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A CASE REPORT ON PEMPHIGUS VULGARIS

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ABSTRACT

Pemphigus vulgaris (PV) is a rare autoimmune blistering disorder characterized by intraepidermal acantholysis, predominantly affecting middle-aged individuals with mucocutaneous lesions. We report a 65-year-old male presenting with throat pain, oral ulcers, bleeding, and skin lesions, eventually diagnosed with PV. This case highlights an atypical presentation without a known autoimmune history, stressing the need for clinical suspicion and timely dermatological consultation. The patient responded well to a combination of corticosteroids, antibiotics, antifungals, and symptomatic care. Early diagnosis and supportive management are key to preventing complications in PV. This case adds to the limited data on late-onset PV in immunocompetent males without prior history.

KEYWORDS:

Pemphigus vulgaris, autoimmune blistering disease, mucocutaneous lesions, elderly male, corticosteroids, case report.

INTRODUCTION

Pemphigus vulgaris (PV) is a rare, chronic, potentially life-threatening autoimmune blistering disease that primarily affects the skin and mucous membranes. It is characterized by the production of IgG autoantibodies directed against desmoglein 3 (Dsg3) and, in some cases, desmoglein 1 (Dsg1), which are critical components of desmosomes involved in keratinocyte adhesion [1]. Loss of these proteins causes intraepidermal acantholysis, resulting in flaccid blisters and erosions. PV has an annual incidence of approximately 0.1–0.5 cases per 100,000 population globally, with a higher prevalence in Ashkenazi Jews, people of Mediterranean descent, and individuals from India and the Middle East [2,3].

The typical age of onset is between 40 and 60 years, with a slight female predominance [4]. Clinically, patients often present initially with painful mucosal erosions, particularly in the oral cavity, followed by cutaneous involvement [5]. Diagnosis is confirmed through clinical examination, histopathology revealing suprabasal acantholysis, direct immunofluorescence (DIF), and enzyme-linked immunosorbent assay (ELISA) for anti-Dsg antibodies [6,7].

Treatment strategies have evolved from systemic corticosteroids alone to include adjuvant immunosuppressants (e.g., azathioprine, mycophenolate mofetil), biologics like rituximab, and newer targeted therapies [8]. Prompt diagnosis and intervention are crucial to prevent morbidity and mortality, which was historically high before the advent of modern immunosuppressive therapies [9].

CASE REPORT

A 65 year old male patient was admitted to General Medicine department with the complaints of throat pain for 4 days, oral ulcers, history of bleeding from oral cavity, skin lesion at chest and upper back. He had no past medical history. The patient was conscious and oriented, heart sounds were heard, chest was clear, was able to move all limbs and GI was non-tender. During admission, she had a Pulse Rate of 70 beats/min, Respiratory Rate of 22 breaths/min, Blood Pressure of 140/90mmHg. His laboratory investigation showed an elevation in HbA1C(9.2%), ESR(34mm/hr) and all other parameters were normal.USG Abdomen and Pelvis showed that Grade I fatty liver, Grade I prostatomegaly. Dermatology consultation was taken and confirmed as Pemphigus Vulgaris.

The patient was treated with IV antibiotics and supportive management including INJ. CEBACTUM (CEFTRIAXONE+SULBACTUM, 1.5gm, IV, BD) for treating infection. TAB. ISIBRO D (TRYPSIN (48MG) + BROMELAIN (90MG) + RUTOSIDE (100MG) + DICLOFENAC (50MG), TDS) for pain and T. AF(FLUCONAZOLE, 150mg, P/O,1-0-1) for fungal infection. TAB. HAPPI (RABEPRAZOLE 20mg P/O 1-0-0) for preventing gastric irritation, CHLORHEXIDINE MOUTHWASH for throat pain, SYP. MUCAINE GEL P/O 1-1-1 for treating ulcers in mouth, MUPIROCIN CREAM L/A 1-0-1 for

treating lesions. INJ. BETNESOL (BETAMETHASONE 4mg IV 1-0-1) for treating severe allergic reaction. Finally the patient was stable and discharged with TAB. ISIBRO D (TRYPSIN (48MG) + BROMELAIN (90MG) + RUTOSIDE (100MG) + DICLOFENAC (50MG), TDS), T. AF(FLUCONAZOLE, 150mg, P/O,1-0-1). TAB. HAPPI (RABEPRAZOLE 20mg P/O 1-0-0), CHLORHEXIDINE MOUTHWASH, SYP. MUCAINE GEL P/O 1-1-1, MUPIROCIN CREAM L/A 1-0-1 for 5 days and review after 5 days in General Medicine Outpatient Department.

DISCUSSION

Pemphigus vulgaris is most commonly observed in the 40–60-year age group with a slight female predominance.[10] However, our case features a 65-year-old male with no significant autoimmune or familial history, aligning with the demographic shift observed in recent studies from Eastern Europe and India.[11] For instance, a retrospective study by Baican et al. (2010) in Romania reported similar presentations in older patients without prior autoimmune disease.[12] Similarly, a large cohort study by Sinha et al. (2021) from India highlighted that environmental and dietary triggers, along with underlying metabolic disturbances like diabetes (as in our patient with elevated HbA1c), might influence PV onset in elderly males.[13]

In our case, the diagnosis was confirmed by dermatological evaluation without direct immunofluorescence or biopsy, which are considered gold standards. This reflects real-world constraints where resource limitations affect diagnostic thoroughness, yet clinical judgment remains vital. Treatment followed a standard empirical regimen with systemic corticosteroids (betamethasone) and antibiotics for secondary infection control. Compared to Ahmed et al. (2006), who emphasized rituximab and IVIG as effective first-line treatments for moderate to severe PV, our case demonstrates that conventional management may still be effective in mild-to-moderate cases.[14]

Unlike some patients who require prolonged immunosuppressive therapy or hospitalization due to complications, our patient stabilized within days and was discharged on oral maintenance therapy. This supports Werth's (2011) findings that early intervention significantly reduces morbidity in PV patients. The lack of mucosal progression or systemic involvement suggests a favorable prognosis, especially when diagnosed early and treated appropriately.[15] This case serves as a reminder for clinicians to consider PV even in atypical demographics and presentations, reinforcing the necessity of a multidisciplinary approach for timely management.

CONCLUSION

This case underscores the importance of clinical vigilance in patients with autoimmune diseases presenting with head and neck infections, as their immunocompromised state can lead to atypical presentations like left-sided quinsy with cervical lymphadenopathy. A multidisciplinary approach involving infection control and autoimmune disease management is vital. Early intervention with antibiotics, close monitoring, and continuation of baseline medications can result in complete recovery

without complications. Documenting such rare presentations is essential to broaden clinical understanding and guide future diagnostic and therapeutic decisions.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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