Dermatology Life Quality Index (DLQI) in Libyan pemphigus patients

Abstract:

Introduction:

Pemphigus refers to a family of rare acantholytic autoimmune dermatoses of the mucocutaneous membranes in which acantholysis, or the loss of cell-to-cell adhesion, causes potentially lethal bullae and erosion formation. Multiple subtypes of pemphigus disease have been identified based on their distinct clinical features and pathophysiology, including pemphigus vulgaris (PV), pemphigus foliaceus (PF), IgA pemphigus, and paraneoplastic pemphigus (PNP).

Aim of the study:

To measure the Dermatology Life Quality Index (DLQI) in Libyan pemphigus patients and assess the impact of the disease on their lives.

Materials and methods:

In this descriptive study, 25 patients with pemphigus attending Bullous clinic, dermatology department, JumhoriaHospital, Benghazi - Libya, over ~~the course of~~one year were enrolled.

All patients were registered in the clinic, and the diagnosis of pemphigus was confirmed by histopathology.

For every patient, the DLQI score was measured by filling out the questionnaire.

Results:

Among the 25 pemphigus patients included in this study, 20 patients (80%) were females and 5 patients (20%) were males. The age ranged from 30 to 72 years, and the mean age was 43.64 years. The duration of the study ranged from 1 to 408 months (mean: 82.12 months). Pemphigus vulgaris was encountered in 13 patients (52%). Concerning disease severity, 44% of cases presented with a severe illness. In 15 patients (60%), mucous membranes were not involved. Eight patients (32%) were treated with steroids and cytotoxic drugs. Dermatology Qualify Life Index (DLQI) scores showed a large effect on life quality in 56% of the study cases and a moderate effect in 32% of them. The mean DLQI scores for pemphigus vulgaris were 12.67 and for pemphigus folaiceous were 10.31.

Conclusion: Pemphigus significantly affects DLQI scores in Libyan patients and lower scores were reported in treated patients with systemic combined therapy.

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Keywords: Pemphigus, pemphigus vulgaris, Demographic features, Dermatology life quality index (DLQI), Libya.

Introduction:

Pemphigus refers to a family of rare acantholytic autoimmune dermatoses of the mucocutaneous membranes in which acantholysis, or the loss of cell-to-cell adhesion, causes potentially lethal bullae and erosion formation. Multiple subtypes of pemphigus disease have been identified based on their distinct clinical features and pathophysiology, including pemphigus vulgaris (PV), pemphigus foliaceus (PF), IgA pemphigus, and paraneoplastic pemphigus (PNP; ~~). (~~1,2). Pemphigus occurs worldwide but has a disproportionate geographic and ethnic distribution, with a significantly higher prevalence in patients of Ashkenazi Jewish or Mediterranean descent.
(1,2,3,4). Despite its increased prevalence in these populations, pemphigus universally affects all racial and ethnic groups (3,4,5). The prevalence of pemphigus disease in Ashkenazi Jewish

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populations may be attributed to the increased presence of several distinct HLA class II genes, specifically HLA-DRB1*04 and HLA-A*10 (6, 7, 8)

Aim of the study

1) To study the demographic features of pemphigus in Libyan patients.
2) To measure the Dermatology Life Quality Index (DLQI) in pemphigus patients and assess the impact of the disease on their lives.

Materials and methods:

In this descriptive study, 25 patients with pemphigus attending Bullous clinic, dermatology department, Jumhoria~~hospital~~Hospital, Benghazi - Libya over one year were enrolled.
Each patient was exposed ~~to~~detailed disease and treatment history and complete dermatological examination according to the prepared proforma. All patients were registered in the clinic and the diagnosis of pemphigus was confirmed by histopathology.
For every patient, the DQLI score was measured by filling out the questionnaire which includes 6 items: symptoms and feelings (Questions 1 and 2), daily activities (Questions 3 and 4), leisure (Questions 5 and 6), and personal relationships (Questions 8 and 9), each with a maximum score of 6, work and school (Question 6), and treatment (Question 10), each with a maximum score of 3.
The scoring for each question is as following:
Very much:~~-~~ 3
Alot:~~-~~ 2
A little:~~-~~ 1
Not at all:~~-~~ 0
Not relevant:~~-~~ 0
Unanswered:~~-~~ 0
Question 7 "prevented work or studying" scored 3.
DLQI is calculated by summing the scores of each questions resulting in a maximum of 30 and a minimum of 0. The higher the score, the more quality of life is impaired.

