

**A Retrospective Clinical Analysis of  
Korean Patients with Bullous  
Pemphigoid**

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# **A Retrospective Clinical Analysis of Korean Patients with Bullous Pemphigoid**

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<ABSTRACT>

**A Retrospective Clinical Analysis of Korean Patients  
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**Background :** Bullous pemphigoid (BP) is a potentially fatal autoimmune bullous disease which commonly occurs in elderly patients. The mortality rate and prognosis of BP have been controversial in the populations of different countries. However, there are few studies to date that have investigated BP in Korea.

**Objective :** To evaluate the mortality rates of Korean patients with BP and compare them with age-matched general population in Korea, and to identify prognostic factors associated with overall survival.

**Methods :** We conducted a retrospective analysis of 168 patients diagnosed with BP between 1993 and 2013 in Gangnam Severance Hospital. The mortality rates of patients with BP were compared with that of age-matched general Korean population.

**Results :** The 1-, 2-, and 5-year mortality rates of BP patients were 19.46%,

29.13%, and 58.03%, respectively. The standardized mortality ratio (SMR) varied from 2.43 to 9.56, depending on the age group. Sex, age at the time of diagnosis, stroke, cancer, and Karnofsky performance status score were associated with decreased overall survival on univariate Cox regression analysis. However, only age at the time of diagnosis and stroke were associated with increased mortality on multivariate analysis.

**Conclusions :** The mortality rates of patients with BP are significantly higher than those of the general population in Korea. Advanced age at the time of diagnosis and stroke are strong prognostic factors associated with increased mortality.

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Key words : Bullous pemphigoid, Korea, Mortality, Prognosis, Autoimmune bullous disease



# **A Retrospective Clinical Analysis of Korean Patients with Bullous Pemphigoid**

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## **I. INTRODUCTION**

Bullous pemphigoid (BP) is the most common autoimmune blistering disease in the United States and Europe.<sup>1</sup> The disease is characterized by pruritic, tense bullae and urticarial plaques in the elderly. The autoantigens of BP are BP180 and BP230 which are components of hemidesmosome. Several studies examining the mortality rates and prognosis of BP have been conducted in American and European patients,<sup>2-5</sup> and have produced conflicting results. In these studies, first year mortality rates reportedly range from 6% to 41%.<sup>2-20</sup> Many European studies have reported a poor prognosis of BP, while studies conducted in the United States have described a better outcome. However, the reason for these large discrepancies of prognosis is unclear. There is a dearth of studies regarding the prognosis of BP in Asian population, although BP is also one of the most common autoimmune

blistering diseases in Asian countries. The objective of this study was to determine the mortality rates of patients with BP in a representative cohort from a single, large clinical referral center in Korea, and to identify clinical and therapeutic factors that exhibit potential prognostic value.

## **II. MATERIALS AND METHODS**

### **1. Study populations**

The medical records of all patients with BP diagnosed at Gangnam Severance Hospital in Seoul, Korea, between January 1993 and March 2013 were retrospectively reviewed. Skin biopsy specimens and sera were evaluated in patients with clinically suspected BP for diagnostic confirmation. The following criteria were used for the diagnosis of BP. (1) clinical suspicion of BP due to characteristic features; (2) histopathology consistent with BP (subepidermal blisters with infiltration of inflammatory cells, especially eosinophils); (3) direct immunofluorescence showing linear C3 or/and IgG along the basement membrane zone; (4) circulating IgG autoantibodies binding to the epidermal side of salt-split skin, as detected by indirect immunofluorescence study; and (5) IgG autoantibodies against BP180 antigen detected by enzyme-linked immunoabsorbent assay (ELISA) or by Western blot analysis. Diagnosis of BP was confirmed when the patient fulfilled the clinical (1), histological (2), and immunological (3) criteria, as well as at least one of the serological (4, 5) criteria. A total 168 BP patients who satisfied the diagnostic criteria were included in this study. After an initial diagnosis, patients treated with corticosteroid with or without corticosteroid sparing agents, such as dapsone, doxycycline and nicotinamide. Some severe patients were treated with high dose intravenous immunoglobulin therapy.

