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Incidence of Bullous Pemphigoid and Pemphigus in Korea

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Dear Editor:

Bullous pemphigoid (BP), pemphigus vulgaris (PV), and pemphigus foliaceus (PF) are autoimmune disorders that cause blistering of the skin and mucous membranes; they are characterized by the presence of tissue-bound and circulating autoantibodies. Few studies focusing on the incidence of these disorders have been reported[1-5].

Through this study, we aimed to evaluate the incidence of BP and compare with the incidences of PV and PF in Korea, as the primary endpoint[5]. In addition, we sought to compare the incidences of patients with BP, PV, and PF with those of age- and sex-matched statistical data using the Health Insurance Review & Assessment service (HIRA) from 2010 to 2016.

Information regarding all newly diagnosed cases of BP and all cases of the pemphigus group that occurred at Chonnam National University Hospital (CNUH) in Gwangju, Korea, between 2001 and 2016 was collected. In total, 296 patients (160 male and 136 female) with these autoimmune bullous diseases, with a diagnosis based on clinical, histological and immunopathological criteria, were finally included. The study was approved by the Institutional Review Board of the Chonnam National University Hospital (IRB no. CNUH-2021-028), and informed consent was obtained.

A total of 187 patients with BP (ICD-10 codes L12.0), 68 patients with PV (ICD-10 codes L10.0), and 41 patients with PF (ICD-10 codes L10.2) were identified. The incidence of BP increased with time; the average annual rate of increase in incidence was 1.67 (incidence rate ratio = 1.11; 95% confidence interval [CI], 1.1 ~ 1.2). In contrast, the incidence of pemphigus group did not show any significant change; the average annual rate of increase in the incidence of PV was 0.22 (incidence rate ratio = 1.04; 95% CI, 1.0 ~ 1.2) and that of PF was 0.19 (incidence rate ratio = 1.02; 95% CI, 1.0 ~ 1.2). The incidence rate ratio was the incidence rate among the exposed portion of the population, divided by the incidence rate in the unexposed portion of the population, and the average annual rate of increase was calculated by dividing the number of events by at-risk person-years.

As shown in Fig. 1, the incidence of BP in both our cohort and the general population tends to be in general agreement. This result could be attributed to the increasingly aging Korean population since 2010 and the advanced diagnostics and improved awareness regarding the disease.

In addition, the incidence of BP increases significantly over time in proportion to aging, and the risk for BP increases rapidly beyond the age of 60 years[4]. This trend is similar to the results of statistical data from age- and sex-matched controls from the HIRA (Fig. 2). In the recently published national literature, the incidence and mortality rates of PV and PF have increased in older age[5].

This study was the first to determine the actual incidence rate of BP, PV, and PF in Korea using a nationwide population-based database. Our patient cohort is sufficiently large to establish BP as the most frequent autoimmune bullous disease. Based on its significantly increasing incidence, more attention should be paid to BP as a public health concern.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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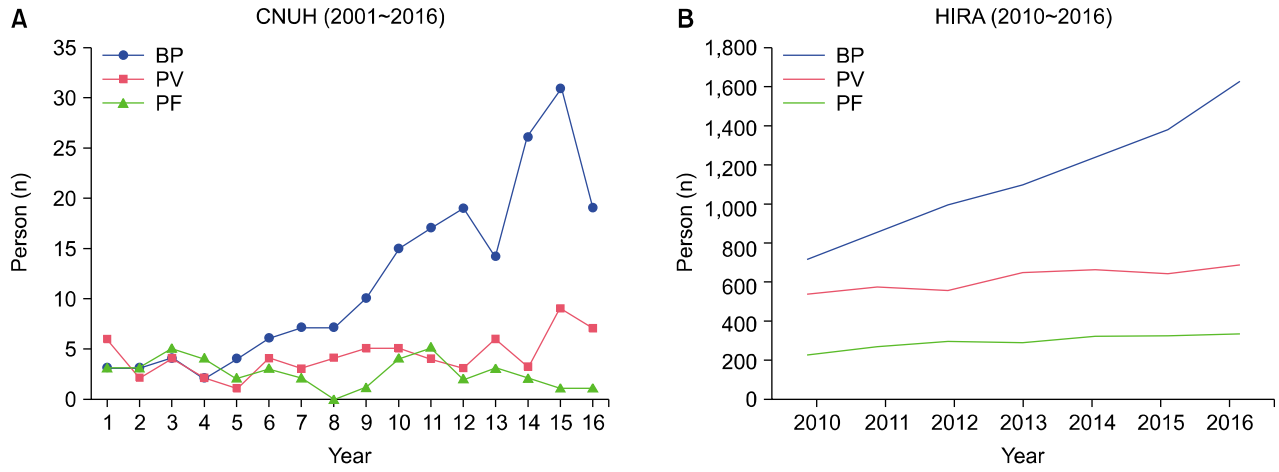

Fig. 1. Incidence of bullous pemphigoid, pemphigus vulgaris and pemphigus foliaceous in Chonnam National University Hospital (CNUH) (A), and Health Insurance Review & Assessment service (HIRA) (B). The incidence of bullous pemphigoid in both our cohort and the general population has increased significantly in the last 16 years, particularly since 2010. BP: bullous pemphigoid, PV: pemphigus vulgaris, PF: pemphigus foliaceus.

