SUBCORNEAL PUSTULAR DERMATOSIS
(A case report and review of Literature)

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Summary

A 50 year old male patient with subcorneal pustular dermatosis is reported. Patient has been succesfully treated with dapsone 100 mg twice a day followed by 50 mg daily as a maintenance dose. The relevant literature is reviewed.

In 1956, Sneddon and Wilkinson[1] described 7 cases of a hitherto unknown dermatosis and coined the term 'subcorneal pustular dermatosis (SPD), for it.

SPD usually occurs in women and the average age of onset is between 40 and 50 years. The incidence ratio of women to men in the published cases has been about 4 : 1[2]. The cases which have been reported from India[3-8] have all been males ; most of them in the relatively younger age group. The youngest age at which the disease has been reported to occur is $3\frac{1}{2}$ months[9]. Johnson and Cripps[10] make mention a patient with SPD who gave birth to a child with similar skin lesions that lasted 7 days.

The primary lesion in subcorneal pustular dermatosis is a pustule. Occasionally a transient vesicular phase may be seen. It develops within a few hours and measures 7-10 mm. There is little or no surrounding erythema. The tops of the pustules readily rupture leaving behind a ragged thin edge which later gets covered by a thin crust. The pustules spread peripherally, leaving concentrically arranged annular and serpiginous lesions.

The eruption may be accompanied by mild to moderate itching or may cause only mild soreness. Individual lesion lasts for about 3-5 days, does not leave behind atrophy or scarring but results in some brown pigmentation. Fresh lesions however, continue to appear.

The chief sites of involvement are axillae, groins, abdomen and flexor aspects of upper and lower limbs. Face and mucous membranes are never affected. Rarely, palms and soles may be involved.

The condition usually lasts for 6-8 years with periods of remission which vary from several days to several weeks. There are no constitutional symptoms whatsover. The disease has to be clinically differentiated from impetigo, miliaria pustulosa, pemphigus foliaceus, dermatitis herpetiformis and pustular psoriasis.

Aetiopathogenesis

The exact cause of SPD is not known[11-15]. Immunofluorescent studies

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(direct and indirect) showed no fluorescence in the dermis, dermoepidermal junction or epidermis[10]. The pustule is formed within a few hours because of a rapid transepidermal migration of neutrophils that collect beneath the stratum corneum without stopping at the granular layer or disturbing the malpighian layer cells[16].

Electron microscopic studies show that the accumulation of polymorphs below the stratum corneum is only a secondary event, since degenerative changes in the granular cell layer have been demonstrated[17]. The subcorneal split is formed as a result of dissolution of the plasma membrane and of the cytoplasm of granular cells.

Immunoelectrophoresis study[18] showed an increase of $\propto$A globulins in sera from 2 patients with SPD suggesting a resemblance to DH. However, Krogh and Tondor[15] did not find any increase of $\propto$A globulins in the sera of 2 of their patients with SPD. They, however, showed the formation of immune complexes in vivo by immune adherence test and postulated that this ag/ab/ $c_{1,4,2,3}$ complex leads to the chemotactic trimolecular complex C 5, 6,7 of the complement system which then triggers off the selective transepidermal migration of polymorphonuclear leucocytes and thereby resulting in the formation of subcorneal blisters. However, further studies are needed.

Histopathology

The pustule is located directly below the stratum corneum and is filled predominantly by neutrophils. An occasional eosinophil may be present. The edidermis is normal except for mild spongiosis and intracellular edema of the cells of the malpighian layer. There may be dilatation of superficial dermal vessels with a surrounding infiltrate of polymorphonuclear leucocytes and occasional eosinophils. Rarely, a few acantholytic cells may be seen at the base of the pustule. These are believed to be due to secondary acantholysis caused by the action of the proteolytic enzymes present in the pustular contents[17].

Therapy of subcorneal pustular dermatosis

Out of the various forms of therapy employed for the treatment of SPD, dapsone remains the best[2]. In fact the response to dapsone is even used as a therapeutic test[17]. Sulfapyridines are also successful in some cases and are the drugs of choice next to dapsone. However, dapsone and sulfapyridine are not uniformly successful in the treatment of SPD[10]

Corticosteroids administered topically, orally or parenterally appear to be helpful[10].

Case Report

A 50 year old farmer presented with recurrent attacks of pustular eruptions on the trunk and extremities of 3 years' duration. Initially the lesions appeared on the trunk (scapular region) and 10-15 days after the onset, similar lesions appeared elsewhere on the body. The lesions would disappear in 4-6 weeks time leaving behind normal skin. The eruption was associated with moderate to severe itching and a burning sensation. There was no history of any seasonal variations, and various forms of treatment were of no help. There was no history of any systemic complaints.

Examination revealed multiple, very superficially situated pustular lesions (Fig. 1 Page No. 344) on the trunk, chest and proximal parts of the upper and lower extremities. The acral parts of the extremities, the face and the mucous membrane were spared. Some of the lesions were discrete while others were grouped in a circinate pattern with

surrounding erythema. At sites of healed lesions, hyperpigmented macules were present.

Routine laboratory examination of the blood and urine were within normal limits. Immunoglobulin levels were normal. Repeated smears and cultures from pustular lesions were negative for pyogenic organisms. Histopathology of a fresh lesion revealed a subcorneal pustule which was filled with mostly polymorphs (Fig. 2 Page No. 344). The epidermis except for mild spongiosis was normal. There was no acantholysis and the dermis was normal.

Patient was put on dapsone 100 mg twice a day with which the lesions were controlled in one week's time. However it relapsed on discontinuation of therapy. Reinstitution of dapsone therapy led to control of disease and the patient was put on 50 mg daily as a maintenance dose.

Comments

The morphology of the lesions and and the distribution on the classical sites suggested a diagnosis of subcorneal pustular dermatosis which was confirmed by histopathology and a therapeutic response to dapsone. As in the earlier reports from India[3-8], this patient again is a male. It was possible to keep this patient free of the disease on a maintenance dose of 50 mg dapsone per day.

The response to dapsone is rather characteristic. The disease being chronic in nature, the duration of therapy has to be long. A direct relationship exists between the dose of dapsone and the response of the lesions.

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