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Views and Practice

Subcorneal pustulosis: what's in a name

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The conventional view of subcorneal pustular dermatosis (SPD), also known as Sneddon-Wilkinson disease, is a rare, chronic relapsing, vesiculopustular dermatosis with fairly characteristic clinical and pathological findings. Clinically, the patients are usually female, with a predilection for the trunk, intertriginous areas and the flexor aspect of the arms. There are recurrent crops of sterile pustules, usually distributed in annular or serpiginous patterns. The aetiology is unknown, although some cases are associated with a monoclonal gammopathy.

Pathologically, the lesions are characterised by subcorneal collections of neutrophils and pustules, with no indentation of the underlying epidermis, and no association with spongiform pustules. There is negative immunofluorescence staining, and no fungal elements are seen. The pustules are also typically sterile on microbiological culture. The pathological differential diagnoses include pustular psoriasis and IgA pemphigus. Pustular psoriasis typically has other features of psoriasis, including suprapapillary plate localisation and spongiosis, dermal papillary capillary dilatation and epidermal proliferation. IgA pemphigus has very similar clinical and pathological features to SPD but has demonstrable intercellular IgA on direct immunofluorescence studies. This has led some authors to consider all cases of SPD to represent IgA pemphigus, but there is a very definite subset of patients with no demonstrable IgA present in biopsy specimen, in which the designation of SPD or subcorneal pustular dermatosis should be retained. Consequently, the clinical and pathological findings of SPD remain characteristic, and justify the retention of this diagnosis as a specific diagnostic entity.

We saw two patients with possible subcorneal pustulosis in the dermatology clinic of a tertiary referral hospital in Sydney. One was a 54-year-old woman of European background with a 5-month history of a generalised pruritic vesiculopapular eruption. The other patient was a 42-year-old man of Asian background with a 1-year history of a relapsing and remitting pruritic eruption of pustules

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in flexural regions and over his chest, which later followed with desquamation.

In the first case, the skin biopsy demonstrated a focus of marked spongiosis with neutrophil infiltration and formation of a subcorneal/ intraepithelial pustule containing neutrophils, of which SPD was a differential provided other neutrophilic dermatoses were excluded. The skin biopsy of the second case demonstrated intracorneal pustule formation, with an inflammatory infiltrate of neutrophils and lymphocytes. Both cases responded to treatment with dapsone.

We note that whilst both these cases were suggestive of SPD from a clinical and pathological point of view, they are not "perfect fits". In the real world, life for dermatologists and pathologists is not always black and white. There are examples of confounding cases which may defy categorical diagnosis: the above two clinical cases are apt illustrations. Furthermore, the apparent rarity of this disease makes one wonder about its homogeneity: is it not possible for there to be a group of pustular dermatoses that are dapsone responsive but without the classic pathological features, such as our two cases here? Perhaps we should open our minds to the possibility that in the 2019 dermatological universe, there exist "no names" awaiting nosologic refocusing and redefinition.

Medicine cannot stand still and new or alternate ideas should be welcome. For this reason, the senior author feels that the two patients with interesting pustular eruptions should be accepted as at least variants of SPD and can be successfully treated as such. Progress through an open mind may prove to be a worthwhile intellectual exploration.