Demographic data were collected along with sex, age at the time of diagnosis, diagnosis delay, date of death, treatment modalities, severity of disease, disease-related symptoms, medications, performance status, and comorbidities. We defined diagnosis delay as a time period of >6 months between the onset of symptoms and a confirmation of diagnosis. In order to measure the patient's overall condition and performance status, we used the Karnofsky performance status scale, with higher scores indicating better performance status.<sup>21</sup> For statistical analysis, comorbidities were grouped as follows: heart disease (coronary artery disease, cardiac arrhythmia, congestive heart failure), diabetes, dementia, hypertension, hypothyroidism, stroke, other neurologic disease (seizures, psychosis, depression, Parkinson's disease), malignancy, chronic renal failure, and chronic lung disease. BP disease severity was graded based on the percentage of body surface area involvement as follows: mild <10%, moderate 10% to 30%, and severe >30%.<sup>5</sup> Patients who had more than 6 months of follow-up data were included in this study to avoid bias.

## **2. Statistical analysis**

All analyses were conducted with the aid of statistical software (SAS, Version 9.2, SAS Institute Inc., Cary, NC), using methods appropriate for time-to-event data. Survival time in days was measured from the date of diagnosis of BP to either the date of death or the end of the analysis. Data for patients alive at the time of final analysis were considered right-censored. Cumulative mortality was estimated using the Kaplan-Meier method. A Cox regression model was used to determine whether there was a significant difference in overall survival among patients with BP based on baseline characteristics. Baseline data included sociodemographic information, disease severity, performance status, treatment modalities and presence of comorbidities. Hazard ratios (HR) were estimated with 95% confidence intervals (CI).

To determine expected mortality in a general population of Korean patients based on age, life tables from the Korean Statistical Information Service were used. This publication presents age-specific death rates for a given year (2011). These rates were applied to the observed person-years of follow-up. The ratio of the observed to expected death rates, or the standardized mortality ratio (SMR), was calculated for patients in 10-year age intervals; an SMR of 1 would indicate that the number of observed and expected deaths was the same. We divided our BP cohort into groups based on age and applied the SMR to each subgroup. The 95% CI for the SMR was determined using exact methods,

which assume the observed number of deaths had a Poisson distribution. *P*-values  $<0.05$  were considered to be statistically significant.

### III. RESULTS

#### 1. Demographic data and clinical features of patients

A total 168 patients with BP who met the diagnostic criteria were identified. A total of 84 patients (50%) were female. The median age at the time of diagnosis was 73 years, and the mean age was 69.15 years (SD = 15.70). The age range at the time of diagnosis 6-99 years, with the majority of patients (96%) being older than 40 years of age. Disease severity was classified as mild, moderate or severe. There were 54 (32.14%) patients with mild BP and 45 (26.79%) patients with severe BP (Table 1).

**Table 1.** Baseline characteristics of study population

	Total N = 168
<b>Mean age at diagnosis (yr)</b>	69.15 ± 15.70
<b>Sex</b>	
<b>Male</b>	84 (50%)
<b>Female</b>	84 (50%)
<b>Disease severity</b>	
<b>Mild</b>	54 (32.14%)
<b>Moderate</b>	69 (41.07%)
<b>Severe</b>	45 (26.79%)

\*Data are presented as mean±SD or N (%)

