Original Research

Illness perception of patients with pemphigus vulgaris ☆

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ABSTRACT

Objective: Little is known about illness perception in patients with pemphigus vulgaris (PV). We designed a cross-sectional study to clarify the beliefs about PV.
Methods: A total of 100 patients with PV (45 men, 55 women) completed the Illness Perception Questionnaire-Revised to assess beliefs about seven aspects of illness perception, including chronicity, recurrence, consequences, self and medicine role in controlling illness, coherence, and emotional representation. The relationship between illness perception and clinical and demographic variables was evaluated.
Results: Patients viewed PV as a chronic and cyclical disease with important impression on their life and emotions. Patients had a good understanding of the disease and supposed an acceptable role for themselves and medical treatment. Interestingly, the clinical subtype and severity of the disease did not influence any aspect of illness perception, but some differences on the basis of demographic data were demonstrated.
Conclusion: Our patients had a relatively good understanding of their illness and a correct perception about chronicity and the cyclical identity of illness. The patients believed that their life and emotions had been strongly influenced by the disease but were hopeful for a cure. Because correction of misconceptions about a disease may improve treatment outcomes, an assessment of patients' illness perception may be useful to try and modify perception.

Introduction

Pemphigus vulgaris (PV) is a rare, chronic, and debilitating bullous disease that involves the skin and mucosa. Distressing symptoms that come with visible lesions can negatively affect patients' mental health (Caccavale and Lo Schiavo, 2014; Wohl et al., 2015) and significantly impair the quality of life (QoL) of affected patients (Ghodsi et al., 2012; Sebaratnam et al., 2012; Terrab et al., 2005). However, the clinical severity of the disease is not a good indicator for impaired QoL and the psychological wellbeing of patients with chronic medical conditions. Fortune et al. (2002) showed that cognitive psychological factors have a more important role than medical variables such as severity, chronicity, and extent of disease in explaining variance in stress, distress, and disability in patients with psoriasis.

In addition, Leventhal et al. (1984) explained that patients can process information about their illness and construct a cognitive model of the illness that is an important factor in functioning and coping behaviors. Thus, the term *illness perception* has been defined as a patient's personal belief about his or her disease. Illness perception may affect psychosocial responses to the disease and adherence to therapy in PV. Studies have shown this effect in similar conditions such as psoriasis (Richards et al., 2006).

To the best of our knowledge, illness perception has not been studied thoroughly in PV. Thus, the aim of this study was to evaluate illness perception in patients affected with PV and its relationship with demographic and clinical factors.

Methods

Participants

A total of 100 patients with confirmed PV, age 18 to 60 years, participated in this cross-sectional study. The study was performed at Razi Dermatology Hospital in Tehran, Iran, from July 2016 to July 2017. Written informed consent was obtained from each patient.

The study was reviewed and approved by the ethics committee of Tehran University of Medical Sciences (8911112105).

Instruments

Patients completed a questionnaire that collected demographic and clinical data. The physician used a simple scoring system of 0 to 3 (0 = quiescent, 1 = mild, 2 = moderate, and 3 = severe) to assess disease severity for the skin and oral cavity, separately (Weinman et al., 1996). The oral cavity was scored as 0 = quiescent, 1 = minor activity (up to three erosions), 2 = moderate activity (>3 but <10 erosions, or generalized desquamative gingivitis), and 3 = severe (>10 discrete erosions, or extensive confluent erosions, or generalized desquamative gingivitis with discrete erosions at other oral sites). The skin was scored as 0 = quiescent, 1 = minor activity (<5 discreet lesions), 2 = moderate activity (>5 but <20 discreet lesions), and 3 = severe (>20 discreet lesions, or extensive confluent areas of eroded skin).

The validated Persian version of the Illness Perception Questionnaire-Revised (IP-R) was used to evaluate illness perception. The questionnaire consists of the following subscales (Aflakseir et al., 2012; Moss-Morris et al., 2002): 1) Timeline-acute/chronic course (6 items) to evaluate perceptions about how long the disease will last, with a higher score corresponding to a stronger perception of disease as a long-lasting situation; 2) timeline-cyclical identity (4 items) to assess the patient's awareness of flare-up periods and remission of disease, with a higher score indicating a greater belief in cyclical identity of the disease; 3) consequences (6 items) to estimate the expected effects of the illness on a patient's life, with a higher score indicating higher negative consequences; 4) personal control (6 items) concerning patients' beliefs about their personal role in the control of their illness—patients with this belief have a higher score in this component; 5) treatment control (5 items) with regard to the patient's beliefs about the efficacy of the medical interventions in recovery—patients with a greater score have a stronger perception of treatment control over their illness; 6) illness coherence (5 items) to evaluate the patients' understanding of their disease, where a higher grade means a better understanding of disease identity; and 7) emotional representation (6 items) to weigh the patients' perceptions about the impact of their illness on their emotions, where patients with higher scores experience greater emotional distress.