Meaning of DLQI scores:
0-1 = No effects at all on the patient's life.
2-5 = Small effect on patient's life.
6-10 = Moderate effect on the patient's life.
11-20 = Very large effect on the patient's life.
21-30 = Extremely large effect on the patient's life.

Statistical analysis:
Data were analyzed using the statistical package for S~~s~~ocial S~~s~~cience (SPSS), version 23. Descriptive statistics: the mean, standard deviation, and median were estimated. Inferential statistics were applied as needed. Student t-tests were used to find the difference between the means of the groups and Chi-square (X^2) to find the difference in the distribution of the categorical variables between the groups. P-value was considered significant when ≤ 0.05.

Results:

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Among the 25 pemphigus patients included in this study, 20 patients (80%) were females and 5 patients (20%) were males. The age ranged from 30 to 72 years, and the mean age was 43.64 years. The duration of the study ranged from 1 to 408 months (mean: 82.12 months) (Table 1).

Pemphigus vulgaris was encountered in 13 patients (52%) (Table 2). Concerning disease severity, 44% of cases presented with a severe illness (Figure 1). In 15 patients (60%), mucous membranes were not involved (Figure 2). Eight patients (32%) were treated with steroid and cytotoxic drugs, and 7 patients (28%) were treated with steroid only (Figure 3).

Dermatology Life Qualify Index (DLQI) scores showed a large effect on life quality in 56% of the study cases and a moderate effect in 32% of them (Figure 4). The mean DLQI scores for pemphigus vulgaris were 12.67 and for pemphigus folaiceous were 10.31 (Table 3).

DLQI scores were lower in cases where pemphigus had partial skin involvement or was generalized without mucous membrane involvement (mean score: 9.6 and 9.22, respectively) (Table 4).

In~~(~~Table 5~~)~~ shows the mean scores for different pemphigus treatments received, in which patients who were on systemic steroids and cytotoxic drugs had a lower mean score (10.50), followed by systemic steroids (10.71).

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Discussion:

Pemphigus largely affects patients between the ages of 50 and 60, although the mean age at diagnosis can differ significantly based on the country of origin and ethnic background. In some Western Asian nations, such as Kuwait, the mean age at diagnosis is 36.5 years, whereas in European nations such as Bulgaria, the mean age at diagnosis is 72.4 years (9). In our study, the mean age of the participants was 43.6 years, and the majority of them were females (80%).

Importantly, disease onset in the pediatric population has also been described, including in patients as young as 6 years old (10).

In France, pemphigus vulgaris accounted for 73% of all cases; incidence increased with age; and the female-to-male sex ratio was 1:2. (2). In Tunisia, pemphigus foliaceus was more frequent (61%), the female-to-male sex ratio was 4:1, and the incidence rate was higher in young women (2). The incidence rate was 15.5 cases per million per year for pemphigus foliaceus among women aged 25 to 34 years and was even higher in some rural areas. (2). In our patients, pemphigus foliaceus is slightly more common than pemphigus vulgaris, and the majority of our patients were females (80%), with a mean age of 43.6 years.

Quality of life (QOL) has been defined as "the individual's perception of their position in life, in the context of the cultural and value systems in which they live and ~~in relation to~~about their goals, expectations, standards, and concerns (11).