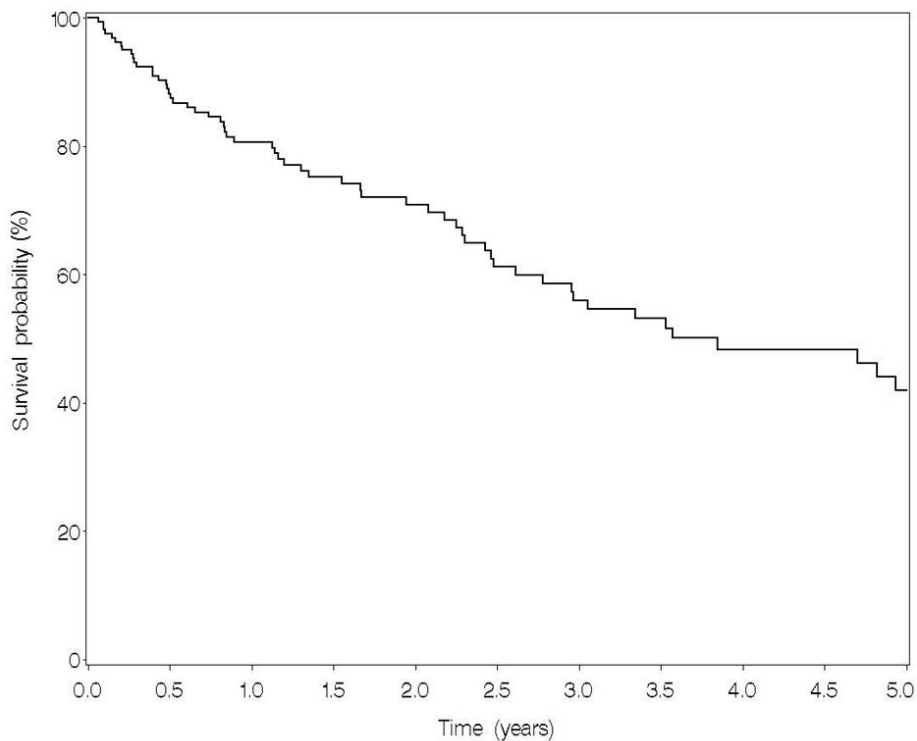
The median length of follow-up for those alive at the end of the analysis was 2.45 years, and for all individuals included in our analysis the median length of follow-up was 2.19 years.



## 2. Mortality and prognostic factors

### A. Mortality

Of the 168 patients included in this study, 103 (61.31%) were still alive at the time of final analysis. The 1-year mortality rate for our BP cohort was 0.19 (95% CI = 0.14, 0.27), the 2-year mortality rate was 0.29 (95% CI = 0.22, 0.38), and the 5-year mortality rate was 0.58 (95% CI = 0.47, 0.69). The survival curve is shown in Figure 1.



**Figure 1.** Kaplan-Meier curve of overall survival of patients with bullous pemphigoid.

## B. Standardized mortality ratio (SMR)

The results of the SMR analysis are given in Table 2.

**Table 2.** Standardized mortality ratios of observed deaths in patients with bullous pemphigoid versus expected deaths in the 2011 general Korean population.

Age group (yr)	No. patients	Expected rate	No. mortality	Observed rate	SMR
≤60	38	0.0109	1	0.0263	2.4254
60-70	34	0.0092	3	0.0882	9.5596
70-80	51	0.0262	10	0.1961	7.4736
≥80	45	0.0925	14	0.3111	3.3651

The SMR varied from 2.43 - 9.56 depending on age. Based on these data, the mortality of patients in our BP cohort was higher than what would be expected in age-matched individuals in the general population. The difference between expected and observed mortality was particularly high for patients with BP between 60-80 years of age.

### **C. Prognostic factors**

To identify the prognostic factors associated with mortality, we used Cox logistic regression for 1-year mortality. Among the variables studied, only age at the time of diagnosis (HR, 1.07; 95% CI, 1.02-1.13,  $P=0.0051$ ), diabetes (HR, 4.11; 95% CI, 1.69-9.97,  $P=0.001$ ) and diagnosis delay (HR, 0.1; 95% CI, 0.01-0.78,  $P=0.02$ ) were independent predictors of 1-year mortality on multivariate analysis. However using a mean age of 70 years as a cut-off, older age was not statistically significant on multivariate analysis for 1 year mortality (HR, 2.961; 95% CI, 0.958-9.154,  $P=0.0595$ ) (Table 3).

