AUTOIMMUNE BULLOUS DISEASES

THE USHER-SENEAR SYNDROME IN 19-YEAR-OLD FEMALE

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Background: Senear and Usher who published the first report about this condition called it "An Unusual Type of Pemphigus". The clinical picture of the disorder, which accounts for less than 10% of all pemphigus cases, embraces features of both lupus erythematosus and pemphigus foliaceus. Mucous membrane involvement is uncommon.

Observation: A 19-year-old female, presented with flaccid lesions, superficial vesicles and bullae of the skin as well as shallow erosions that commenced as erythematous papules over ears, scalp, upper back and "V" of the chest. Lesions were associated with sun exposure. The patient denied taking any drugs. The patient reported a lasting contact with plants, therefore phytophotodermatitis was suspected at first. Laboratory examinations revealed positive desmoglein 1 antibodies as well as positive ANA antibodies and low C3 level. Scalp biopsy showed superficial acantholysis and a perivascular dermal infiltrate with granular immune complex deposition at the dermoepidermal junction. The patient was diagnosed with pemphigus erythematosus based on clinical and immunological findings. The patient was refractory to standard oral steroid treatment and required extensive immunosuppressive therapy.

Key message: We report a case of pemphigus erythematosus in an otherwise healthy 19-year-old male. The clinical presentation of this condition may easily overlap with other forms of pemphigus, seborrheic dermatitis, impetigo or phytophotodermatitis. Patients and clinicians should be aware that there is no universally accepted treatment algorithm.