Subcorneal Pustular Dermatosis Masquerading as Pustular Psoriasis- A Rare Entity in Childhood

B. C. Sharath Kumar[1], T. Chaitra[2]

[1]Professor and HOD, Department of Dermatology, Venerology and Leprosy, Kempegowda Institute of Medical Sciences Hospital and Research Centre, Bengaluru, Karnataka, India.
[2]Resident, Department of Dermatology, Venerology and Leprosy, Kempegowda Institute of Medical Sciences Hospital and Research Centre, Bengaluru, Karnataka, India.

INTRODUCTION

Subcorneal pustular dermatosis (SCPD) also known as neutrophilic dermatosis is characterized by superficial pustular lesions.[1] It was first described by Ian Sneddon and Darrell Wilkinson in 1956.[1,2] The primary lesions are pea-sized pustules classically described as half clear, half pustular, flaccid blisters. The exact cause of SCPD is not yet known. It is a rare, chronic, relapsing pustular disorder with a predilection for flexural aspects of the limbs, trunk, and intertriginous areas. Usually it spares the face, palms, soles and mucous membrane. This condition is frequently seen in adults, especially in women, in the age group 40-50 years.[3] In rare cases, unusual involvement of the face, palms, and soles has been described.[4] Dapsone is the treatment of choice for SCPD. Others includes retinoids, colchicine, phototherapy with psoralen, ultraviolet A (PUVA), broad or narrow band UVB, corticosteroids and cyclosporine. The anecdotal use of tetracycline, minocycline, vitamin E, tacalcitol, infliximab, ketoconazole, mizoribine, mebhydrolin and mycophenolate mofetil.[4] However this disease can occur rarely during the childhood. Only few pediatric SCPD are described in literature.[5,6] We report a case of SCPD in a 3-year-old child which mimicked pustular psoriasis and was treated with topical dapsone showing good results.

PRESENTATION OF CASE

A 3-year-old pre-school male child presented to skin OPD with complaints of multiple pus-filled lesions over reddish raised areas all over the body since 6 months, aggravated since 1 week and it was associated with itching. The lesions initially developed over the upper extremities and trunk later, progressed to involve the whole-body surface. The palms, soles and the mucous membrane were spared. History of photosensitivity was present and there was no history of atopy. Mother gave history of similar complaints since one and a half year of age which was present on and off. No significant family history was noted. On general examination, lymphadenopathy was present and geographic tongue was seen.

On dermatological examination, multiple flaccid pustules varying in size from 2 to 7 mm over an erythematous base were seen. Few coalescing to form annular and serpiginous pattern. Superficial crusting were present over trunk, extremities, face and scalp. Scaling were present at some areas. Multiple post inflammatory hyperpigmentation was present on healed areas. Nails were normal. Routine investigations were done. Hemoglobin was 6.9 g%, TLC –1, 5, 550, differential count showed neutrophilia and urine routine showed albumin in traces.

Corresponding Author:
Dr. T. Chaitra,
Resident, Department of Dermatology, Venerololgy and Leprosy,
Kempegowda Institute of Medical Sciences Hospital and Research Centre, Bengaluru, Karnataka, India.
E-mail:
chaitra.friendz@gmail.com

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Pus culture didn't yield any organism. Differential diagnosis of pustular psoriasis, Subcorneal pustular dermatosis (SCPD), Langerhans cell histiocytosis, atopic dermatitis was made. Biopsy was done. Histopathology showed hyperkeratosis with pustule comprising of neutrophils, few eosinophils, and few acantholytic cells. All features were suggestive of SCPD. Patient was then started on oral antihistamines with topical dapsone gel twice a day. There was 40% improvement after 1 month. After 1 month, patient was advised to use the dapsone gel for another 2 months. After 3 months, the patient is still monitored for the follow-up at our hospital every 4 weeks.

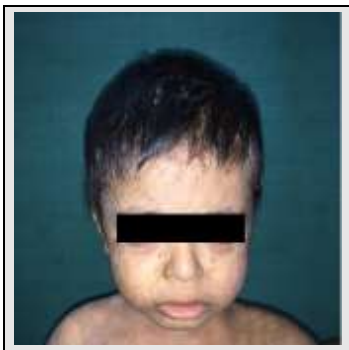

Figure 1. Multiple Flaccid Pustules and Superficial Crusts on the Normal Skin or Erythematous Skin Seen Over the Face

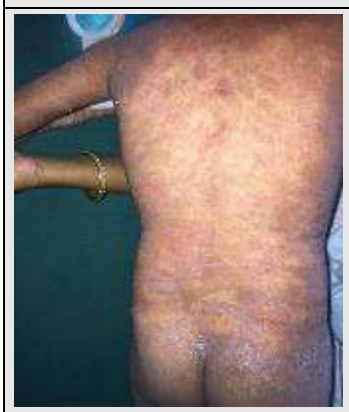

Figure 2. Multiple Grouped Flaccid Pustules Coalescing to Form Circinate or Serpiginous Pattern and Superficial Crusts on the Normal Skin or Erythematous Skin Over the Trunk

