GENETICS AND GENODERMATOSES

RECALCITRANT HAILEY-HAILEY DISEASE TREATED SUCCESSFULLY WITH HYDROSURGICAL DEBRIDEMENT

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Background: Hailey-Hailey disease, or familial benign chronic pemphigus, is a rare blistering genodermatosis caused by an autosomal dominant mutation in the ATP2C1 gene leading to suprabasal acantholysis and giving the appearance of a dilapidated brick wall in the epidermis.
Intertriginous areas are more commonly symmetrically affected in young adults (third and fourth decades). Misdiagnosis are common (intertrigo, candidiasis, inverse psoriasis or frictional/contact dermatitis) and therefore inappropriate treatments are undertaken.
A correct diagnosis, however, involves a difficult management of the patient's disease especially due to chronic fluctuating course with subsequent remissions and relapses triggered by a variety of factors such as friction, stress, sweating, heat, moisture, superinfection, ultraviolet radiation, or tissue damage. Current treatments are not particularly effective and any individual's disease course may be difficult to predict.

Observation: A case of a 45 year old woman with a severe form of non responsive Hailey-Hayley disease involving axillary cables, groin abdomen, submammary fold is presented.
Numerous therapies such as oral and topical steroids, oral antibiotics, cyclosporine, methotrexate, photodynamic therapy, botulinum toxin type A, oral retinoids, magnesium chloride hexahydrate solution have been dispensed with partial relief. Given its considerable impact on the quality of life of this young and active patient, surgery has been performed. In particular, hydrosurgical debridement, in collaboration with plastic surgeons, has been adopted in order to achieve a long term remission with minimal scarring formation.

Key message: Although a complete remission is difficult to achieve, surgical debridement has to been taken into consideration for those non responder patients in order to guarantee a better quality of life.