Family dermatology life quality index in patients with pemphigus vulgaris: A cross-sectional study

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Abstract

Background and Aims: Pemphigus vulgaris is a rare autoimmune intraepidermal vesiculobullous disease involving the skin and mucosa. It impacts the quality of life of both patients and their families.

Methods: A total of 70 patients with pemphigus vulgaris (either outpatient or hospitalized) were enrolled using the simple sampling method between 2016 and 2017 from the dermatology clinic at Faghihi Hospital, Shiraz, Iran. A validated Persian version of the Family Dermatology Life Quality Index (FLDQI) questionnaire was filled by a family caregiver. The questionnaire contained 10 items assessing the quality of life of the family. Demographic variables were recorded in a separate form.

Results: The mean age of the patients was 51 ± 11.3 years and that of the family caregivers was 32 ± 8.8 years. The FLDQI score was higher (poorer quality of life) if the patient was male, older, had shorter disease duration or had fewer disease recurrences (P = 0.046, 0.01, 0.001 and >0.001, respectively). Higher scores were also obtained in the less-educated caregivers (P = 0.026) but there was no association with either gender or age (P = 0.399, 0.1).

Conclusion: Pemphigus vulgaris significantly affects the Family Dermatology Life Quality Index. Education and counseling of family caregivers by various support groups such as Pemphigus Family Associations could be effective in improving the quality of life of the caregivers.

Limitations: This study did not assess the effect of comprising domain analysis, severity of disease, patients' Dermatology Life Quality Index (DLQI), mucosal involvement, response to treatment, outpatient or admitted status, socioeconomic status, or the quality of life among the various family members.

Key words: Caregivers, family dermatology life quality index, pemphigus vulgaris

Introduction

Pemphigus vulgaris is a rare autoimmune intraepidermal vesiculobullous disease affecting the skin and mucosa most often in the sixth decade of life.[1,2] Immunoglobulin G autoantibodies targeting desmoglein 1 and 3 cell adhesion molecules are linked to its pathogenesis.[3-8] The disease is rare, with an estimated global incidence of 0.076:100,000 and a male–female ratio of 1:1.1–2.25.[7,9,10]

Owing both to the nature of the disease and the prolonged treatment that it entails, pemphigus vulgaris gravely impacts the quality of life (QOL) of the patient.[11] It also affects the QOL of the caregivers and family and this further negatively impacts patient care. Most studies have assessed the QOL of the patients but not of the caregivers or the families. A number of tools have been developed for assessing the QOL of caregivers of patients with chronic dermatologic diseases.[12,13] In view of the importance of the QOL of caregivers and the family and its influence on patient management,[14-19] we studied the impact of pemphigus on the Family Dermatology Life Quality Index (FDLQI).

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Methods

This cross-sectional study was conducted among patients referred to the dermatology clinic in Faghihi Hospital, Shiraz, Iran between 2016 and 2017. The study was approved by the Ethics Committee of Shiraz University of Medical Sciences (code: 94-01-01-9758). All the patients and their caregivers gave written informed consent.

Seventy patients (outpatient or hospitalized, significant difference = 1.5; standard deviation = 6.4; α = 0.05; 1−β = 0.9) with pemphigus vulgaris were selected by the simple sampling method and their caregivers were enlisted for the study. All patients were diagnosed clinically by a dermatologist, and the diagnosis was confirmed by skin biopsy. A physician confirmed that the selected patients did not suffer from any other health conditions that could affect the quality of life, by history and face-to-face interview.

Family caregivers enrolled for the study were usually first-degrees relative aged 18 years or more. Most patients had a single caregiver; however in the event of multiple caregivers, one of them was randomly selected for the study. The study objectives were carefully explained to each caregiver.

A validated Persian version of the FDLQI questionnaire was filled by the family caregiver. The questionnaire contained 10 items assessing the QOL of the family.[20,21] The scores for each question ranged from 0 to 3 and the total score was the sum of the scores for the 10 questions. Demographic variables were recorded in a separate form.

The Statistical Package for the Social Sciences software (IBM Corp., released in 2013; IBM SPSS Statistics for Windows, version 22.0, Armonk, NY; IBM Corp.) was used for statistical analysis. After checking the normality by Kolmogorov–Smirnov's test, the mean ± standard deviation or median (interquartile range) was reported. Independent *t*-test (or Wilcoxon rank-sum test), one-way analysis of variance (or Kruskal–Wallis test) and Pearson's correlation *r* were used. $P \leq 0.05$ was considered as statistically significant.

Results

The questionnaire was completed by all 70 caregivers. Their ages ranged from 18 to 53 years (mean 32 ± 8.8 years). No correlation was observed between the age of the caregivers and the total scores ($P = 0.1$, Pearson's correlation $r = 0.198$). Demographic and clinical features of the patients and caregivers are shown in Tables 1 and 2. Pemphigus had a moderate to severe impact on the QOL of the caregivers with 39 (55.7%) caregivers scoring 10–20 and 18 (25.7%) caregivers scoring >20. Caregivers often suffered from emotional distress and depression, but other domains of QOL were also affected including time spent on looking after the patient, physical wellbeing such as sleep and rest, recreation and leisure activities, and extra house-work. Caregivers were less affected by issues such as staying away from job or study, reduction in work hours, peoples' reactions to the patient, social life problems, household expenditures and disruption of family relationships.

