SUBCORNEAL PUSTULAR DERMATOSIS (SNEDDON-WILKINSON): REPORT OF A CASE

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In 1956 Sneddon and Wilkinson[1] identified and named a new vesiculo-pustular disease, subcorneal pustular dermatosis (SPD), which bears certain resemblances to some of the acquired bullous diseases (dermatitis herpetiformis, pemphigus, pemphigoid, benign familial pemphigus, and erythema multiforme) and to impetigo herpetiformis, parapsoriasis, pustular psoriasis, acrodermatitis continua of Hallopeau, and the pustular bacterides.

SPD affects women more often than men and appears mainly in the middle decades of life. It is a chronic disease

of exacerbations and spontaneous remissions, usually unassociated with any constitutional disorders. Pruritus is suffered by some patients but is not as striking a feature of the syndrome as it is in dermatitis herpetiformis.

The initial lesions of SPD are pustules that appear in crops surrounded by erythematous haloes and situated primarily in the flexures and on the upper parts of the trunk. The face, palms, soles and mucous membranes are almost invariably spared. The lesions rapidly rupture and coalesce to form crusted, circinate areas with actively spreading, festooned edges, eventually covering large portions of the body. In time the crusts fall off leaving dusky red or brownish hyperpigmented stains with no scarring or atrophy.

On histological examination the constant finding is a subcorneal pustule covered with a thin keratin layer, its base formed by the stratum granulosum and its cavity filled with polymorphonuclear leukocytes. There is no true acantholysis, and spongiosis is usually absent. The rete may be slightly acanthotic, with minimal oedema and exocytosis. In the upper corium there may be an infiltration by polymorphonuclear leukocytes and eosinophils.

The pus is sterile on culture and there is seldom any response to the usual antibiotics or to corticosteroids; sulphapyridine and diaminodiphenyl-sulphone (dapsone) are the only drugs that lead to improvement.

SPD may be differentiated histologically from dermatitis herpetiformis, erythema multiforme and bullous pemphigoid by the subcorneal situation of the bullae and by the absence of vascular dilatation and oedema. In pemphigus, including Senear-Usher syndrome, the bullae arise as the result of intraepidermal cleavage and acantholytic cells are therefore seen. In benign familial pemphigus. dyskeratosis is found besides acantholysis.

The bullae of SPD are very similar to those of impetigo herpetiformis, which, however, is a rare fulminating disease seldom seen in any but pregnant women, although Hall[2] has seen it in males and non-pregnant females. Unlike impetigo herpetiformis, SPD has sterile pustules and does not improve with antibiotics.[3] The mucous membranes are frequently involved in impetigo herpetiformis, erythema multiforme, and pemphigus, but never in SPD; a few cases, like that of Beck *et al.*,[4] have been reported as having mouth lesions and nail dystrophies, but Wilkinson[43] states that the mucous membranes have never been affected in the authoritative cases of Sneddon-Wilkinson disease.

The histological pictures of impetigo contagiosa and SPD are very similar, but in the former the bullae contain bacteria and the condition clears rapidly under treatment with ordinary antibiotics.

Sneddon, discussing the case Wilkinson[5] reported in 1951, said he originally thought that his first case of SPD was a variant of parapsoriasis appearing in a pustular form, but later decided that the peculiar distribution of the eruption and its response to sulphone therapy ruled this out.

SPD resembles dermatitis herpetiformis more closely than any other disease, particularly in its protracted course and its therapeutic response to sulphapyridine and sulphone; but SPD is seen in women more than in men, its pruritus is not pronounced, and in its flexural distribution and histological picture it is entirely unlike dermatitis herpetiformis.

Hellier[7] reported a case of SPD with a pustular bacteride on the palms and soles and postulated that SPD was more closely related to pustular bacterides than to dermatitis herpetiformis, in view of the differences in histological appearances and the absence of exacerbation in SPD when potassium iodide was given internally or locally applied to the skin. Greenbaum and Lee[8] disagree with the bacteride theory because in the pustular bacteride the pustule is situated more deeply and the lesions are essentially acral, as indeed they are in acrodermatitis continua.