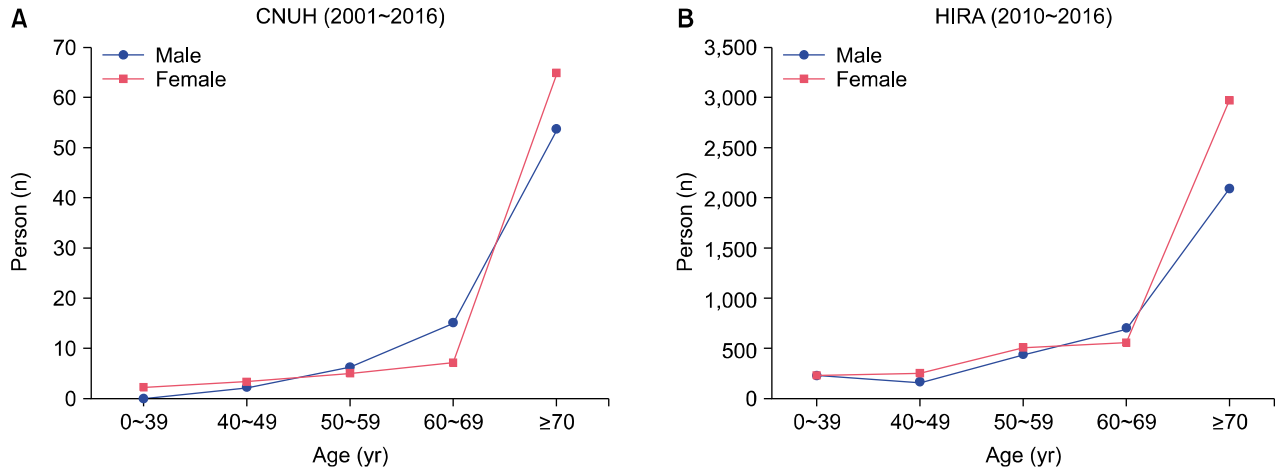

Fig. 2. Age- and sex-specific incidence of bullous pemphigoid based on data obtained from Chonnam National University Hospital (CNUH) (A) and Health Insurance Review & Assessment service (HIRA) (B). The incidence of bullous pemphigoid in both groups has increased significantly in proportion to aging, especially in individuals aged above 60 years.

DATA SHARING STATEMENT

Research data are not shared.

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A Case of Extranodal Natural Killer/T-Cell Lymphoma, Nasal Type with Dermatomyositis

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Dear Editor:

A 27-year-old male presented to our dermatology clinic with recurrent facial rash for five years, aggravated swelling for six months, and myalgia for the last two months. He presented with erythema and edema of the face (Fig. 1A). There was no rash on his trunk or limbs. Proximal dominant muscle weakness in his upper limbs (Medical Research Council grade 4) with grasping pain was noted. In addition, he complained that he had a history of rhinitis lasting 3 years.

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Laboratory investigations showed elevated creatine kinase, 1,108.2 U/L (55～170 U/L); lactate dehydrogenase, 1,283 U/L (98～192 U/L); alanine transaminase, 74.2 U/L (9～50 U/L); aspartate transaminase, 85.5 U/L (15～40 U/L); erythrocyte sedimentation rate, 19 mm/h (0～15 mm/h). Needle electromyography revealed fibrillation potentials, positive sharp waves, short duration low amplitude motor unit potential with increased polyphasic potential, and early recruitment in proximal muscles of the upper limb. A muscle biopsy of left bicep brachii on frozen sections suggested inflammatory myopathy (Fig. 1B). The right mandible skin lesions biopsy revealed epidermal focal parakeratosis, and liquefaction degeneration of basal cells, lymphocytic cell infiltration with mild atypia around vascular regions, appendages and in the fat lobules (Fig. 1C～E). Immunohistochemistry analysis showed positive of Ki67 (30%), LCA, CD2, CD3, CD8, CD4, TIA-1, and Epstein-Barr encoding region (EBER); negative of CD79a, CD20, CD3; weak positive of CD56 and granzyme B (Fig. 1F～J). We suspected that it was extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENKTL) but not sure, then the patient was admitted for study and treatment. After admission the patient developed a high fever, pharyngalgia, and dysphagia. A nasopharyngeal biopsy was performed and showed massive cells infiltration which were stained positive for cytoplasmic CD3, CD56, EBER (by *in situ* hybridization), granzyme B, TIA-1, and Ki67 (about 50%) with