Subcorneal Pustular Dermatosis in Paediatrics: A Case Report and Review of the Literature

Mais A. Alhafi 1, Mohamed I. Janahi 2, Zainab N. Almossalli 1

1. Dermatology, Salmaniya Medical Complex, Manama, BHR 2. Orthopedic Surgery, Salmaniya Medical Complex, Manama, BHR

Corresponding author: Mohamed I. Janahi, mimjanahi@gmail.com

Abstract
Subcorneal pustular dermatosis (SPD) is a rare chronic condition rarely seen in adolescence and childhood. The exact etiology of the disease remains unknown. In this paper, we report the case of a 14-year-old girl who came with a history of itchy skin lesions confined to the upper and lower extremities, thighs and pubic area for two months. Physical examination showed well-demarcated annular brownish plaques, ranging in size from 5cm to 7cm, in addition to a scaly and elevated border with few pustules noted over the upper and lower extremities, thighs and pubic area. Some lesions also showed central clearing. New annular vesicular lesions were also noted on the lower extremity and inner thigh. She was diagnosed with SPD based on the characteristic clinical and histological features. The patient was treated with Dapsone and showed good clinical response.

Introduction
Subcorneal pustular dermatosis (SPD) or Sneddon-Wilkinson disease, is a rare, chronic yet benign inflammatory neutrophilic dermatosis (ND), which was first described by Lan Sneddon and Darell Wilkinson in 1956 [1]. Its specific etiology and pathophysiology remain largely unknown. The condition is four times more common in women than in men [2,3]. Although it typically presents in middle-aged women, there have been a few cases reported in children [4,5]. SPD has been associated with a wide spectrum of systemic disorders, including other neutrophilic dermatosis, hematologic disorders, connective tissue diseases, and neoplasms [6]. To the authors’ knowledge, a case of SPD without any medical background has been reported only once in the literature [7]. We report a case of a child diagnosed with SPD without any systemic illness.

Case Presentation
A 14-year-old female presented with a two-month history of itchy skin lesions confined to the upper and lower extremities, thighs and pubic area. Her system review was negative for any other complaints or associated symptoms.

Physical examination revealed well-demarcated annular brownish plaques, ranging in size from 5cm to 7cm. Borders were scaly and elevated with few pustules noted over the upper and lower extremities, thighs and pubic area (Figure 1). In addition, some lesions showed central clearing. There was no hair, nail, mucosal or genital lesions.
In view of the previous findings, the patient diagnosis was Tinea corporis and she was started on terbinafine 250 mg tab plus topical miconazole cream twice daily for two weeks. She came back after two weeks with more lesions noted on the lower extremity and inner thigh despite her compliance with the treatment given.

A skin biopsy was taken from the left and right lower limbs. Histopathology demonstrated pustules located immediately below the stratum corneum and contained mainly polymorphonuclear leucocytes with a few eosinophils, acantholytic cells in the cavity, and spongiosis in the epidermis (Figure 2).

Periodic acid-Schiff (PAS) stain was negative for fungal infection. A complete blood count, erythrocyte sedimentation rate (ESR) activity, G6PD activity and liver function test showed normal results.

In view of the histopathological and clinical findings, the diagnosis was subcorneal pustular dermatosis (Sneddon-Wilkinson disease). The patient was started on dapsone 50mg once daily for two weeks then the dose was increased to 100mg once daily with good response.

**Discussion**

Sneddon-Wilkinson or subcorneal pustular dermatosis (SPD) is a rare skin disorder, which typically presents in middle-aged women and is rarely seen in children [8]. Sarkany reported the first case of pediatric SPD in a
Subcorneal pustular dermatosis has no cure. However, treatment is available to palliate it and it centers on the anti-neutrophilic sulfone, dapsone. Dapsone is the treatment of choice and in our case, it showed a good response [20]. Other treatments include acitretin, psoralen + ultraviolet light A (PUVA), narrowband ultraviolet B (UVB) [21], broadband UVB [22], colchicine [23], etretinate [24], cyclosporine, prednisone [25], and infliximab [26]. Potent topical corticosteroids may also be used as a treatment alone [27] or in combination with dapsone. Sulfapyridine and sulfamethoxypyridazine can also be used. However, they are not as effective [23]. To our knowledge, this is the first case report about pediatric subcorneal pustular dermatosis done in Bahrain.

**Conclusions**

Although Sneddon-Wilkinson disease is extremely rare in children, it should be considered while investigating for the differentials. Proper investigations are crucial and should be done at first presentation in order to prevent poor outcome. Referral to other specialties must be initiated if any underlying rheumatologic or hematologic conditions have been identified. No curable treatment has been discovered yet and the management is purely palliative. Dapsone remains the first line of management and it should be started as soon as the diagnosis is made.