Of the 50 or more cases recorded in the world literature[1-42] we believe ours to be the first South African case to be reported.

CASE REPORT

A 43-year-old European male clerk noted in January 1962 the sudden appearance in his axillae and groins of blisters that rapidly spread to the anterior aspect of the trunk. During the following weeks the condition was subject to remissions and exacerbations and he was troubled with some pruritus. His general health remained good. He described the blisters as being very thin-walled and delicate, usually clear at first, but tending to turbidity after a day or two and some becoming confluent. There was no history of his having taken any drugs before the onset and he had never suffered any previous skin disorder. He was, however, known to be allergic to penicillin. There was no family history of blistering of the skin. Before being seen by us the patient had been admitted on 14 January 1962 to another hospital with a provisional diagnosis of bullous impetigo. There was an initial temperature of 99·5°F, which returned to normal in 24 hours.

He was treated with oral antibiotics and an aerosol spray containing bacitracin and neomycin. At that time the urine showed no porphyrins or other abnormality, and on general physical examination he appeared to be normal except for his bullous skin lesions.

A direct examination of the contents of a pustule stained by Gram's method showed no micro-organisms, and appeared sterile on culture. A blood count was normal. He was discharged on 22 January slightly improved but with fresh lesions tending to erupt from time to time.

The patient was first seen by one of us (J.J.W.) in mid-February, when he had a very distinctive clinical pattern of skin lesions. There was an extensive bullous eruption mainly involving the anterior aspect of the trunk. The bullae appeared to be spreading over the chest and lower abdomen from the axillae and groins, the lesions tending to have a circinate and gyrate pattern with very clearly demarcated active borders. Central healing with some scaling and erythema was apparent in most of the affected areas. The bullae situated in the spreading borders were flaccid and thin-walled and, except for a few small fresh lesions, contained a turbid fluid, although in the larger bullae a clear supernatant fluid could be seen. There were some pigmented scars indicating the sites of previous healed lesions. There were no fresh bullae or scars on the back. Although individual bullous lesions did show some superficial resemblance to bullous impetigo, the general pattern was very similar to the clinical picture described so clearly by Sneddon and Wilkinson. (Fig. 1.)

Direct examination of skin scrapings showed no evidence of fungus. A biopsy was taken of a fresh lesion that had been present for less than 48 hours. Section showed it to be a subcorneal pustule. PAS stains for fungi were negative. The bullae contained neutrophils, but no eosinophils. No acantholytic cells were seen. (Fig. 2.)

A blood count showed a haemoglobin level of 16·8 G per 100 ml.; PCV 49%; MCHC 34%; white cells 10,000 per cu.mm. (polymorphs 46%, lymphocytes 42%, monocytes 6%, and eosinophils 6%).

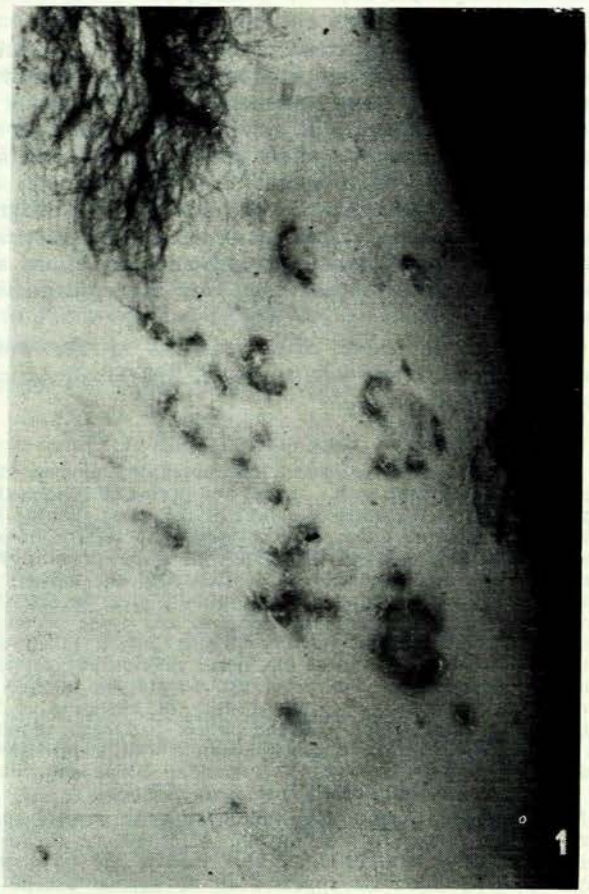

Fig. 1. The skin lesions of the subcorneal pustular dermatosis of Sneddon and Wilkinson. The photograph includes the left axilla.