**Table 3.** Results of the univariate and multivariate analyses for 1-year mortality in patients with bullous pemphigoid

	No. of patients (%)	No. of patients with lethal outcome(%)	Univariate Hazard Ratio	<i>P</i> , Univariate	Multivariate Hazard Ratio	<i>P</i> , Multivariate
<b>Sex</b>						
Female	84 (50%)	13 (15.48%)	1.212	0.6124		
Male	84 (50%)	15 (17.86%)				
<b>Age (yr)</b>						
<70	72 (42.86%)	4 (5.56%)	5.359	0.0019	2.961	0.0595
≥70	96 (57.14%)	24 (25%)				
<b>Disease severity</b>						
Mild	54 (32.14%)	6 (11.11%)	1.13	0.6711		
Moderate to severe	114 (67.86%)	22 (19.30%)				
<b>Diagnosis delay</b>						
Yes	45 (26.79%)	1 (2.22%)	0.088	0.0171	0.104	0.0275
No	123 (73.21%)	27 (21.95%)				
<b>Diabetes</b>						
Yes	57 (34.76%)	16 (28.07%)	4.291	0.0008	4.110	0.0018
No	107 (65.24%)	8 (7.48%)				
Missing	4					

Heart disease						
Yes	11 (6.67%)	5 (45.45%)	4.56	0.0025	0.457	0.089
No	154 (93.33%)	20 (12.99%)				
Missing	3					
Hypertension						
Yes	53 (32.12%)	11 (20.75%)	1.661	0.208		
No	112 (67.88%)	14 (12.5%)				
Missing	3					
Dementia						
Yes	20 (12.12%)	3 (15%)	1.006	0.9926		
No	145(87.88%)	22 (15.17%)				
Missing	3					
Stroke						
Yes	28 (16.97%)	8 (28.57%)	2.758	0.018	2.011	0.1657
No	137 (83.03%)	17 (12.41%)				
Missing	3					
Neurologic disease						
Yes	13 (7.88%)	1 (7.69%)	0.455	0.4409		
No	152 (92.12%)	24 (15.79%)				
Missing	3					
Cancer						
Yes	9 (5.45%)	3 (33.33%)	2.471	0.1419		
No	156 (94.55%)	22 (14.10%)				
Missing	3					

CRF						
Yes	10 (6.06%)	2 (20%)	1.097	0.9006		
No	155 (93.94%)	23 (14.84%)				
Missing	3					
Lung disease						
Yes	13 (7.88%)	1 (7.69%)	0.444	0.4268		
No	152 (92.12%)	24 (15.79%)				
Missing	3					
Admission						
Yes	45 (26.79%)	11 (24.44%)	1.699	0.1709		
No	123 (73.21%)	17 (13.82%)				
IVIG						
Yes	10 (5.95%)	2 (20%)	1.121	0.8759		
No	158 (94.05%)	26 (16.46%)				
Combination Therapy						
Yes	128 (76.19%)	26 (20.31%)	3.985	0.0596		
No	40 (23.81%)	2 (5%)				
Karnofsky scale			0.981	0.0206	1.014	0.5017
Admission duration			1.018	0.0944		
Initial steroid dose			1.001	0.9657		

Factors related to decreased overall survival were also evaluated using univariate and multivariate Cox logistic regression (Table 4).



**Table 4.** Results of the univariate and multivariate analyses for lethal outcome in patients with bullous pemphigoid

	No. of patients (%)	No. of patients with lethal outcome(%)	Univariate Hazard Ratio	<i>P</i> , Univariate	Multivariate Hazard Ratio	<i>P</i> , Multivariate
Sex						
Male	84 (50%)	41 (48.81%)	1.905	0.0136	1.702	0.0609
Female	84 (50%)	24 (28.57%)				
Age (yr)						
≥70	96 (57.14%)	52 (54.17%)	7.475	<0.0001	5.09	<0.0001
<70	72 (42.86%)	13 (18.06%)				
Disease severity						
Mild	54 (37%)	18 (33.33%)				
Moderate to severe	114 (73%)	47 (41.23%)	1.765	0.2176		
Diagnosis delay						
Yes	45 (26.79%)	15 (33.33%)				
No	123 (73.21%)	50 (40.65%)	0.636	0.126		
Diabetes						
Yes	57 (34.76%)	21 (36.84%)				
No	107 (65.24%)	40 (37.38%)	1.247	0.4148		
Missing	4					