**Additional Information**

**Disclosures**

**Human subjects:** Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.
References

1. Sneddon IB, Wilkinson DS: Subcorneal pustular dermatosis. Br J Dermatol. 1956, 68:385-394. 10.1111/j.1365-2133.1956.tb12774.x
2. Scalvenzi M, Palminsano F, Annunziata MC, Mezza E, Cozzolino I, Costa C: Subcorneal pustular dermatosis in childhood: a case report and review of the literature. Case Rep Dermatol Med. 2013, 2013:424797. 10.1155/2013/424797
3. Naik HB, Cowen EW: Autoinflammatory pustular neutrophilic diseases. Dermatol Clin. 2013, 31:405-425. 10.1016/j.det.2013.04.001
4. Johnson SAM, Cripps DJ: Subcorneal pustular dermatosis in children. Arch Dermatol. 1974, 109:73-77. 10.1001/archderm.1974.01650100140012
5. Yalici S, Bahadır S, Alpay K, Cimjrit G, Reis A: A case of juvenile subcorneal pustular dermatosis successfully treated with acitretin. Int J Dermatol. 2006, 45:1131-1133. 10.1111/j.1365-4632.2006.02984.x
6. Watts PJ, Khachemoun A: Subcorneal pustular dermatosis: a review of 30 years of progress. Am J Clin Dermatol. 2016, 17:663-671. 10.1007/s40257-016-0202-8
7. Yamaguchi Y, Oyama N, Koizumi H, Chino T, Hasegawa M: Successful treatment of recalcitrant subcorneal pustular dermatosis with oral nicorinamide. J Dermatol. 2019, 46:438-440. 10.1111/1346-8138.14980
8. Kashaa EE Jr, Epinette WW: Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) in association with a monoclonal IgA gammopathy: a report and review of the literature. J Am Acad Dermatol. 1988, 19:854-858. 10.1016/S0190-9622(88)70244-5
9. Sarkany I: Subcorneal pustular dermatosis. Br J Dermatol. 1958, 70:507.
10. Bordignon M, Zattra E, Montesco MC, Alabac M: Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) with absence of desmoglein 1 and 3 antibodies: case report and literature review. Am J Dermatopathol. 2008, 9:51-55. 10.2165/00128071-200809010-00006
11. Tajiri K, Nakajima T, Kawai K, Minemura M, Sugiyma T: Sneddon-Wilkinson disease induced by soroferin in a patient with advanced hepatocellular carcinoma. Intern Med. 2015, 54:597-600. 10.2169/internalmedicine.54.3675
12. Scerri L, Zaki I, Allen BR: Pyoderma gangrenosum and subcorneal pustular dermatosis, without monoclonal gammopathy. Br J Dermatol. 1994, 130:398-399. 10.1111/j.1365-2133.1994.tb02941.x
13. Scarpà R, Lubrano E, Cozzi R, Ames PR, Oriente CB, Oriente P: Subcorneal pustular dermatosis (Sneddon-Wilkinson syndrome): another cutaneous manifestation of SAPHO syndrome?. Br J Rheumatol. 1997, 36:601-605. 10.1093/rheumatology/36.5.602
14. Delaporte E, Colombel JF, Nguyen-Maifer C, Piette F, Cortot A, Bergoend H: Subcorneal pustular dermatosis in a patient with Crohn’s disease. Acta Derm Venereol. 1992, 72:301-302.
15. Tsuruta D, Matsumura-Oura A, Ishii M: Subcorneal pustular dermatosis and Sjögren’s syndrome. Int J Dermatol. 2005, 44:955-957. 10.1111/j.1365-4632.2004.02990.x
16. Butt A, Marge SM: Sneddon-Wilkinson disease in association with rheumatoid arthritis. Br J Dermatol. 1995, 132:513-515. 10.1111/j.1365-2133.1995.tb05037.x
17. Taniguchi S, Tsuruta D, Kutsuna H, Hamada T: Subcorneal pustular dermatosis in a patient with hyperthyroidism. Dermatology. 1995, 190:64-66. 10.1159/000246638
18. Reed J, Wilkinson J: Subcorneal pustular dermatosis. Clin Dermatol. 2000, 18:301-315. 10.1111/j.1070-4790.1998.tb00121-2
19. Hashimoto T, Inamoto N, Nakamura K, Nishikawa T: Subcorneal pustular dermatosis (Sneddon-Wilkinson disease induced by sorafenib in a patient with advanced hepatocellular carcinoma). Br J Rheumatol. 1997, 36:601-605. 10.1093/rheumatology/36.5.602
20. Patel R, Cafardi JM, Patel N, Sami N, Cafardi JA: Tumor necrosis factor biologics beyond psoriasis in dermatology. Expert Opin Biol Ther. 2011, 11:1341-1359. 10.1517/14712598.2011.590798
21. Orton Di, George SA: Subcorneal pustular dermatosis responsive to narrowband (TL-01) UVB phototherapy. Br J Dermatol. 1997, 137:149-150. 10.1046/j.1365-2133.1997.tb01937.x
22. Park YK, Park HY, Bang DS, Cho CK: Subcorneal pustular dermatosis treated with phototherapy. Br J Dermatol. 1986, 25:124-126. 10.1111/j.1365-4632.1986.tb04556.x
23. Pavithran K: Colchicine in the treatment of subcorneal pustular dermatosis. Indian J Dermatol Venereol Leprol. 1995, 61:56-57.
24. Iandoli R, Montefrecola G: Treatment of subcorneal pustulosis by etretinate. Dermatologica. 1987, 175:253-258. 10.1159/000248910
25. Zachariae COC, Rassens K, Weismann K: An unusual severe case of subcorneal pustular dermatosis treated with cyclosporine and prednisolone. Acta Derm Venereol. 2000, 80:386-387.
26. Bonifati C, Trento E, Cordiali Fei P, Muscardin L, Amantea A, Carducci M: Early but not lasting improvement of recalcitrant subcorneal pustular dermatosis (Sneddon-Wilkinson disease) after infliximab therapy: relationships with variations in cytokine levels in suction blister fluids. Clin Exp Dermatol. 2005, 30:662-665. 10.1111/j.1365-2230.2005.01902.x
27. Walkden VM, Roberts A, Wilkinson JD: Two cases of subcorneal pustular dermatosis. Response to use of intermittent clobetasol propionate cream. EJD Eur J Dermatol. 1994, 4:44-46.