The causal component consists of 18 items that evaluate personal ideas about the cause of disease, including stress or worry, mental attitude, family problems, overwork, emotional state, personality, hereditary factors, eating habits, poor medical care in the past, personal behavior, ageing, smoking, alcohol, a germ or virus, pollution in the environment, altered immunity, chance or bad luck, and accident or injury. A five-point Likert-type scale with scores from 1 (strongly agree) to 5 (strongly disagree) was used to answer each item.

Statistical analysis

Mean and standard deviation as well as frequency were reported for continuous and categorical variables, respectively. To compare the means of scores among discontinuous independent variables, the independent *t* and one-way analysis of variance tests were used. Pearson's correlation coefficient was estimated to evaluate the correlation between continuous variables. A *p*-value of < .05 was considered significant. Data were analyzed using SPSS software, version 22. A nonparametric equivalent of parametric tests was performed to confirm the results of the parametric test.

Results

A total of 100 patients with PV (45 men, 55 women), with an age range of 18 to 59 years and a median age of 24 years, were enrolled in

Table 1
Baseline characteristics of patients (N = 100)

Sex	n (%)
Male	45
Female	55
Marital status	
Single	15
Married	84
Divorced	1
Occupation	
Employed	45
Unemployed	55
Education	
Elementary school or less	30
High school level	41
University degree	29
Smoking status	
Yes	14
No	86
Cutaneous lesions severity	
Mild	55
Moderate	14
Severe	14
Not available	17
Mucosal lesions severity	
Mild	76
Moderate	8
Severe	3
Not available	13
Disease subtype	
Mucocutaneous	70
Mucosal	17
Cutaneous	13

the study. The median duration of the disease was 51 to 75 months with a range of 1 to 300 months (Table 1).

Table 2 shows the aggregated scores for the IPQ-R in patients with PV. Higher scores mean stronger perceptions of acute/chronic and cyclical illness identity, more negative consequences, greater emotional distress, stronger perceptions of personal and treatment control over the illness, and a higher level of understanding of disease identity.

Beliefs about chronicity

Most patients experienced their illness as a long-lasting condition. Married patients had stronger beliefs about chronicity compared with patients who were single (18.5 ± 5.3 vs. 15.0 ± 5.7; p = .021). Furthermore, a positive correlation was detected between the duration of the disease and beliefs about chronicity (r = .438; p < .001).

Beliefs about recurrence

Patients mostly perceived their illness as a timely variable condition. We observed that women had a stronger trend toward perceptions about cyclical identity of disease compared with men (14.3 ± 2.9 vs. 13.0 ± 2.8; p = .02).

Table 2
Aggregated scores of Revised Illness Perception Questionnaire in study sample

Questionnaire dimension	Score range	Mean ± standard deviation
Timeline: Acute/chronic	6-30	18.1 ± 5.6
Timeline: Cyclical	4-20	13.7 ± 2.9
Consequences	6-30	18.6 ± 4.9
Personal control	6-30	23.6 ± 4.1
Treatment control	5-25	20.1 ± .2
Illness coherence	5-25	15.0 ± 5.0
Emotional representations	6-30	20.9 ± 5.9

Beliefs about consequence

Patients perceived that their illness has a strong negative effect on their lives. Perceptions of negative consequences become stronger with increasing disease duration (r = .25; *p* = .01).

Beliefs about personal control

Patients showed a more effective role for themselves in control of their illness, especially with a higher educational level (r = .23; *p* = .02).

Beliefs about medication role

We observed a strong belief by patients in the role of medicine to control their illness. There was a weak negative correlation between the duration of the disease and the belief in the ability of treatment to control the disease (r = -.21; *p* = .03).

Beliefs about illness coherence

Patients had a relatively good understanding of their illness. Furthermore, there was a difference based on marital status (15.4 ± 4.9 vs. 12.3 ± 4.7 in married and single patients, respectively; *p* = .024).

Beliefs about emotions

We demonstrated a strong negative impact of disease on patient emotions. Additionally, women had higher scores than men (22.2 ± 5.8 vs. 19.3 ± 5.7; *p* = .01).

Causal component

Patients assumed that the cause of their disease was stress (89%), family problems (72%), altered immunity (64%), emotional conditions (56%), personal behavior (54%), mental attitude (50%), environmental pollution (47%), dietary habits (40%), overwork (39%), personality (34%), chance (28%), poor medical care in the past (26%), a germ or virus (17%), smoking (12%), hereditary factors (11%), ageing (8%), and alcohol (4%). An analysis of the causal component based on demographic data indicated that men identified stress (*p* = .041) and smoking (*p* = .001) as the leading cause of their illness more often than women. There was no significant difference in causal ideas based on the subtype of the disease, but the severity of the disease indicated some differences with increasing cutaneous and mucosal involvement.

An analysis based on cutaneous involvement demonstrated a positive correlation for bio-organism and environmental pollution items, but a negative correlation for emotional statement. Thus, bio-organism (r = .21; *p* = .03) and environmental pollution (r = .28; *p* = .004) were considered more responsible and emotional statement (r = -.2; *p* = .03) as less responsible for disease causes by patients with severe cutaneous involvement. Smokers considered smoking as the cause of their disease more often than nonsmoking patients (*p* < .001).

