FIRST REPORTED CASE OF PATIENT WITH HAILEY-HAILEY DISEASE (HHD) DEVELOPING PEMPHIGUS FOLIACEUS

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Hailey-Hailey Disease (HDD) is a rare autosomal dominant skin condition discovered in 1939 by the Hailey brothers.\(^1\) It is usually observed in the third or fourth decade of life but has been reported at any age. Recent evidence shows HHD is due to a malfunctioning calcium pump.\(^2\) Protein SPCA1 regulates this process. Disruption of the level of calcium in the intracellular matrix leads to compromised desmosomes. Patients can suffer painful erosive and crusted skin due to suprabasilar acantholysis of the epidermis.\(^3\)

Pemphigus foliaceus (PF) is a rare autoimmune disease that has a blistering effect on the skin. The patient’s immune system mistakenly produces antibodies that attack the desmosomes of the cell which then separates keratinocytes. Fluid then fills the separated void and creates a blister. This disease can be induced by drugs or increased exposure to sunlight.\(^3\)

CASE REPORT

A 61-year-old-female presented to the dermatology clinic in April 2016. She was diagnosed with HHD at the age of 59. The patient also has a strong positive family history of the disease. The condition affects her son and mother. It is reported that most of her family members were diagnosed with HHD in the third decade of life. The patient consulted with numerous dermatologists before her visit to the clinic, but had found minimal relief. Her prior treatments included various oral and topical antibacterial agents, antiviral agents, antifungal agents, and steroidal agents.

A skin biopsy performed at the clinic demonstrated acantholytic dermatosis consistent with pemphigus foliaceus.

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Fig 1. Truncal rash on upper body

Fig 2. Crusted rash on lower and upper back

Fig 3. Rash on chest and upper arms

Fig 4. Rash with lesions on the face

Fig 5. Patient’s face after one week of prednisone.

Fig 6. Sample under immunofluorescence (IF)
Acantholytic dermatosis is known for causing a truncal rash consisting of crusted patches (Fig 1, Fig 2, Fig 3 and Fig 4). Direct immunofluorescence revealed intercellular IgG and C3 deposition (Fig 6). Indirect immunofluorescence (IIF) results showed 1:160 titters of IgG antiepithelial cell surface antibodies on monkey esophagus and normal results for other substrates.

The decision was made to treat the patient with systemic prednisone and azathioprine. Prednisone use would be gradually reduced. Within one week, the patient’s condition improved significantly (Fig 5). The patient still continues to improve.

DISCUSSION

After much debate, it was decided that the patient initially suffered from HHD and then later developed pemphigus foliaceus. Her case was of much interest because she is the first patient documented in the literature to have both conditions. It is possible that the patient’s HHD caused a reaction in her body that resulted in epitope unveiling.

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