Heart disease						
Yes	11 (6.67%)	6 (54.55%)	1.559	0.3057		
No	154 (93.33%)	56 (36.36%)				
Missing	3					
Hypertension						
Yes	53 (32.12%)	24 (45.28%)	1.334	0.2702		
No	112 (67.88%)	38 (33.93%)				
Missing	3					
Dementia						
Yes	20 (12.12%)	10 (50%)	1.848	0.0805		
No	145 (87.88%)	52 (35.86%)				
Missing	3					
Stroke						
Yes	28 (16.97%)	15 (53.57%)	2.297	0.0059	2.113	0.0321
No	137 (83.03%)	47 (34.31%)				
Missing	3					
Neurologic disease						
Yes	13 (7.88%)	5 (38.46%)	1.161	0.7506		
No	152 (92.12%)	57 (37.50%)				
Missing	3					
Cancer						
Yes	9 (5.45%)	8 (88.89%)	3.797	0.0005	2.039	0.0809
No	156 (94.55%)	54 (34.62%)				
Missing	3					

CRF						
Yes	10 (6.06%)	5 (50%)	1.108	0.8261		
No	155 (93.94%)	57 (36.77%)				
Missing	3					
Lung disease						
Yes	13 (7.88%)	5 (38.46%)	0.835	0.6996		
No	152 (92.12%)	57 (37.50%)				
Missing	3					
Admission						
Yes	45 (26.79%)	22 (48.89%)	1.324	0.2865		
No	123 (73.21%)	43 (34.96%)				
IVIG						
Yes	10 (5.95%)	5 (50%)	1.43	0.4443		
No	158 (94.05%)	60 (37.97%)				
Combination Therapy						
Yes	128 (76.19%)	54 (42.19%)	1.499	0.2229		
No	40 (23.81%)	11 (27.5%)				
Karnofsky scale			0.976	0.996	0.6281	
Admission duration			1.01	0.3394		
Initial steroid dose			0.999	0.9294		

On the univariate cox logistic regression analysis, sex and age at the time of diagnosis were significantly associated with overall survival. Males had a significantly increased risk of mortality on univariate analysis, but male sex was not independently predictive of mortality on multivariate analysis. However, age at the time of diagnosis appeared to independently predict overall survival on multivariate analysis (HR, 2.2; 95% CI, 1.2-4.3,  $P<0.0001$ ). Using a mean age of 70 years as a cut-off, older age remained a significant risk factor for death (HR, 5.09; 95% CI, 2.27-10.66,  $P<0.0001$ ). Among the comorbidities studied, only stroke and cancer showed significant differences in overall survival. However, only stroke had an increased risk of mortality on multivariate analysis (HR, 2.1; 95% CI, 1.07–4.19,  $P<0.05$ ). The other comorbidities did not significantly impact overall survival on univariate analysis.

A higher Karnofsky performance status score was strongly associated with survival on univariate analysis, but not multivariate analysis. However, disease severity, hospitalization, and treatment modality did not appear to be associated with overall survival.

## IV. DISCUSSION

Regarding the mortality of BP patients, conflicting results have been reported from different countries. In our study of 168 patients with BP, the 1-year mortality rate was 19%, the 2-year mortality rate was 29%, and the 5-year mortality rate was 58%. These rates are lower than those reported in European studies. However, the mean age of the patients in our study was 69.15 years (SD = 15.70), which was approximately 10 years younger than that of previous European studies. Considering the younger age of our patient population, our results are consistent with previous data from Switzerland.<sup>2</sup> Several European studies reported higher mortality rates than US studies,<sup>22</sup> however, a recent large US cohort study reported higher mortality rates that were closer to those demonstrated in previous European data.<sup>5</sup> Although there are few studies in Asian countries, two recent Chinese cohort studies reported 1-year mortality rates of patients with BP.<sup>19,20</sup> One study<sup>20</sup> showed a slightly higher mortality rate than our study (23.4%). Another study from China<sup>19</sup> reported a significantly lower mortality rate (12.9%). However the latter is difficult to compare with results from other studies, because this included only hospitalized patients. A summary of 1-year mortality rates in BP cohorts from recently published studies is shown in Table 5.