Spouses had higher scores (poorer QOL) as compared to other relatives ($P = 0.0005$). Twenty of the family caregivers were poorly educated (illiterate and under high school diploma) and 50 were well educated (high school diploma and above) - a poorer QOL was seen in the the less educated caregivers (19.9 ± 7 vs. 14.3 ± 7.3; $P = 0.005$). There was no significant difference between the genders (17 ± 8.6 vs. 15.7 ± 4.2; $P = 0.399$) [Table 2], but a poorer quality of life was noted in caregivers of male patients (18 ± 8.6 vs. 14.4 ± 6.5; $P = 0.046$) and older patients ($P = 0.01$, Pearson's $r = 0.305$).

All patients were treated with prednisolone and none of them had received rituximab. The FDLQI was not influenced by the type of adjuvant used: azathioprine (15.7 ± 8 vs. 16.9 ± 5.8; $P = 0.554$) or cyclosporine (18.8 ± 9 vs. 15.7 ± 7.6; $P = 0.436$). The duration of the disease (2 ± 2.7 years) was negatively correlated with scores ($P = 0.001$) i.e. a lower impact on the FDLQI was seen in caregivers of patients with a shorter disease duration. The impact on quality of life

Table 1: Demographic and clinical features of the pemphigus patients in the study

Variable	*n* (%)	Mean score obtained by family caregiver±SD	*P*
Sex			
Male	30 (42.9)	18±8.6	0.046[a]
Female	40 (57.1)	14.4±6.5	
Age	51±11.3	16.7±7	0.01 (0.305)[b]
Disease duration			
<1 month	25 (35.7)	20.56±7	0.001[c]
1-12 months	20 (28.6)	14.7±25.2	
1-5 years	14 (20)	13.5±7.2	
>5 years	11 (15.7)	11.55±5.3	
Recurrence episodes			
1	28 (40)	19.5±7.4	<0.001 (−0.494)[b]
2	21 (30)	16.2±7.6	
3	13 (18.6)	12.1±4.5	
4	2 (2.8)	15.5±3.5	
5	6 (8.6)	6.7±4.9	
Medications			
Prednisolone	70 (100)	-	-
Azathioprine	58 (82.9)	15.7±8 vs. 16.9±5.8[e]	0.554[d]
Cyclosporine	6 (8.6)	18.8±9 vs. 15.7±7.6	0.436[d]
Rituximab	0	-	-

[a]Wilcoxon rank-sum test, [b]Pearson's correlation *r*, [c]Kruskal-Wallis, [d]Independent *t*-test, [e]Medication positive vs. medication negative.
SD: Standard deviation

Table 2: Demographic features of the family caregivers

Features	*n* (%)	Mean score±SD	*P*
Gender			
Male	23 (32.9)	17±8.6	0.399[a]
Female	47 (67.1)	15.7±4.2	
Age (years)	32±8.8	16±7.7	0.1 (0.198)[b]
Relationship			
Spouse	14 (20)	20±8.2	0.0005[c]
Child	38 (54.3)	17.1±7.3	
Sibling	18 (25.7)	10.4±4.8	
Educational level			
Below high school diploma	20 (28.6)	19.9±7	0.026[c]
High school diploma	34 (48.5)	13.8±6.8	
Bachelor `s degree	13 (18.6)	14.6±8.7	
Master`s degree and higher	3 (4.3)	19±9.6	

[a]Independent *t*-test, [b]Pearson's correlation *r*, [c]Kruskal-Wallis. SD: Standard deviation

was less in caregivers of patients with more frequent recurrences ($P < 0.001$, Pearson's $r = -0.494$) [Table 1].

Discussion

Chronic diseases can affect many aspects of the quality of life of both patients and their families including their social, mental and physical health, living expenses, time spent with the patient and issues related to job or studies.[16,21-26]

In the present study, the most affected aspects of the quality of life of caregivers were emotional distress and depression, time spent on looking after the patient, physical wellbeing such as sleep and rest, recreation and leisure activities, and extra house-work. The least affected domains were staying away from job or study, working-hour reduction, peoples' reactions to the patient, social life problems, household expenditures and disruption of family relationships. However, in atopic dermatitis and epidermolysis bullosa household expenditure and leisure time were most affected,[26-28] while in caregivers of psoriatic patients major concerns were the impact on job and reduced working hours.[24] Higher divorce rates were also seen in the families of children with atopic dermatitis, but the family quality of life was better in daughters.[29] Caregivers of patients with multiple sclerosis believed that the disease had a negative impact on intra-family relationships including tension and decreasing mutual understanding.[30]

We found that caregivers of male patients were more dissatisfied which may be due to the nature of the disease, patients' responsibilities to the family (especially household expenditure) and local culture.[29,31] Spouses were more impacted as compared to other relatives, possibly due to a higher burden on the spouse to manage the responsibilities which the patients could no longer handle. Higher levels of education were directly linked to a better quality of life - the more educated caregiver may be able to understand the disease better and thus cope better; the higher income of these caregivers may also translate into improved care and facilities.

Decreased caregivers scores were observed with longer disease duration and more recurrences. The shock of being diagnosed with a dreaded chronic disease, initial denial, and subsequent acceptance and adjustment to the illness may also explain this observation. It is likely that with time both patient and caregiver learn to cope with the chronicity of the disease and return to a near normal life.

Limitations of our study include the fact that we did not assess such factors as the quality of life in different family members or the effects of socioeconomic status. These will be addressed in future studies by interviewing more than one caregiver. Several variables comprising domain analysis, severity of disease, patients' DLQI, mucosal involvement, response to treatment and outpatient or admitted status could be evaluated to define the family caregiver score, in more detail.

Conclusion

We studied the FDLQI among patients with pemphigus vulgaris and found significant impact on the QOL of the caregivers. Pemphigus family associations and support groups may be encouraged to step up support and educate family caregivers regarding the disease in order to improve the quality of life of both caregivers and patients.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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