SUBCORNEAL PUSTULAR DERMATOSIS

Certain skin diseases, such as subcorneal pustular dermatosis, are observed mostly in mid-aged and mature women.

What is subcorneal pustular dermatosis (SPD)?

SPD, also called Sneddon-Wilkinson syndrome, is a rare rash that shows sterile pustules. The reason for this rash is unknown. It affects predominantly middle-aged or mature women. Development in younger women and males has been reported. It is rare in children and was reported only recently in pregnancy. The rash usually occurs symmetrically over the trunk, folds, especially the armpits, and flexures of the upper and lower limbs. The lesions are painless papules or vesicles with the size of a pea that later develop into flaccid pustules. The bottom half of the pustule contains pus whereas the top half clear fluid ('half-half' appearance or hypopyon pustule). The lesions merge together to form geographic or annular patterns.

Which conditions are associated with SPD?

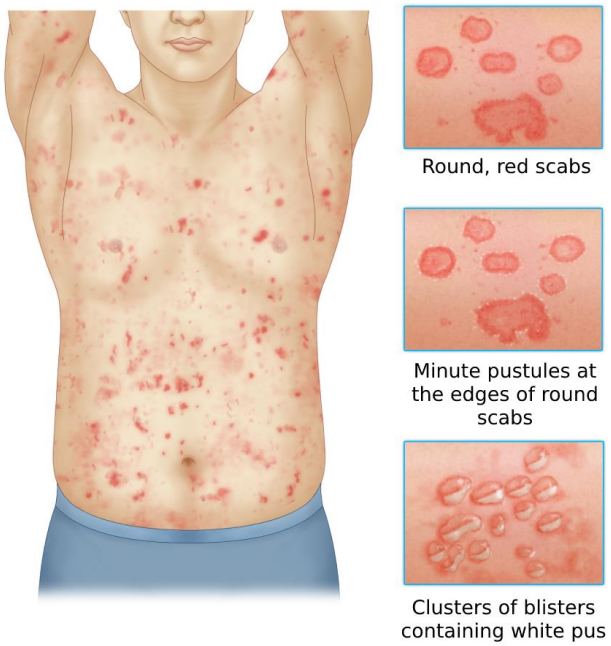
SPD can be associated with skin conditions called neutrophilic dermatoses, such as *pyoderma gangrenosum* and inflammatory bowel disorders (ulcerative colitis and Crohn's disease), hematologic disorders like lymphomas, multiple myeloma, chronic lymphocytic leukemia, aplastic anemia, and monoclonal gammopathies, solid organ malignancies, and connective tissue disorders such as Sjögren's syndrome, systemic lupus erythematosus, and rheumatoid arthritis. Also, drug reactions and infections, for example *Mycoplasma Pnemoniae* and primary pulmonary coccidiodomycosis, can trigger or aggravate SPD.

How is SPD diagnosed?

Diagnosis of SPD is established through the combination of typical clinical features of the rash and findings from skin biopsy that show a pustule sitting underneath the very top layer (stratum corneum) of the epidermis. Bacterial cultures from the lesions are negative.

Which conditions should be differentiated from SPD?

The SPD lesions can mimic pustular psoriasis, acute generalized exanthematous pustulosis which is usually triggered by a drug, blistering conditions such as pemphigus foliaceous and subcorneal type of IgA pemphigus, skin infections such as impetigo, dermatitis herpetiformis, and genetic syndromes with pustule formation. A skin biopsy is required for differentiation from other conditions. Also, a type of skin testing called direct immunofluorescence is needed to differentiate SPD from blistering disorders. The differentiation from other skin conditions requires dermatologic expertise. Therefore, generalized rashes with pustules should be referred to Dermatology.



Management

SPD is best treated with dapsone 50-150 mg per day taken by mouth. The response to dapsone is typically rapid, and lesions usually respond within 1-4 weeks. However, maintenance at lower dosage is required to prevent relapses. Other oral medications that can be beneficial include colchicine, sulfapyridine, and retinoids like acitretin and etretinate. Topical steroid medications can be used as adjunctive therapy. Newer drugs including biologic medications, such as adalimumab, infliximab and etanercept, are reserved for treatment resistant cases.

FOR MORE INFORMATION

Bhargava S, Kumar U, Kroumpouzos G. Subcorneal pustular dermatosis: comprehensive review and report of a case presenting in pregnancy. Int J Womens Dermatol. 2020:6(2):129-133.

Watts P, Khachemoune A. Subcorneal pustular dermatosis: a review of 30 years of progress. Am J Clin Dermatol. 2016;17(6):653-671.

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