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The Dermatology Life Quality Index (DLQI) is the most commonly applied questionnaire to measure health-related quality of life (HRQoL) in dermatology (12). Since its development in the early 1990s, it has been used for over 40 different skin conditions worldwide. (13,14). It is recognized as being useful in various health service settings, including primary care, day-case treatment, outpatient consultations, and inpatient care. (15,16).

Although a previously deadly diagnosis because of skin barrier breakdown leading to infection, the advent of steroids and steroid-sparing agents has allowed pemphigus to be considered a less-fatal, chronic disease (17). In this study, 32% of patients were treated with steroids and cytotoxic drugs, and 28% were treated with steroids only. Therefore, assessing a pemphigus patient's quality of life (QOL) has become an important part of monitoring the disease (3, 17).

Despite the limited literature on QOL in pemphigus (17–26), it is clear that this disease greatly affects patients' emotions, physical health, and social functioning (26, 27). The number of clinical trials in pemphigus has increased immensely over the past decade (28), creating a growing need for objective measurements in both QOL and disease severity to monitor improvement.

The QOL of patients with pemphigus vulgaris was evaluated in one study of 27 German participants using a dermatology-specific instrument. (29). The dermatology-specific Dermatology Life Quality Index (DLQI) score for the study cohort averaged 10.1 ± 6.6 compared with healthy individuals with a mean score of 0.5 ± 1.1, and the authors concluded that a diagnosis of pemphigus vulgaris conferred a large impairment in QOL. The study pointed out that DLQI Patients with mucosal involvement were found to have a higher DLQI, averaging 10.4 ± 7.3, compared with those patients with mucosal sparing, who averaged 9.3 ± 5.1, indicating a poorer QOL. Ghodsi et al. found a similar mean DLQI score of 10.9 ± 6.9 in 61 newly diagnosed untreated PV patients in Tehran using the Persian DLQI (12). Our study showed that the mean DLQI among 25 patients was 11.4. The highest subscores were related to symptoms and feelings (2.8) and daily activities (2.2). Investigators found that the DLQI score was significantly increased in patients with severe disease and mucosal involvement.

These results were consistent with our study, which found that DLQI in pemphigus patients with generalized skin involvement or mucous membrane involvement was more impaired than in those without mucous membrane involvement or localized pemphigus. Also, we found a higher DLQI in patients with pemphigus vulgaris than those with pemphigus foleceous. Disease severity and extent of symptoms likely affect the ability to partake in daily activities and therefore result in lower QOL. A negative correlation between DLQI score and duration of disease was also noted, suggesting increased impairment in the initial stages of the disease. This is further supported by Wysocynska et al. (year??), who reported an average DLQI of 4.0 ± 5.9 in a patient population mainly composed of patients with >5 years of disease (30). Patients likely undergo an adjustment period upon initial diagnosis, which affects QOL scores early on in the disease course. In 2015, a meta-analysis across four studies surrounding QOL in pemphigus patients found a mean DLQI of 12.0 (95% CI 11.1–12.9) with symptoms/feelings and daily activities subscores most consistently affected (31). The mean DLQI scores in our study for different pemphigus treatments received showed patients who were on systemic steroids and cytotoxic drugs had a lower mean score (10.50), followed by systemic steroids (10.71).

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Conclusion: Pemphigus is not just a physical disease and has various aspects. It is responsible for many changes in health related quality of life. in this study ,pemphigushad significant effects on DLQI scores in Libyan patients and lower score scores were observed in treated patients with systemic steroids and cytotoxic drugs.

Ethical approval and consent:The study was approved by Jumhoria teaching hospital ethical committee - Every patient signed an informed written consent for acceptance of participation in the study.

Competing interests:Authors have declared that no competing interests exist.

