Case Letter

Long-term Follow-up in a Case of Pemphigus

To the Editor:

Pemphigus is a chronic, autoimmune, epidermal immunobullous disorder with potentially fatal outcomes. It is caused by autoantibodies directed against components of the intercellular adhesion complex, desmoglein 1, and desmoglein 3. In India, pemphigus occurs in a younger age group compared to other populations, and childhood pemphigus also is common.[1] The natural course of the disease is characterized by remissions and relapses. Most relapses occur within 2 years after the diagnosis,[2] but late recurrences are well-documented.[3]

In 1992, Kanwar et al[4] reported a 35-year-old woman (who was part of our nursing staff) with pemphigus vulgaris that was diagnosed at 3 years of age; she was treated with oral corticosteroids. She became lesion free in 2 years and was in clinical remission for 30 years.[4] On further follow-up she continued to be in remission for the next 8 years when she had a relapse. No triggering factors could be identified. She was treated with dexamethasone-cyclophosphamide pulse therapy monthly for 6 months, followed by oral cyclophosphamide 1 mg/kg daily for 6 months. The patient was free of disease activity after therapy, and direct immunofluorescence was negative. At present, the patient is 57 years of age and free of disease. Enzyme-linked immunosorbent assay of recombinant baculoproteins of desmoglein 1 and desmoglein 3 for IgG antibodies, immunofluorescence study, and immunoblotting are negative.

The relapse seen in our case after a 38-year disease-free period is interesting. Reasons for the relapse are speculative. It could be because of memory B cells or perhaps some environmental trigger in a genetically susceptible individual.[5] Our case emphasizes that relapses may occur in pemphigus following a long period of remission and these patients require regular follow-up. Because the patient was part of our nursing staff, we were able to facilitate management and long-term follow-up. However, such close follow-up may not be possible with other patients. Nevertheless, we would like to share our experience with other clinicians in the dermatology field and emphasize the need for patient education on risk for relapse and need for regular clinical and immunological monitoring.

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