





## Here is the answer to last month's case

### Meet Vlad

- Vlad is a 44-year-old male who has had a red sore with occasionally pruritic papules on the axillae for about 10 years. He also has skin tags on the axillae
- His doctors have suggested that he use a milder deodorant or no deodorant at all, but neither strategy seems to make much of a difference
- The rash is embarrassing, painful, and sometimes smells
- He has no personal or family history of skin problems
- He does not take any medications, nor is he allergic to any medications



### What is your diagnosis?

- |                                |                                |                       |
|--------------------------------|--------------------------------|-----------------------|
| a) Impetigo                    | c) Allergic contact dermatitis | e) Pemphigus vulgaris |
| b) Irritant contact dermatitis | d) Hailey-Hailey disease       |                       |

### Answer: D

Hailey-Hailey disease (**answer d**), or benign familial chronic pemphigus, is a rare, genetic, blistering skin condition. It typically presents in the third or fourth decade of life, but it can occur at any age. It often begins as painful erosive skin affecting skin folds, such as the axillae, groin, neck, intergluteal, or inframammary fold. The rash comes and goes, and often macerates or cracks. A secondary bacterial infection is a common occurrence, resulting in a foul odour and increasing local discomfort.

Heat, sweating, and friction appear to exacerbate the condition, and this explains why many

patients have worsening symptoms during the summer. This is an inherited condition, although sporadic cases have been reported as well. The defect is a gene called *ATP2C1* that encodes a calcium pump.

The condition is diagnosed by its clinical appearance and family history, although a biopsy is a useful confirmatory test. While there is no cure for Hailey-Hailey disease, management involves minimizing symptoms and flares. Avoidance of triggers, such as sunburn, sweating, and friction, is important. Topical steroids,

with or without topical antibiotics, can be useful. Occasionally, long-term courses of oral tetracyclines can be helpful. Phototherapy, ablative lasers, and botulinum toxin have also shown some benefit.

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*cme*

### Congratulations

to our winner for the month of  
March 2012

**Dr. Talita Strumpfer**  
Victoria British Columbia