**Jon’s Big Lump**

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**Meet Jon**

- Jon is a 28-year-old male with a large lump on his back that has been increasing in size over the past year.
- The lesion is occasionally tender but is nonpruritic.
- There is no discharge from the lesion.
- There is no personal or family history of malignancy.
- He is otherwise healthy, though he occasionally takes NSAIDs for headaches.

**What is your diagnosis?**

- a) Keloid
- b) Deep acne nodule
- c) Milia cyst
- d) Epidermoid cyst
- e) Lipoma

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See page 2 for the answer to last month’s case
Haeley-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 Haeley-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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