**Table 5.** One-year mortality rates in bullous pemphigoid cohorts from recently published studies

Authors	No. of Patients	Mean age	1-year Mortality (%)	Country	Type of study	Year
Parker <i>et al.</i> <sup>5</sup>	223	75.2	23	US	Retrospective	2008
Cortes <i>et al.</i> <sup>2</sup>	115	76.4	20.9	Switzerland	Prospective	2011
Cortes <i>et al.</i> <sup>18</sup>	60	79.5	26.7	Switzerland	Retrospective	2012
Gual <i>et al.</i> <sup>3</sup>	101	77.8	12.9	Spain	Retrospective	2012
Joly <i>et al.</i> <sup>4</sup>	312	82.1	38	France	Prospective	2012
Li <i>et al.</i> <sup>19</sup>	140	67	12.9	China	Retrospective	2013
Zhang <i>et al.</i> <sup>20</sup>	94	71	23.4	China	Retrospective	2013
Present study	168	69.2	19	Korea	Retrospective	2013

We were unable to identify disease-specific mortality of bullous pemphigoid in this study. As an alternative, we compared the mortality rates of patients with BP with those of an age-matched general population. We found that patients with BP had greatly increased rates of mortality. For all age groups, the rate of observed deaths was higher than that expected in the general population, particularly in BP patients in 60-80 years of age. Several cohort studies have reported that patients with BP have increased mortality compared with the age-adjusted general population,<sup>2,4,19</sup> with the exception of one study conducted in the US<sup>5</sup> which reported that the mortality rate of patients with BP was not higher than that of the general population. Because the US study was a large retrospective cohort study, the study might have a selection bias. Two other large prospective cohort studies,<sup>2,4</sup> on the other hand, reported a SMR that was significantly higher than the general population. Our data support the results reported in the latter studies.

Previous studies have reported prognostic factors for mortality in patients with BP.<sup>2,3,5,16-20</sup> However, some of these data are based on 1-year mortality rates, while others are based on overall mortality. Therefore, we analyzed prognostic factors associated with both 1-year survival and overall survival. The mean age in our study (69.15 years) was lower than that of other previously published studies. However, similar to other reports, advanced age at the time of diagnosis was a significant risk factor for mortality in our cohort. This was consistently true for both 1-year survival and overall survival. And

using a mean age of 70 years as a cut-off, older age remained a significant poor prognostic factor for overall survival. Because the mean ages of the patients in previous studies were older than ours, they proposed that an age of 80 years or older was a poor prognostic factor for BP patients.<sup>2,3,18</sup> Our study, on the other hand, showed that age could be a risk factor for poor prognosis in a younger patients as well.

Interestingly, other groups have reported diabetes to be a poor prognostic factor for overall survival,<sup>5,18</sup> however, a history of diabetes showed an increased risk of 1-year mortality, but not overall mortality in our study. Diagnosis delay was also an independent predictive factor of 1-year mortality, but not overall mortality. This might be due to the fact that patients who were diagnosed 6 months after the onset of symptoms might have had a relatively less severe disease. However, we could not find the statistically significant relation between disease severity and mortality.

Stroke, which commonly occurs in advanced old age, was shown to be a poor prognostic factor of overall survival in our patients. Many studies have previously reported neurologic disease to be a poor prognostic factor as well.<sup>2,5,18-20</sup> However, some of these studies have classified dementia, cerebrovascular disease and psychosis collectively as neurologic disease. Thus, in this study we analyzed each disease individually, and showed that only stroke was significantly associated with overall survival.

There are some limitations to our study. First, it was retrospective in nature



and extended over approximately 20 years. Thus, improvement of treatment may have significantly altered mortality rates of patients with BP during this time period. Second, even though our hospital is the largest center for patients with bullous pemphigoid in Korea, this study is limited by the fact that it was based at a single institution.

## **V. CONCLUSION**

In conclusion, the mortality rate of patients with BP is significantly higher than that of an age-matched general population in Korea. Advanced age at the time of diagnosis and stroke are poor prognostic factors associated with overall survival. This is the first largest Korean-based study that assessed mortality and prognosis in patients with BP and compared the mortality with BP patients from other countries.