Higher-educated patients considered hereditary factors, environmental pollution, overworking, and altered immunity as the leading causes of their illness more often than patients with less education (*p* = .009; .004; .003; and .02, respectively). Environmental pollution was considered more as a cause of disease by employed patients (*p* = .001).

Discussion

Our findings revealed that patients with PV correctly experienced their illness as a chronic and periodic condition. Married patients and those with long-lasting disease perceived their disease as more chronic in nature. Eder et al. (2016) reported that women with cutaneous T-cell lymphoma assumed their illness to be more chronic than did men, but we did not detect any sex-related differences in the chronicity item (Tabolli et al., 2014). However, women assumed their illness to be more periodical. Patients perceived their illness as such because of the strong effect on their lives, which could be explained by the chronic course of the disease, visible lesions, distressing symptoms, and prolonged immunosuppressant therapy.

Our findings are in accordance with those of Ghodsi et al. (2012), who showed that the QoL of patients with PV is significantly impaired, and with Tabolli et al. (2014), who indicated that QoL remains low even during quiescent periods of the disease. Two internationally validated scores that measure patient QoL change due to disease and treatment that are specific to patients with autoimmune bullous disease are available (Sebaratnam et al., 2013; Tjokrowidjaja et al., 2013). The scores were translated recently into the Farsi but have not been published yet; therefore, we could not use them for this article. We suggest an evaluation of changes in patients' QoL due to alternative illness perceptions in future studies.

Patient perceptions about the consequences of their disease became stronger with increasing durations of their disease, but interestingly there was no significant difference based on the severity of the disease. Firooz et al. (2005) demonstrated the same results in a survey of patients with alopecia areata, which revealed that the chronicity of disease affects patients' life more than the severity of the disease.

Our patients were hopeful about a cure, but their concepts of treatment outcomes became weaker with increased duration of disease. Although PV is not curable, the disease can be controlled with appropriate treatment modalities, especially new biologic agents, so our patients should be educated about the efficacy of various medications (Joly, 2017). Furthermore, patients assumed an acceptable role for themselves on the control of the disease, and this concept became stronger with increased levels of education; thus, education gives patients a greater sense of capability and power.

Despite the uncommon nature of PV, patients reported a relatively acceptable understanding of the illness. However, Eder et al. (2016) reported a poor understanding of the disease in patients with cutaneous T-cell lymphoma, possibly due to the low incidence of the disease in society and its unknown identity for the general population. Interestingly, understanding of the disease was not influenced by level of education, but married patients had a better understanding compared with patients who were single, which may be due to the advocacy of their partners and their role in asking the doctor more questions or performing research on their partner's behalf.

PV had strong negative impacts on patients' emotions. Although harboring negative perceptions about the consequences of an illness is not exactly equal to psychiatric disorders such as depression and anxiety, the likelihood of these situations occurring increases (Fortune et al., 1998; Wohl et al., 2015). We demonstrated that the emotions of women were influenced by their illness more than those of men. Stewart et al. (1997) explained that women have more sensitivity to physical concerns because of their acceptance of different sex-related roles in society.

Interestingly, the severity of PV did not affect patients' illness perception. This parallels the findings by Fortune et al. (1998), who did not observe any association between the clinical severity of psoriasis and illness perception.

Patients in this study assumed that stress was the most common causal factor, similar to reports of prior studies about illness

perception on skin disorders such as vitiligo (Firooz et al., 2004; Topal et al., 2016), psoriasis (Fortune et al., 1998), and alopecia areata (Firooz et al., 2005). Patients with severe cutaneous involvement pinpointed external agents such as bio-organisms and environmental pollution as the leading causes of their illness, but this did not hold true for patients with severe mucosal involvement. This finding could be explained by the fact that the skin is the most outer human body organ and has the most interaction with the environment; therefore, patients may assign environmental factors as the causal agent of the disease. Interestingly, patients assumed that their emotional statement was the cause of severe mucosal lesions.

Among the list of causal factors that patients were asked to score, altered immunity was the most reasonable real cause, and highly educated patients believed altered immunity to be a causal factor more often than lower educated patients. Topal et al. (2016) demonstrated the same results in a sample of 100 patients with vitiligo. An unclear understanding of disease identity, consequences, and treatment outcomes leads to anxiety and depression and consequently can worsen the disease (Bystryn and Rudolph, 2005). Furthermore, a correction of misconceptions about the disease may improve treatment outcomes (Chen et al., 2018; Etain et al., 2018; Keogh et al., 2011; Lee et al., 2018).

A limitation of this study was that no validated score of severity was used. More studies are needed to focus on clearing illness perception in patients with PV and designing follow-up studies to assess treatment adherence changes with psychological interventions.

Conclusion

Our patients had a relatively good understanding of their illness and correct perception about chronicity and the cyclical identity of the disease. They believed that their life and emotions are strongly influenced by the disease but were hopeful for a cure. Because correction of misconceptions about the disease may improve treatment outcomes, an assessment of patients' illness perception could be useful to try and modify misconceptions.

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