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Letter to the Editor - Study Letter

Demography and health-related quality of life of patients with pemphigus vulgaris and bullous pemphigoid treated at a tertiary care hospital: A cross-sectional study

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Dear Editor,

Pemphigus vulgaris and bullous pemphigoid are autoimmune vesiculobullous diseases. The reported annual incidence of pemphigus vulgaris is 0.1–0.5/100,000 population.[1] A systematic review and meta-analysis documented the incidence rate of bullous pemphigoid as 34.2 per million person-years.[2] Certain population groups (including Indians) are reported to be at a higher risk for pemphigus vulgaris and bullous pemphigoid.[3]

The remitting and relapsing nature of skin lesions, pain, burning, and pruritus associated with the diseases, widespread bullae and erosions, and severe mucosal involvement can adversely affect the life of the patients. Moreover, the financial burden of treatment and the adverse effects associated with the treatment modalities (systemic corticosteroids and immunosuppressants) affect the quality of life, even when the disease is under control. In addition, systemic steroids (mainstay of treatment for autoimmune bullous diseases) themselves can cause depression and psychiatric disturbances.[4]

In this setting, we did a cross-sectional study among patients who attended the dermatology department of our tertiary referral center from January 2019 to June 2020 and who received a diagnosis of pemphigus vulgaris or bullous pemphigoid (based on clinical, histopathological, and direct immunofluorescence findings).[5,6] The study aimed to document the patient profile, disease characteristics, and treatment received by the individual study participant. We also assessed the health-related quality of life using the dermatology life quality index (DLQI) questionnaire.[7] The ethics committee of the institution approved the study and individual study participant gave written, informed consent.

The study instruments included a standard data collection form (a triple validated data collection form by authorities in our department), a questionnaire to assess the DLQI {the permission and license for using the questionnaire were obtained (license ID – Cuqol2402)} and modified Kuppuswamy's socioeconomic scale (to assess the socioeconomic background of the study population).[7,8]

We collected data on demography and disease characteristics, investigation findings, and treatment received. To evaluate the health-related quality of life, the patients were asked to fill out the Malayalam version of the DLQI questionnaire. The questionnaire was very simple and self-explanatory. Filling up the questionnaire required only 2–3 minutes. Based on the score calculated by the questionnaire, the health-related quality of life was determined ("score of 0–1: no effect at all on patient's life, 2–5: small effect on patient's life, 6–10: moderate effect on patient's

life, 11–20: very large effect on patient's life, and 20–30: extremely large effect on patient's life").[7]

Details collected were recorded into Microsoft Excel sheets. Analysis of data was performed by SPSS Inc IBM company version 18 Chicago, SPSS Inc. (United States of America). Continuous variables were stated as mean and standard deviation (SD). Discrete variables were described as frequency and percentage. Unpaired *t*-test and ANOVA were applied to compare variations between continuous variables. $P < 0.05$ was considered statistically significant.

The study participants included 14 males (50%) and 14 (50%) females. The age ranged from 24–82 years (mean 52.6 years, SD 15.2 years.). Most of the patients (24, 85.7%) were married, two (7.1%) were unmarried, and two (7.1%) were widowed. Eighteen patients (64.3%) resided in urban areas and 10 (35.7%) were from rural areas.

As per Modified Kuppuswamy's socioeconomic status scale, 14 (50%) patients belonged to the upper lower socioeconomic class, 13 (46.4%) belonged to the lower middle socioeconomic class and one patient (3.6%) belonged to the upper middle socioeconomic class.

The comorbidities noted in the study participants were diabetes mellitus (11, 39.3%), hypertension (9, 32.1%), and dyslipidemia (4, 14.3%). Eight patients (28.6%) had other diseases which included chronic obstructive pulmonary disease, hypothyroidism, glaucoma, and coronary artery disease.

Four (14.3%) patients were smokers. Nine patients (32.1%) used to consume alcohol.

Eleven patients (39.3%) had pemphigus vulgaris and 17 (60.7%) had bullous pemphigoid. Twenty-two patients (78.6%) had a disease of recent onset and six (21.4%) had recurrent disease. Seventeen patients (60.7%) were on dexamethasone cyclophosphamide pulse therapy, 10 (35.7%) were on daily systemic corticosteroids, and 1 (3.6%) was on treatment with rituximab.