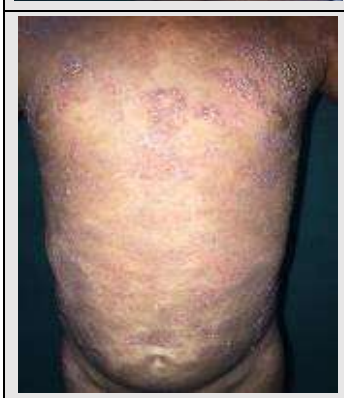

Figure 3. Multiple Flaccid Grouped Pustules Forming Annular Pattern Over the Anterior Aspect of the Trunk with Superifical Crusting Over Normal or Erythematous Skin Seen

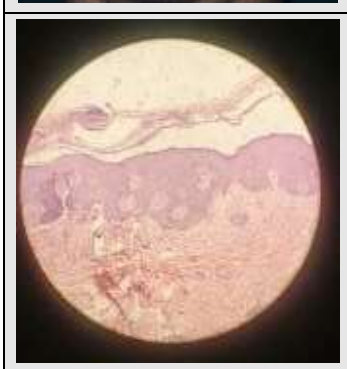

Figure 4. Histopatholgical Examination Shows Subcorneal Pustule in Stratum Corneum Containing Neutrophils, and the Underlying Epidermis Shows Intercellular Edema

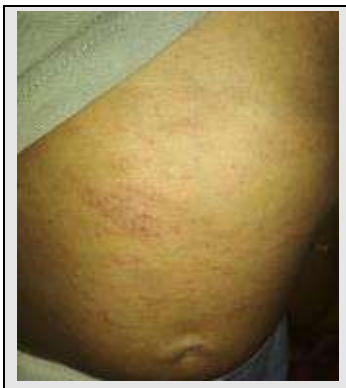

Figure 5. Followup after 4 Weeks. The Patient was Treated with Dapsone. Cutaneous Lesions over the Trunk were Almost Healed with Few Erythematous Papules

Figure 6. Follow-Up after 4 Weeks with Dapsone Treatment. There was Disapperance of the Flaccid Pustules and Crusting over the Face. There was 60% Improvement after 4 Weeks of Dapsone Treatment

CLINICAL DIAGNOSIS

Subcorneal pustular dermatosis.

DIFFERENTIAL DIAGNOSIS

- Pustular psoriasis.
- Subcorneal pustular dermatosis.
- Langerhans cell histiocytosis.
- Severe atopic dermatitis.

PATHOLOGICAL DISCUSSION

Subcorneal pustular dermatosis (SPD) is a rare, chronic, relapsing pustular eruption characterized by subcorneal sterile pustules that contain neutrophils on histopathology.[7] Histopathological features of SCPD includes subcorneal neutrophilic pustules, occasional eosinophils, and a perivascular neutrophilic infiltrate. Generalized pustular eruption in a child can be a diagnostic challenge. Various dermatoses which present with generalized pustular and bullous lesions include pustular psoriasis, subcorneal pustular dermatosis (SCPD), severe atopic dermatosis, staphylococcal scalded skin syndrome (SSSS), acute generalized exanthematous pustulosis (AGEP), pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid and also dermatitis herpetiformis.[8,9,10]

Some cases of subcorneal pustular dermatosis have been considered a variant of pustular psoriasis. The clinical and histological differentiation of subcorneal pustular dermatosis from pustular psoriasis can be difficult, although

spongiform changes on histology favours the latter. Furthermore, a significant number of cases initially diagnosed as subcorneal pustular dermatosis are later diagnosed as psoriasis.[11,12]

The exact pathophysiology of subcorneal pustular dermatosis (SPD) is unknown. The microscopy suggests the presence of chemoattractants such as TNF-alpha in the uppermost layers of epidermis.[13] Direct immunofluorescence should exclude IgA staining because of autoantibodies to desmocollin-1.[14] Cases of SCPD in association with pyoderma gangreosum,[15] benign monoclonal IgA/IgG gammopathy,[16,17] multiple myeloma,[18] lymphomas, connective tissue diseases[19] and inflammatory bowel disease (IBD)[20] are well documented.

In our case, the history, physical examination, and laboratory results did not reveal any systemic associations. Histopathologically, the pustule containing neutrophils and absence of spongiform pustule of Kogoj and regular elongation of rete ridges all suggested the features of SCPD. In literature, only 15 cases of paediatric SCPD have been documented.[3, 21-23] 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.

DISCUSSION OF MANAGEMENT

Dapsone is the first choice of therapy.[6,7,24] The response is slower when compared to dermatitis herpetiformis. The resolution time is about 4 weeks. The dosage needs to be adjusted according to the severity of the disease. Once the patient starts showing the response the dose needs to be tapered to the lower dose. Acitretin can be used as an alternate or additional drug for the treatment of subcorneal pustular dermatosis specially in people who do not respond or intolerant to dapsone. The dose needs to be tapered to the lowest dose once the disease is under control.[25]

Other alternatives that has been proposed includes broadband or narrowband (ultraviolet) UVB phototherapy, PUVA (psoralen and UVB),[26,27] corticosteroids and cyclosporine,[28,29] infliximab and colchicine.[24] Other anecdotal usage include etanercept, tacalcitol,[30] sulfapyridine, sulfamethoxypyridazine, maxacalcitol,[31] ketoconazole,[32] azithromycin,[33] tetracycline, minocycline, vitamin E,[34] mizoribine,[35] mebhydrolin, adalimumab and mycophenolate mofetil.

FINAL DIAGNOSIS

Subcorneal pustular dermatosis (SCPD).

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