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< ABSTRACT(IN KOREAN)>

한국인 유천포창 환자에 대한 후향적 임상 연구

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이지혜

수포성 유천포창 (Bullous pemphigoid)은 60세 이상의 노령에서 호발하며 표피하 수포를 형성하는 만성 수포성 질환으로서 기저막대의 반결합체 (hemidesmosome) 성분에 대한 자가 항체를 가지는 자가면역 피부질환이다. 수포성 유천포창 은 한국인에서의 발생빈도는 자가면역 수포성 질환 중 천포창 다음으로 발생률이 높다고 알려져 있으며 당뇨병, 건선, 류마티스 관절염, 궤양대장염, 자가면역 감상샘염 등과도 동반될 수 있다고 알려져 있다. 또한 치료하지 않을 경우 사망률이 높은 위중한 피부병이기 때문에 치료와 예후에 대한 분석이 매우 중요하나 현재까지 한국인 수포성 유천포창 환자에 대한 연구들은 비교적 적은 환자를 대상으로 시행하였고, 추적관찰의 기간도 짧아 한국인 수포성 유천포창 환자의 임상적 특징을 알기에는 부족한 점이 있었다.

이에 본 연구는 지난 20년간 강남세브란스병원 피부과 수포성질환 클리닉에서 진단 및 치료한 수포성 유천포창 환자의 임상적 특징과 사망률 및 예후인자를 분석해 보고자 하였다.

총 168명의 환자군이 모집되었다. 남자 84명 여자 84명이었으며 평균 연령은 69.15 세였다. 수포성 유천포창으로 진단받은 환자의 1년 사망률은 19.46%, 2년 사망률은 29.13%, 그리고 5년 사망률은 58.03%로 확인되었다. 또한 2011년 통계청 자료를 근거로 하여 한국인의 연령별 사망률과 환자의 사망률을 비교한 표준화 사망비를 구하여 보았을 때 전 연령에서 일반 인구에 비해 환자군의 사망률이 높게 나타났으며 특히 60대에서 80대 사이의 연령군에서는 7배 이상으로 높게 나타났다. 사망률과 관련한

예후인자로는 1년 생존률과 관련해서는 나이, 당뇨, 초기 진단이 늦어진 경우가 밝혀졌다. 당뇨를 동반한 수포성 유천포창 환자에서는 1년 사망률이 높게 나타났으며 오히려 초기 진단이 늦었던 환자들의 경우 1년 생존률이 낮게 나타났다. 전체 생존률에 대한 예후인자로는 나이와 뇌경색의 과거력이 있는 경우로 밝혀졌다. 나이가 많을수록 유의하게 사망률이 높아졌으며 뇌경색의 과거력이 있는 경우도 통계적으로 유의하게 사망률이 높아지는 것을 확인할 수 있었다.

본 연구에서 밝혀진 수포성 유천포창 환자의 사망률은 기존에 밝혀져 있는 수포성 유천포창 환자의 사망률과 크게 다르지 않았다. 유럽에서의 결과보다는 약간 낮은 사망률을 보였지만 대상군의 평균연령이 10세 정도 낮았다는 점을 고려할 때 한국인 수포성 유천포창 환자의 사망률도 유럽의 결과와 비슷하게 높다는 것을 확인해 볼 수 있었다. 또한 기존의 결과와 마찬가지로 수포성 유천포창 환자에서 일반 인구에 비해 높은 사망률을 확인할 수 있었다. 수포성 유천포창의 사망률을 높이는 예후인자로 고령과 뇌경색의 동반이 확인되었으며 이 결과 역시 기존에 미국과 유럽의 보고와 다르지 않음을 확인할 수 있었다. 또한 본 연구를 통해 질병의 중증도와 치료 방법 등은 전체적인 사망률을 결정하는데 영향을 주지 못한다는 것을 확인할 수 있었다.

본 연구는 한국인에서는 최초로 수포성 유천포창 환자를 대상으로 한 대규모 연구이며 아시아에서 최대 환자군을 대상으로 한 연구로 큰 의의를 지니며 아시아 수포성 유천포창 환자의 이해에 중요한 의의를 지닐 것으로 생각된다.

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핵심되는 말 : 수포성 유천포창, 자가 면역 수포성 피부질환, 유천포창, 사망률, 예후