DLQI index of study participants ranged from 4–22 (mean 14.1, SD 4.5). Disease had a very large effect on the life of most of the study participants (20 patients, 71.4%). Disease had a moderate effect on the quality of life of 5 patients (17.9%), an extremely large effect on the quality of life of 2 patients (7.1%), and a small effect on the life of 1 patient (3.6%).

No significant association was noted between DLQI and age, gender, socioeconomic status, or residential area of the study participants.

The mean DLQI index of patients with the recent-onset disease was 15.09 ± 4.3, while the same was 10.5 ± 3.6 in those with recurrent disease. The difference was statistically significant ($P = 0.02$).

The equal gender distribution among study participants as noted by us was consistent with the literature on pemphigus vulgaris and bullous pemphigoid; however, a female predominance was noted by El Hafeez *et al*.[3,9] Our observation of a higher proportion of patients manifesting bullous pemphigoid than pemphigus vulgaris was discordant with the findings of Sobhan *et al*.[10] The previous authors have drawn attention to the adverse impact of pemphigus vulgaris and bullous pemphigoid on the quality of life of the affected.[11]

Specific tools to assess the quality of life in autoimmune bullous diseases are introduced (autoimmune bullous disease quality of life and treatment of autoimmune bullous disease quality of life), since it was suggested that the DLQI questionnaire is not disease-specific.[12,13] However, Ferries *et al*., after their prospective analysis of the correlation between disease severity scores in different autoimmune bullous diseases and quality of life assessed by different tools, did not find any added benefit for autoimmune bullous disease-specific tools over DLQI.[14]

The mean DLQI index of patients with recent onset disease (15.09 ± 4.3), as observed by us, was higher than the same noted by others (10 ± 6.7 and 10.9 ± 6.9) in new-onset, pemphigus vulgaris, and in patients with bullous pemphigoid (9.45 ± 3.34).[15-17]

The lack of association noted between the age of the affected and the quality of life in our study was consistent with the findings of Ghodsi *et al*. and Tabolli *et al*., but contrary to the observation of Paradisi *et al*., who reported a poorer quality of life among geriatric patients.[16,18,19] The lack of association noted between the gender and the quality of life was similar to the findings of many previous authors; however, Paradisi *et al*. reported that male patients enjoyed a better quality of life in comparison to females.[16,18-20]

Tabolli *et al*. and Ghodsi *et al*. did not find any association between quality of life and socioeconomic status in their studies, which was consistent with our findings.[16,18] Arbabi *et al*. documented that those with higher levels of education enjoyed a better quality of life.[20] The lack of association recorded by us between the residential area and the quality of life was consistent with the literature.[18]

The significant association noted between a high DLQI and a recent onset disease was consistent with the literature.[16] It is suggested that a patient with a disease of long duration would be better adjusted to the disease, while a patient with a recent onset disease may find it difficult to accept and live with the reality of a chronic disease. The disease would be better controlled with medication in patients with long-term disease, since the clinicians might have had adequate time to titrate the drugs according to the requirements of the specific patient.[16,21] However, another study noted no association between quality of life and duration of illness in patients with pemphigus.[22]

Cross-sectional study conducted in a single-center, limited number of cases, and considering pemphigus vulgaris and bullous pemphigoid as one entity (autoimmune bullous disease) for analysis were the major limitations.

Autoimmune vesiculobullous diseases showed a significant adverse impact on the health-related quality of life of those affected. Patients with a disease of recent onset were more affected than those with a recurrent disease. Prospective and multicenter studies with large sample size are needed to gather more information on the impact of autoimmune bullous diseases on the quality of life, which may help the clinician to provide comprehensive care to the affected.

Declaration of patient consent

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

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Conflicts of interest

Dr Anuja Elizabeth George is on the editorial board of the Journal.

