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Pemphigus vulgaris– A case report

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Abstract
The uncommon autoimmune disease known as Pemphigus vulgaris (PV) is typified by autoantibodies that attack desmosomal proteins, especially desmogleins, which are essential for preserving cell-to-cell contact in epithelial tissues. Usually, the illness is characterized by excruciating erosions of the mucosa, which start in the mouth and spread to the skin, where they cause bullae and vesicles. Clinical assessment, histological analysis demonstrating acantholysis, and immunological testing, including enzyme-linked immunosorbent assay and direct immunofluorescence to identify anti-desmoglein antibodies, are the mainstays of the diagnosis process. The mainstay of management is immunosuppressive treatments, mainly systemic corticosteroids, supplemented with rituximab or azathioprine in instances that are not responding to treatment. To maximize patient outcomes, long-term care entails managing treatment side effects, keeping an eye out for disease activity, and offering supportive care. Notwithstanding advancements in treatment, PV still presents difficulties, necessitating multidisciplinary cooperation and continuous research to enhance patient care and quality of life.

Keywords: Acantholysis, epithelial tissues, immunofluorescence, assay, quality of life

Introduction

The Greek word for blister, pemphix, is the source of the name Pemphigus. Stephen Dickson initially wrote about pemphigus in 1788 after seeing a woman with a blister on her tongue. While PV is not as contagious as previously believed, people with other autoimmune illnesses may be susceptible to PV due to known triggers. [1, 1]. Acantholysis is the main feature of Pemphigus vulgaris (PV), an autoimmune illness that causes blisters on mucosal and cutaneous surfaces. Although the exact cause of pemphigus vulgaris is uncertain, several studies have connected the condition to autoantibodies that target cadherins, which in turn cause acantholysis and keratinocyte dissociation. Several investigations have connected PV to alleles of the HLA class II. Ashkenazi Jews have an association between PV and HLA-DRB1 0402; non-Jewish patients of European or Asian heritage have an association between PV and the HLA alleles DRB1 1401/04 and DQB1 0503. [1, 1]. A PV flare-up can be caused by a variety of environmental variables, including pesticides, food, stress, viruses, drugs, UV radiation, ionizing radiation therapy, and allergies.

Even while PV is common everywhere, it may also be correlated with geographic location and ethnicity. Nonetheless, certain ethnic groups report a greater rate. It has been discovered that Ashkenazi Jews are more likely to get PV, which typically manifests itself between the ages of 40 and 60. Additionally, the Middle East, Southeast Europe, and India have the highest rates of PV. In general, PV affects both men and women equally. Nonetheless, in Tunisia, women are four times more likely than males to have PV. Although familial occurrences of PV are rare, a genetic component has been demonstrated. [10] Compared to healthy controls, patients with PV had a higher proportion of nonsymptomatic first-degree relatives with circulating PV–immunoglobulin G (IgG) antibodies. Two of the most prevalent alleles linked to PV are DRB1*0402 and DQB1*0503; the latter is protective against rheumatoid arthritis. PV has been linked to ulcerative colitis and myasthenia gravis [3]. IgG autoantibodies against desmogleins cause disruptions in desmosome-mediated cell adhesion in Pemphigus vulgaris (PV), an autoimmune disease that results in blister development and acantholysis. PV mostly affects mucosal membranes at first; it generally starts with painful mouth erosions and then spreads to the skin, mainly excepting the palms and soles. Histologically, eosinophilic infiltrates and suprabasilar acantholysis are typical. Histopathologic analysis and direct immunofluorescence are used in the diagnosis process [4, 5]. Immunosuppressive therapy is used as part of the treatment to lower the formation of antibodies and maintain skin integrity, with an emphasis on the significance of early detection and management for better patient results. [3, 4]. The main symptom of Pemphigus vulgaris (PV) is painful intraoral blisters that burst into erosions, which can lead to malnutrition because oral intake is difficult. Later-developing vesicles, erosions, or bullae on different parts of the body— apart from the palms and soles- are indicative of cutaneous lesions. PV can cause considerable damage to mucosal surfaces, sometimes affecting nails, or manifest as pemphigus herpetiformis, which is marked by annular cutaneous vesicles and urticarial plaques. [2, 9]. A combination of clinical assessment, histopathologic testing (H&E biopsy exhibiting acantholysis), and direct immunofluorescence (DIF) revealing a distinctive netlike pattern of IgG in the epidermis are used to diagnose pemphigus vulgaris (PV). Serum IgG antibodies against desmoglein 1 (Dsg 1), desmoglein 3 (Dsg 3), or both are confirmed by ELISA testing. Tzanck smear, indirect immunofluorescence (IIF) on the monkey esophagus, and serological testing to identify circulating autoantibodies are further assessments. [2, 4]. For patients on systemic glucocorticoids, a thorough workup may include vital signs, pregnancy testing, CBC, metabolic panel, ANA, urinalysis, and bone density scan for early osteoporosis prevention. Tools such as the PDAI and ABSIS can be used to evaluate the quality of life and severity of the disease.

