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Case Report

A case report on the management of pemphigus vulgaris along with comorbid conditions in geriatric patient

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ABSTRACT

Pemphigus vulgaris (PV) is a rare autoimmune blistering disease characterized by intraepidermal acantholysis, which affects the skin and mucous membranes. The patient was a 65-year-old female presenting with a history of type 2 diabetes mellitus and hypertension. There were fluid-filled, pruritic lesions that progressively spread from the forearms and neck to the face and other areas. Histopathological and immunofluorescence studies confirmed PV. She was treated with intravenous hydrocortisone, antibiotics, antihistamines, and supportive measures. The basic chronic kidney disease needed to be considered, and thus, conservative treatment was opted for. On being discharged, she was given a tapering dose of prednisolone along with other drugs that were prescribed for antidiabetic and antihypertensive drugs. It is a case that emphasizes the early and multidisciplinary need for diagnosis in PV, especially when comorbidities are present. Further research is also sought to improve the procedures of diagnosis and treatment of PV.

Keywords: Pemphigus vulgaris, Autoimmune blistering disease, Corticosteroids, Histopathology, Immunofluorescence

INTRODUCTION

The pemphigus diseases are a group of rare autoimmune blistering disorders affecting the skin and mucous membranes.[1] The word "pemphigus" originated from the Greek word "pemphix" meaning bubble or blister. It was introduced by Wichmann in 1791 to describe a chronic blistering disease now known as pemphigus vulgaris (PV).[2] The global incidence is 0.5–3.2 new cases per million persons yearly, with varying prevalence rates by region and ethnicity. In India, it is seen from 0.09% to 1.8%, which is lower than in other parts of the world. Pemphigus is more common in women than men; the ratio is 2:1, and it is most frequently observed in individuals in their 50s and 60s. The disease results from pathogenic autoantibodies that are usually of the immunoglobulin G (IgG) class against desmosomal proteins like desmogleins.[3-5] This autoimmune response disrupts intraepidermal adhesion, leading to acantholysis with the subsequent formation of vesicles, blisters, and erosions on the skin and mucous membranes. Major variants are PV and pemphigus foliaceus, these are distinguished by the level of acantholysis, suprabasilar in PV, and subcorneal in pemphigus foliaceus. Other variants include pemphigus vegetans, pemphigus erythematosus, paraneoplastic pemphigus, drug-induced pemphigus, and IGA pemphigus.[6] However, the treatment at an early stage of the disease would prevent progression to extensive skin involvement.

CASE REPORT

A 65-year-old female was admitted to Government Erode Medical College and Hospital with complaints of fluid-filled skin lesions accompanied by itching for 1 month. One month ago, she was in her usual state of health. She began developing these lesions initially on her forearm, V-area of the neck, and back. The lesions then spread to the face, ear lobules, and the back of the neck. The patient also reported constipation and itching of an intense nature over the lesions. No history of aggravation of lesions or itching by exposures to sunlight, and irritants like parthenium, chemicals, food allergies, and drug allergies is present. She has had type 2 diabetes mellitus and hypertension for the past 5 years for which she has been taking amlodipine 5 mg twice a day, glimepiride 1 mg twice a day, and metformin 500 mg twice a day. She had a personal history of mixed diet, with a good appetite. Her bowel and bladder habits were normal, except that she passed stool only once in the past 2 days and had decreased sleep due to itching. Her family history is negative for a similar illness. She has been married for the past 50 years, has two sons who are deceased due to renal tubular acidosis and alcoholic liver disease, and lives alone without family support. She has worked as a domestic servant, previously in agriculture.

General examination

The patient was conscious, oriented, and afebrile, with no signs of pallor, jaundice, cyanosis, clubbing, lymphadenopathy, or edema. Systemic examination was normal.

Cutaneous examination

Multiple well-defined erosions were measuring from 1 × 1 cm to 2 × 2 cm on the anterior aspect of the neck and medial aspect of the left wrist, with multiple vesicles/bullae and post-inflammatory hyperpigmentation. Similar lesions were present on her face, bilateral forearms, thighs, legs, and abdomen. Her hair and scalp were appropriate for her age, with dystrophic toenails. Oral and other mucosa, external genitalia, thyroid, and breasts were normal. There was no restriction of movement or joint tenderness.

Random blood sugar – 150, blood urea – 120, and serum creatinine – 2.4 are the parameters high in patients.

A skin biopsy from the bilateral forearms showed histopathological features of PV: Intraepidermal blister formation with acantholysis and the "tombstone" appearance of basal keratinocytes. Direct immunofluorescence revealed intercellular deposits of IgG and C3, which confirmed the diagnosis. Indirect immunofluorescence showed circulating IgG autoantibodies, and enzyme-linked immunosorbent assay indicated significantly raised levels of anti-desmoglein 1 and 3 antibodies. Chest X-ray revealed unfolding of the aorta and mild cardiomegaly.