REFERENCES

1. Ingold CJ, Khan MA. Pemphigus Vulgaris. Treasure Island, FL: StatPearls; 2021.
2. Persson MS, Begum N, Grainge MJ, Harman KE, Grindlay D, Gran S. The global incidence of bullous pemphigoid: A systematic review and meta-analysis. Br J Dermatol 2022;186:414-25.
3. Available from: https://www.rarediseases.org/rare-diseases/pemphigus [Last accessed on 2022 Jul 05].
4. Thibaut F. Corticosteroid-induced psychiatric disorders: Genetic studies are needed. Eur Arch Psychiatry Clin Neurosci 2019;269:623-5.
5. Hertl M, Jedlickova H, Karpati S, Marinovic B, Uzun S, Yayli S, *et al*. Pemphigus. S2 guideline for diagnosis and treatment--guided by the European dermatology forum (EDF) in cooperation with the European academy of dermatology and venereology (EADV). J Eur Acad Dermatol Venereol 2015;29:405-14.
6. Feliciani C, Joly P, Jonkman MF, Zambruno G, Zillikens D, Ioannides D, *et al*. Management of bullous pemphigoid: The European dermatology forum consensus in collaboration with the European Academy of dermatology and venereology. Br J Dermatol 2015;172:867-77.
7. Finlay AY, Khan G. Dermatology life quality index (DLQI) a simple practical measure for routine clinical use. Clin Exp Dermatol 1994;19:210-6.
8. Wani RT. Socioeconomic status scales-modified Kuppuswamy and Udai Pareekh's scale updated for 2019. J Family Med Prim Care 2019;8:1846-9.
9. El Hafeez NA, Hafez MK, Ahmed RF. Quality of life of patients with pemphigus vulgaris in Alexandria Egypt. World J Nurs Sci 2017;3:95-103.
10. Sobhan M, Farshchian M, Tamimi M. Spectrum of autoimmune vesiculobullous diseases in Iran: a 13-year retrospective study. Clin Cosmet Investig Dermatol 2016;9:15-20.
11. Padniewski JJ, Shaver RL, Schultz B, Pearson DR. Patient quality of life improvement in bullous disease: A review of primary literature and considerations for the clinician. Clin Cosmet Investig Dermatol 2022;15:27-42.
12. Sebaratnam DF, Hanna AM, Chee SN, Frew JW, Venugopal SS, Daniel BS, *et al*. Development of a quality- of-life instrument for autoimmune bullous disease: The autoimmune bullous disease quality of life questionnaire. JAMA Dermatol 2013;149:1186-91.
13. Tjokrowidjaja A, Daniel B, Frew J, Ebaratnam DF, Hanna AM, Chee S, *et al*. The development and validation of the treatment of autoimmune bullous disease quality of life questionnaire, a tool to measure the quality of life impacts of treatments used in patients with autoimmune blistering disease. Br J Dermatol 2013;169:1000-6.
14. Ferries L, Gillibert A, Duvert-Lehembre S, Corbaux C, Alexandre M, Prost-Squarcioni C, *et al*. Sensitivity to change and correlation between the autoimmune bullous disease quality-of-life questionnaires ABQOL and TABQOL, and objective severity scores. Br J Dermatol 2020;183:944-5.
15. Patsatsi A, Kokolios M, Kyriakou A, Lamprou F, Stylianidou D, Tsapas A, *et al*. Quality of life in Greek patients with autoimmune bullous diseases assessed with ABQOL and TABQOL indexes. Acta Derm Venereol 2017;97:1145-7.
16. Ghodsi SZ, Chams-Davatchi C, Daneshpazhooh M, Valikhani M, Esmaili N. Quality of life and psychological status of patients with pemphigus vulgaris using dermatology life quality index and general health questionnaires. J Dermatol 2012;39:141-4.
17. Kouris A, Platsidaki E, Christodoulou C, Armyra K, Korkoliakou P, Stefanaki C, *et al*. Quality of life, depression, anxiety and loneliness in patients with bullous pemphigoid. A case control study. An Bras Dermatol 2016;91:601-3.
18. Tabolli S, Mozzetta A, Antinone V, Alfani S, Cianchini G, Abeni D. The health impact of pemphigus vulgaris and pemphigus foliaceus assessed using the Medical Outcomes Study 36-item short form health survey questionnaire. Br J Dermatol 2008;158:1029-34.
19. Paradisi A, Sampogna F, Di Pietro C, Cianchini G, Didona B, Ferri R, *et al*. Quality-of-life assessment in patients with pemphigus using a minimum set of evaluation tools. J Am Acad Dermatol 2009;60:261-9.
20. Arbabi M, Ghodsi Z, Mahdanian A, Noormohammadi N, Shalileh K, Darvish F, *et al*. Mental health in patients with pemphigus: an issue to worth consideration. Indian J Dermatol 2011;56:541.
21. Wysoczyńska K, Żebrowska A, Waszczykowska E. Quality of life in patients with pemphigus. Przegl Dermatol 2013;100:139-45.
22. Tee CT, Lee CS, Gunabalasingam P. Characteristics and quality of life in pemphigus patients. Med J Malaysia 2022;77:324-30.

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