References:

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Table 1: Descriptive statistics of scale variables in the study

Parameter	Age of study case (in years)	Duration of illness (in months)	Number of symptoms coexisting	Number of complication	Score of life quality index
Mean	43.64	83.12	5.72	4.16	11.44
Std deviation	10.404	96.782	2.208	2.749	5.432
Minimum	30	1	0	0	0
Maximum	72	408	9	10	19

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Table 2 : Type of pemphigus encountered in the study

Clinical type	Frequency	~~Percent~~%
Pemphigus vulgaris	12	48.0
Pemphigus folaiceous	13	52.0
Total	25	100.0

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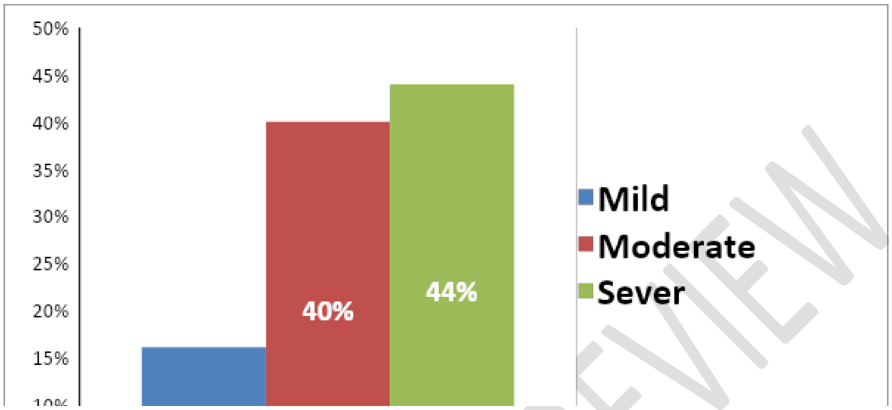