Case description

A 37-year-old female patient reported to the Venereology and Dermatology department, Sri Aurobindo Institute of

Medical Sciences, Indore with a complaint of itchy skin lesion over body. The patient was apparently alright one month when she developed clear fluid filled lesion over chest of around pea size followed by back and thigh lesion associated with burning and itching. The lesions ruptured within a day spontaneously leaving behind raw areas and crust formation. She developed raw painful areas in mouth which slowly spread to whole mouth and was associated with difficulty in swallowing. Her medical history was significant for Rheumatoid arthritis, and the patient was on Azathioprine and Omnacortil for 5 months. On physical examination, diffused edema over face and moonlike face along with multiple erythema plaque with crusting present over back, chest and multiple hyperpigmented patches present over back. Histopathological examination revealed suprabasilar blister formation associated with acantholysis. The patient did not agree for immunofluorescence analysis. Hence, a final diagnosis of oral PV was made based on histopathological pattern and clinical features. Treatment regimen included thorough prophylaxis; use of Fusidic acid cream 3–4 times daily for topical application; along with liquid paraffin, topically as advised Betamethasone ointment 3–4 times daily for topical application, tab levocetirizine twice daily, tab prednisolone twice daily and multivitamins once daily for 1 month. The patient was reviewed every 2 weeks for the first 1 month. The lesions had subsided with topical steroids within 4 weeks of starting the treatment. The patient was asked to stop the topical application, and reinforcement of hygiene instructions was given. Since the lesions can recur, the patient was kept under observation for 6 months and the lesions showed no signs of recurrence. She was advised to add liquid paraffin regularly topically.

Discussion

Autoantibodies against desmogleins cause Pemphigus vulgaris (PV), an autoimmune disease that disrupts cell-cell adhesion in mucous membranes and the skin, resulting in intraepithelial blister development and acantholysis. [6, 7]. Early signs and symptoms include painful erosions of the oral mucosa, which can hinder feeding, and skin lesions including bullae and vesicles. [5, 9]. Clinical evaluation, histology showing acantholysis, and immunological testing such as DIF and ELISA to identify anti-desmoglein antibodies are used in the diagnosis. [2, 3]. To minimize antibody generation and maintain skin integrity, management focuses on immunosuppression. Corticosteroids are frequently the first line of treatment, with rituximab or azathioprine being used as an adjuvant in patients that are refractory. [5, 9]. To improve quality of life, long-term care includes controlling treatment side effects, keeping an eye on disease activity, and making sure patients receive supportive care and education. [8, 10].

Conclusion

In conclusion, because PV is an autoimmune disease that targets desmosomal proteins, it poses a serious challenge to immunology and dermatology. Patients' nutritional condition and quality of life are negatively impacted by the disease, which presents with painful mucosal and skin sores. To confirm antibody-mediated acantholysis, the diagnosis is based on a combination of clinical, histological, and immunological assessments. Effective suppression of autoimmune reactions is the goal of treatment techniques, which also minimize treatment-related problems and highlight the importance of multidisciplinary care and long-term maintenance. To improve results and improve the general well-being of people with PV, further research and breakthroughs in therapy are required.

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