This patient was found to have PV, chronic kidney disease, type 2 diabetes mellitus, and systemic hypertension. The patient was admitted for inpatient treatment with a salt-restricted diet of <2 g/day and fluid restriction of 1.5 L/day, along with an injection of hydrocortisone 100 mg intravenously every 8 h, injection cefotaxime 1 g twice daily intravenously, tablet ranitidine 150 mg twice daily, tablet a Vitamin C and zinc complex once daily, tablet chlorpheniramine maleate 4 mg nightly, tablet amlodipine 1 mg twice daily, tablet glimepiride 1 mg twice daily, tablet metformin twice daily, tablet furosemide 20 mg twice daily, and liquid paraffin in the morning and at night.

The patient was discharged the following week on the following medications and advice, which included a salt-restricted diet of <2 g/day, fluid restriction of 1.5 L/day, a tablet of prednisolone 20 mg twice a day, tablet ranitidine 150 mg twice daily, tablet Vitamin C and zinc complex once daily, tablet chlorpheniramine maleate 4 mg nightly, tablet amlodipine 5 mg twice daily, tablet glimepiride 1 mg twice daily, tablet metformin 500 mg twice daily, liquid paraffin E/A twice daily, and silver sulfadiazine cream E/A twice daily and review after 15 days.

DISCUSSION

PV is a rare, chronic autoimmune blistering condition characterized by intraepidermal blistering and erosions on the skin and mucous membranes.[2] The principal mechanism of pathogenesis involves autoantibodies directed against desmogleins 1 and 33. These are components of the desmosomes in the epidermis, playing a crucial role in cell-to-cell adhesion, which clinically results in flaccid blisters and erosions, as occurred with our patient.[3,7]

This 65-year-old female had a history of type 2 diabetes mellitus and hypertension, whose chief complaint was a 1-month history of pruritic, fluid-filled skin lesions that had spread progressively from the forearm and neck to include her face and other parts. The diagnosis of PV was entertained based on its classic presentation through clinical examination, histopathology, and immunofluorescence studies that showed intraepidermal blister formation, acantholysis, and intercellular deposits of IgG and C3.

The rationale behind the application of non-corticosteroid drugs in the treatment of pemphigus is to reduce the use of corticosteroids as much as possible to avoid or reduce their side effects and achieve better control over the disease. This, therefore, involves using immunosuppressive drugs such as cyclophosphamide, azathioprine, cyclosporine, methotrexate, and the latest one, mycophenolate mofetil. Anti-inflammatory drugs such as gold, dapsone, chloroquine, and a combination

of nicotinamide with tetracycline are also taken into consideration. The main immunosuppressive agents are introduced when the prednisolone dose has been reduced to 40 mg daily. If it is possible to discontinue prednisolone, these immunosuppressive drugs should be tapered gradually over few months as tolerated until a negative result is obtained from a direct immunofluorescence study.[8,9] These drugs are, in general, useless as solo agents and might require 4–8 weeks to act. Compared to azathioprine, cyclophosphamide is more effective but has more toxicity in the form of bone marrow suppression, hemorrhagic cystitis, bladder fibrosis, sterility, and an increased risk for malignancy. Major toxicities of azathioprine include bone marrow suppression, hepatotoxicity, and a possible increased risk of malignancy.[10] The usual dose is 2–3 mg/kg body weight. During therapy, total white cell count and platelet count should be done weekly for the 1st 8 weeks, then biweekly for 8 weeks, and once monthly thereafter. Urinalysis also must be performed in the same manner and cyclophosphamide is to be stopped if red blood cell appears in urine. Pulse cyclophosphamide 500 mg intravenously is also being tried alone' or with a concomitant pulse of intravenous corticosteroids.[11]

Her case was complicated by the presence of chronic kidney disease which necessitated an extremely cautious approach to management. Intravenous hydrocortisone was prescribed to this patient with widespread skin involvement and marked pruritus to control inflammation quickly. Other accompanying measures included antibiotics to prevent secondary infection, antihistamines for itching, and supportive measures such as nutritional supplements and topical treatments.

Comorbid chronic conditions of diabetes and hypertension needed co-management to prevent aggravation of her skin condition, as well as overall stability. This would be achieved by the institution of a salt-restricted and fluid-restricted diet and close monitoring of her glucose levels and blood pressure. She was discharged on a tapering dose of prednisolone to maintain the remission of PV, in addition to her pre-existing conditions, antihypertensives and antidiabetics were added, respectively.

The case underlines that, in patients with autoimmune blistering diseases and complex cases, there needs to be a multidisciplinary approach to managing such diseases, especially when comorbidities are more. Indeed, it is critical to make early diagnoses and institute prompt treatment for PV to prevent its complications and enhance the quality of life in affected subjects.

CONCLUSION

PV is a highly challenging condition, entailing early recognition and an overall treatment plan for the management of the core disease and its linked comorbidities. In this case, a patient who was prescribed a combination of corticosteroids and immunosuppressants, this supportive care was quite effective in demonstrating this approach. Moreover, the patient had hidden chronic conditions such as diabetes and hypertension, which were taken into consideration to have holistic management planning.

This case exemplifies the need for continued research and awareness regarding PV to help in the refinement of diagnostic tools and treatment protocols. Second, it also brings to light the importance of patient education and support, for it is a chronic condition with chances of relapse. It is by ensuring that our patient gets regular follow-ups and access to specialist care including nephrology consultation for her chronic kidney disease that long-term management and outcomes can be improved.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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