Figure 1 : Severity of illness in study cases

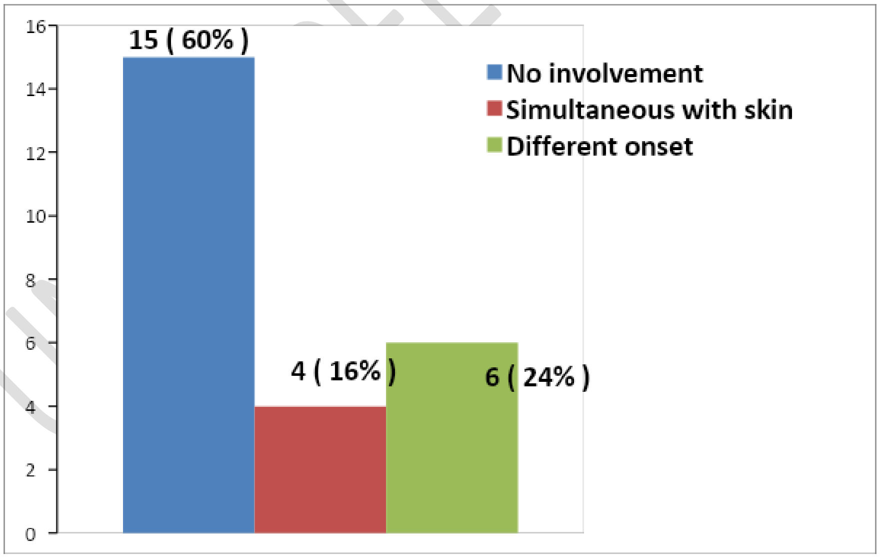

Figure 2 : Mucous membrane involvement in disease in study cases

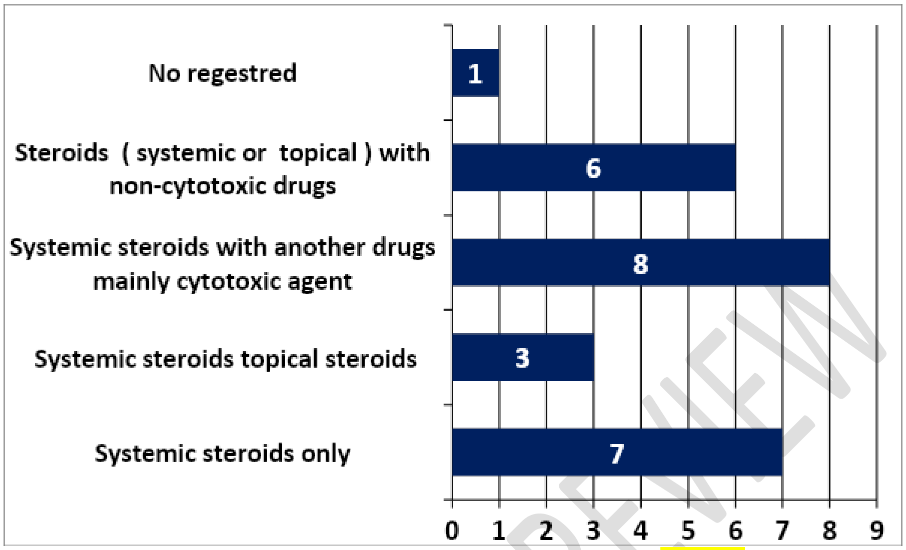

Figure 3 : Treatment used in the study sample (units??)

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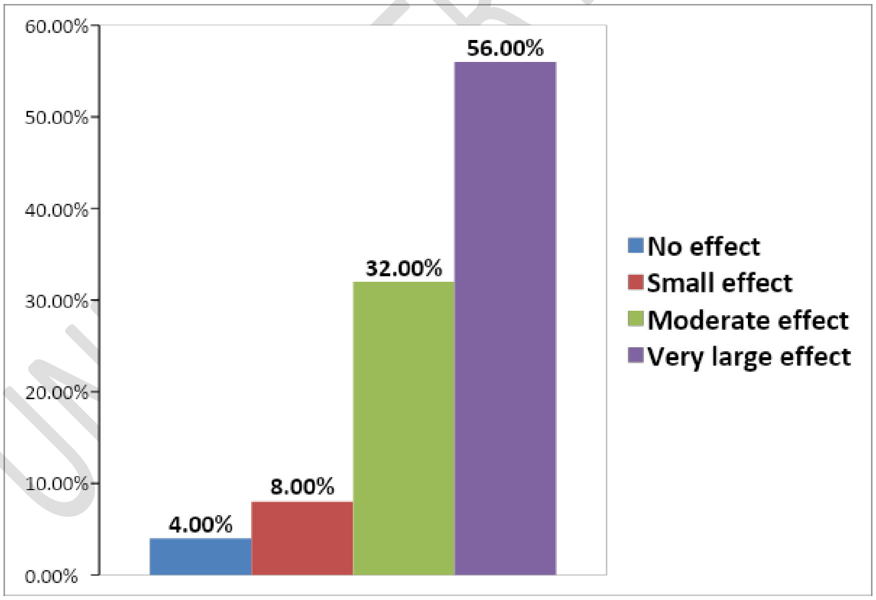

Figure 4 : Distribution of study cases on quality of life index score categories

Table 3 : Descriptive parameters of score of life quality index in categories of pemphigus type

Type of pemphigus	Frequency	Mean	Std. deviation
Pemphigus vulgaris	12	12.67	5.314
Pemphigus foliaceous	13	10.31	5.498

Table 4 : Descriptive parameters of score of life quality index in site of involvement categories

Distribution on the body	Frequency	Mean	Std. deviation	Minimum	Maximum
Generalized skin involvement	9	9.22	4.790	0	15
Generalized skin involvement with mucous membrane	5	16.60	1.517	15	19
Partial skin involvement	5	9.60	5.079	3	17
Partial skin involvement with mucous membrane	5	13.00	6.819	2	19
Total	24	11.63	5.468	0	19

Table 5 : Descriptive parameters of score of life quality index in major treatment categories

Type of treatment	Frequency	Mean	Std. deviation
Systemic steroid	7	10.71	4.499
Systemic + topical Steroids	3	12.00	5.245
Systemic steroids + cytotoxic agent	8	10.50	7.231
Systemic or topical steroids + non-cytotoxic agent	6	14.17	3.371
Total	24	11.67	5.427