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Discovering the cause helps the cure

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Belgi et al. describe a 37-year-old woman with a 4-month history of pemphigus vulgaris refractory to therapy despite several currently used medications. She experienced an exacerbation of her disease, and eventual clinical improvement and reduced antiepidermal antibody titres only 3 months after beginning treatment with rituximab, prednisolone, and mycophenolate mofetil.1

The case report lacks any mention of an attempt to identify the possible cause of the disease, although pemphigus is considered to stem from a genetic predisposition triggered and/or exacerbated by exogenous factors.2 The acronym PEMPHIGUS was proposed to summarize the triggering factors, standing for pesticides, malignancy, pharmaceuticals, hormones, infectious agents, gastronomy, ultraviolet radiation, and stress.2 Eliminating these factors can hasten the recovery and reduce the dosage and duration of pharmacological treatment.

The concept of induced pemphigus has been widely accepted in the literature since the first case report in 1969.3 The culprit drugs were categorized into three main groups by their chemical structure: (i) drugs containing a sulfhydryl radical, thiol drugs (including penicillamine, captopril, gold sodium thiomalate, penicillin and piroxicam) (ii) phenol drugs, containing phenolic compounds (including rifampin, levodopa, aspirin, and heroin); and (iii) nonthiol, nonphenol drugs (including some of the calcium-channel blockers, angiotensin-converting enzyme inhibitors, nonsteroidal anti-inflammatory drugs, dipyrone and glibenclamide).2 Topical contact, or contact pemphigus, is an additional triggering factor that has been attributed to gardening materials and pesticides.

Infectious diseases and immunizations have been implicated in inducing or exacerbating pemphigus, including viruses of the Herpetoviridae family, and bacteria such as coagulase-positive Staphylococcus aureus. Foods can also trigger pemphigus, including the allium family of vegetables (garlic, onion, shallot, chive and leek), the mustard family, the caper family (with a thiol group in their molecule) and the urshiol family [mango, cassava, areca nuts and cashew nuts (with a phenol group)]. Several studies point to the possible contribution of emotional stress as a precipitating factor in pemphigus,4,5 and pemphigus has long been considered to be a photosensitive disease.6

Eliminating the triggering or exacerbating factors is a vital step towards remission prior to pharmacological therapy. Thus, the management of pemphigus should include a thorough search for the culprit, and elimination of possible triggering factors such as sun exposure, emotional stress and certain foods. Furthermore, the continued use of triggering drugs may not only interfere with pemphigus treatment but can be the cause of its failure. The lack of immediate response to rituximab in the patient reported by Belgi et al. could be due to the fact that the cause of the disease had not been addressed. Discovering the exacerbating or triggering factors and their cessation will ameliorate the disease and reduce the need for medications that themselves carry potentially serious side-effects.

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