of the back, and upper portion of the body	Subcorneal pustules containing mainly polymorphonuclear leukocytes with a few eosinophils, acantholytic cells in the cavity, and spongiosis in the epidermis. IIF, DIF: negative for IgG, IgM, IgA, and C3.	Intravenous immunoglobulin 600 mg/kg + Dapsone 1 mg/kg: good response within 1 wk
Jardim et al (2018)	15 y/female	Pustular lesions on the proximal region of the upper limbs, chest, and abdomen	Subcorneal spongiform pustule. DIF: negative for IgA, IgM, IgG, and fibrinogen.	Dapsone 100 mg/d: good response within 1 mo
Saini et al (2020)	16 y/male	Multiple grouped flaccid pustular lesions forming annular, circinate, and serpiginous pattern over trunk, infra-axillary area, and buttocks	Subcorneal accumulation of neutrophils without spongiosis and acantholysis with nonspecific mixed inflammatory cells in dermis. DIF: negative for IgA.	Dapsone 100 mg/d: good response within 4 wk
Present case	6 y/male	Multiple pustular eruptions forming annular and serpiginous patterns on the trunk and extremities SPD relapsed after red ginseng intake, and it also caused pityriasis amiantacea	Subcorneal pustule composed of predominantly neutrophils with occasional eosinophils. Hyperkeratosis and parakeratosis with irregular acanthosis. Papillary dermal edema and perivascular inflammatory infiltrates in the papillary dermis.	Dapsone 25 mg/d: good response within 2 wk

DIF, Direct immunofluorescence; *Ig*, immunoglobulin; *IIF*, indirect immunofluorescence; *SPD*, subcorneal pustular dermatosis.

particular reaction pattern to various inflammatory scalp diseases, such as psoriasis, seborrheic dermatitis, tinea capitis, and atopic dermatitis.[5,6] To the best of our knowledge, PA caused by SPD has not been reported in the previous literature. We should keep in mind PA as a rare complication of SPD.

Finally, this case implies the association between ingesting immunostimulatory herbs, like red ginseng, and flares of inflammatory skin diseases, including SPD. There have been few prior reports of immunostimulatory herbs worsening some skin diseases characterized by an exaggerated immune response, such as lupus erythematosus, dermatomyositis, and autoimmune blistering disorders.[7]

Ginseng is an herbal supplement derived from the root of the *Panax* genus that has been used in Eastern Asia for >2000 years. It is traditionally used as an immunostimulant. Several in vitro and in vivo studies have shown that ginseng activates immune cells and increases cytokines and chemokines, such as interleukin (IL)-1β, IL-6, IL-8, IL-10, and tumor necrosis factor-α. These cytokines are neutrophil chemoattractants that have been found at increased levels in scale extracts of patients with SPD.[2,3,8-10] Given the immunomodulatory properties of red ginseng and pathophysiology of SPD, it is plausible that red ginseng potentiated a susceptible patient's immune response, culminating in the aggravation of SPD. As patients' independent use of herbal therapies continues to grow, dermatologists should screen for functional food use that could aggravate inflammatory skin diseases.

Conflicts of interest

None disclosed.

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