The patient was given sulphapyridine 0·5 G *t.d.s.* and there was an immediate improvement in his skin condition. Unfortunately he was not very cooperative, and he was not seen again for 3 or 4 months in spite of repeated requests to attend the outpatient department. However, messages were received stating that his skin had cleared and had remained clear in spite of his having discontinued taking the sulphapyridine.

He was again seen in June 1962, when he had had no treatment for more than 6 weeks, and about 12 discrete vesicular lesions, 2 - 3 mm. in diameter, were found on various parts of the body, including one on the dorsal aspect of the left ring finger. Itching was still a troublesome feature. There was some pigmentation in old healed areas. At no time had he had any lesions in the mouth. He was then given dapsone, 50 mg. *t.d.s.*

Later he developed an erythematous eruption, which was attributed to sulphones, but he was not seen by us at the time nor has he been seen since in spite of numerous attempts to persuade him to return.

COMMENT

This case appeared to us to conform to the pattern of the subcorneal pustular dermatosis described by Sneddon and Wilkinson. The duration was rather shorter than most of the other cases but in every other way the features were characteristic. The general picture of annular, circinate and gyrate lesions spreading from the axillae and groins over the ventral surfaces, with flaccid pustules rupturing to form crusts, could hardly allow any alternative diagnosis. Additional features such as the recurrent nature, the pruritus, the definite response to sulphapyridine, and the failure to react to other antibiotics, were further confirmatory evidence.

The negative bacteriological findings and the lack of response to antibiotic treatment made the possibility of bullous impetigo remote. The absence of acantholysis, the apparently benign nature of the condition, and the response to sulphapyridine, excluded pemphigus vulgaris and pemphigus foliaceus, and the clinical and histological pictures did not conform to dermatitis herpetiformis.

We did not think that this case could possibly be related to impetigo herpetiformis of the type described in non-pregnant women and in men by Hall.[2] Our patient has so far been remarkably fit, except for his dermatosis, and one of our main difficulties has been to persuade him to attend our outpatient clinic now that he is more or less relieved of his symptoms.

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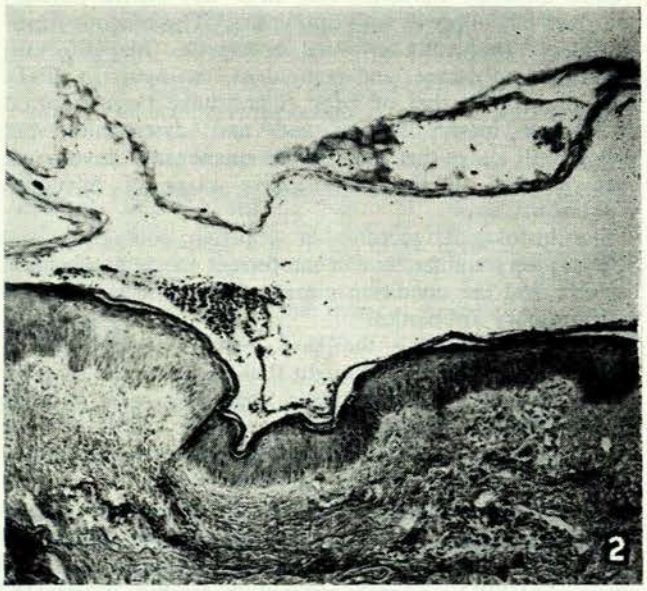

Fig. 2. The section reveals a subcorneal pustule containing neutrophils but no eosinophils. No acantholytic cells are seen.

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