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Commentary

A Brief Note on Pemphigus Vulgaris

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ABOUT THE STUDY

Pemphigus vulgaris is the most frequent variant of pemphigus, a rare persistent blistering skin condition. The name Pemphigus comes from the Greek word pemphix, which means blister. It's a type II hypersensitivity reaction in which antibodies are generated against desmosomes, which are skin components that maintain different layers of skin connected together. The layers of skin split when desmosomes are assaulted, and the clinical appearance resembles a blister.

Acantholysis, or the breaking away of intercellular connections as a result of an autoantibody-mediated reaction, causes these blisters. Without therapy, the illness would definitely worsen: lesions will grow in size and spread throughout the body, acting physiologically like a serious burn.

Prior to the development of contemporary medicines, the illness had a near 90% fatality rate. Due to the introduction of corticosteroids as a main therapy, the mortality rate with treatment is now between 5-15 percent. Despite this, pemphigus vulgaris was the fourth leading cause of mortality from a skin illness in 1998.

The condition primarily affects middle-aged and older individuals, particularly those between the ages of 50 and 60. Women have typically had a greater rate of infection.

Signs and symptoms

The most common symptom of Pemphigus vulgaris is oral blisters (especially buccal and palatine mucosa), however it can also produce cutaneous blisters. Mucosal areas that may be affected include the conjunctiva, nose, oesophagus, penis, vulva, vagina, cervix, and anus. Blisters are prevalent, with flaccid skin and sparing of the skin covering the palms and soles.

Blisters often get infected, resulting in ulcerated sores and erosions. A positive Nikolsky sign indicates that you are sick (induction of blistering in normal skin or at the margin of a blister).

Severe chewing pain can lead to weight loss and malnutrition.

Pathophysiology

Antibodies against the desmoglein 1 and 3 proteins present in desmosomes cause pemphigus, an autoimmune disease. Keratinocyte cohesiveness in the epidermis breaks broken as a result of desmosome loss, compromising the barrier function that intact skin offers. Antibodies bind to antigens found in the body's own tissues, resulting in a type II hypersensitivity reaction. On histology, the basal keratinocytes typically stay attached to the basement membrane, giving the appearance of "tombstoning".

When transudative fluid gathers between keratinocytes and the basal layer (suprabasal split), a blister develops, resulting in a positive reaction. Bullous pemphigoid, on the other hand, is thought to be caused by anti-hemidesmosome antibodies and entails a separation of the epidermis and dermis (subepidermal bullae).

Diagnosis

Because it is an uncommon condition, diagnosis can be difficult and time-consuming. Individuals may develop oral erosions or skin blisters early on in the disease. Blisters that are itchy or painful are possible. The blisters should, in theory, show a positive Nikolsky's sign, in which the skin sloughs off when rubbed lightly, however this is not always the case. The gold standard for diagnosis is a punch biopsy from the area surrounding the lesion that is analysed by direct immunofluorescent labelling, in which cells are acantholytic, meaning they lack the typical intercellular connections that keep them together.

Impetigo and candidiasis are often mistaken with Pemphigus vulgaris. IgG4 is a pathogenic antibody. Testing for the infections that cause these other illnesses, as well as a lack of response to antibiotic therapy, helps confirm the diagnosis.

Treatment

Historically, corticosteroids and other immunosuppressive drugs have been used to treat pemphigus symptoms. However, steroids have substantial and long-term negative effects; therefore their

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use should be minimised as much as possible. Intravenous immunoglobulin, mycophenolate mofetil, methotrexate, azathioprine, and cyclophosphamide have all been tried and failed.

Monoclonal antibodies, such as rituximab, are a well-established alternative to steroids and are increasingly being utilised as first-line therapy. Following a successful fast track assessment, the FDA awarded full clearance to rituximab for this application in summer 2018. Many individuals attain remission with just one rituximab round, according to many case studies. Treatment is more effective if it begins early in the course of the disease, potentially even at the time of diagnosis. Rituximab therapy in combination with monthly IV immunoglobulin infusions resulted in long-term remission, with no disease recurrence 10 years after treatment was stopped. This was a modest trial study with 11 patients, 10 of whom were followed until the end.