Köbner Phenomenon in a Rituximab-Treated Pemphigus Patient: Beware Disease Activity

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Introduction

Pemphigus consists of a group of rare autoimmune blistering diseases that are typically characterized by the appearance of intraepithelial blisters on the skin and/or mucosa. Pemphigus vulgaris (PV) and pemphigus foliaceus (PF) are 2 well-known forms of pemphigus. Although they have distinct clinicopathological features, each of these conditions could switch to the other. The Köbner phenomenon (KP) is described as the appearance of new lesions in the uninvolved skin as a consequence of different kinds of trauma. It has been described in several skin disorders, including psoriasis, vitiligo, lichen planus, and rarely pemphigus [1]. Here we report a patient with previously diagnosed mucocutaneous PV who developed PF after rituximab (RTX) therapy and developed crusted lesions some weeks after surgery for femoral avascular necrosis at the surgical site.

Case Presentation

A 55-year-old man with an 8-year history of mucocutaneous PV (confirmed with pathological findings and direct immunofluorescence) was treated with RTX in February 2017, after his disease had flared. Both anti-desmoglein (anti-Dsg) 1 and anti-Dsg3 were positive (>200 U/mL) at the time of infusion. His disease went into remission within 3 months after the last dose of RTX and remained on minimal therapy (5 mg prednisolone daily and 7.5 mg methotrexate weekly) for more than a year. In May 2018 he complained of the development of dry lesions on the chest, consistent with PF. After checking anti-Dsg1/3 titers, we found that anti-Dsg1 was 63 U/mL, while anti-Dsg3 was negative. In June 2018 a surgical procedure was performed in an attempt to treat femoral avascular necrosis. Despite the successful surgery, after 6 weeks new lesions appeared on the surgery site, which seemed to be related to KP (Figure 1, A and B). At the last follow-up in May 2019, the lesions were about to disappear via low-dose steroid treatment (Figure 2, A and B).

Conclusions

In addition to several suggested triggers for pemphigus, trauma can trigger the onset of new lesions in patients with pemphigus. A variety of procedures from minor skin sur-
If the surgery is urgent and patient is not in remission, increasing the dose of corticosteroid could probably prevent KP or at least mitigate its risk. However, further studies are needed to better understand approaches for preventing KP in patients with pemphigus.

References