

PHOTOTHERAPY, PHOTODYNAMIC THERAPY

HAILEY-HAILEY DISEASE RESPONDING TO PHOTODYNAMIC THERAPY

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Background: Hailey-Hailey disease is an autosomal dominant genetic disease that causes degradation of desmosomes and subsequent acantholysis. It presents with chronic erosions and vegetations in the flexures between the ages of 20 to 40. Treatment is often difficult and there is a limited evidence base for intervention. We describe a case of Hailey-Hailey responding to photodynamic therapy. There are only 5 case reports/series equating to 15 patients where photodynamic therapy has been utilised for Hailey-Hailey. We would like to highlight this as a safe and effective treatment.

Observation: A 48 year old presented to our service in 2007 with a five year history of erosions, maceration and vegetative plaques in the inframammary region and groin. A biopsy confirmed Hailey-Hailey disease. The patient had no family history of such disease. She was treated with topical Clobetasol, and trialled on systemic agents, including Dapsone, Sulphamethoxypyradizine, Sulfasalazine, Lymecycline, Minocycline and Acitretin. She did not respond to or could not tolerate any of these treatments. She had severe complications from her disease, which included chronic pain, recurrent bacterial and herpetic infections and severe depression. We therefore decided to trial treatment with photodynamic therapy. Initially a small area was treated with methyl-amino-laevulinic acid and red light, then repeated seven days later. Within three weeks the lesions fully cleared and her response has been sustained two years later. Four areas in total have now been treated.

Key message: Current interventions for Hailey-Hailey have a limited evidence base and are often ineffective

We demonstrate that photodynamic therapy can be a safe and effective treatment for Hailey-Hailey disease and should be considered in refractory disease.

A single course of photodynamic therapy can give a prolonged period of remission, as demonstrated by our case.





