Symposium on Autoimmune Diseases*

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(1) Epidemiology of SLE, PSS and DM in the Western Part of the Main Island and Shikoku Island in Japan

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For the purpose of study on epidemiology of systemic lupus erythematosus (SLE), progressive systemic sclerosis (PSS) and dermatomyositis (DM), questionnaires about the number of the patients with these diseases hospitalized from 1966 to 1970 and about their related detailed data were sent to physicians and dermatologists at 339 hospitals with more than 200 beds in the Kinki and Chugoku district (the western part of Japan main island) and Shikoku island. The population of this area covers 22% of the total population of Japan. Information was given by 32.2% of the hospitals, which counted 689 cases of SLE, 174 of PSS, and 94 of DM in total. The

Fig. 1. Prefectural distribution of number of patients with SLE hospitalized in Kinki, Chugoku and Shikoku Districts (1966-1970).

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ratios of SLE patient number to the population in each prefecture do not seem to show any significant differences (Fig. 1).

Annual incidence of SLE showed a sharp increase from 1968 on, but those of PSS and DM remained the same level during the five years of checking (Fig. 2-a). Age distribution of patients with SLE ranged from below ten to over 70 years old,

Fig. 2-a. Number of patients with SLE, PSS and DM admitted to 109 hospitals in Kinki, Chugoku and Shikoku districts (1966-1970).

Fig. 2-b. Age and sex distribution of SLE, PSS and DM in Kinki, Chugoku and Shikoku districts.

and 2nd to 4th decade patients made up 90% of the total, showing a peak for the 3rd decade. The peak in the age distribution of patients with PSS shifted to one decade older. Sex differences in patients with SLE, PSS or DM were remarkable, particularly with SLE, the ratio of female to male with SLE, PSS and DM being 12.4, 3.3 and 2.1, respectively (Fig. 2-b). Incidences of abnormalities in the main clinical and laboratory data, including immunology, on patients with SLE (532 cases), PSS (115 cases) and DM (66 cases) were presented.

Analysis of the detailed data on 367 patients in the Kinki district diagnosed as having SLE by doctors in charge shows that 75% of the patients shall be said to have SLE, according to the preliminary criteria for SLE presented by the American Rheumatism Association (ARA) which indicates the existence of four or more of the 14 manifestations selected for SLE (Fig. 3). However, our questionnaires skipped two items of the 14 manifestations, i. e. oral or nasopharyngeal ulceration and discoid lupus, so that four or more manifestations were actually obtained from 12 items, excluding these two manifestations. Comparison of clinical and laboratory findings was made between two defined groups of patients with and without SLE according to the preliminary criteria. Besides the 12 manifestations mentioned above, the group without SLE showed much less frequently myalgia, cardiac enlargement, and

Fig. 3. Classification of 376 patients in Kinki district according to diagnostic criteria of SLE.

various pulmonary lesions on a chest X-ray examination and a positive RA-test than those with SLE did. However, there were observed, with equally high frequency among both groups, manifestations such as running fever, photosensitivity, negative tuberculin skin reaction, high sedimentation rate, hyper- γ -globulinemia and antinuclear factor (Figs. 4-a and 4-b).

Fig. 4-a. Comparison of clinical manifestations between the patients with 4 or more and those with 3 or less items of ARA preliminary criteria.

Fig. 4-b. Comparison of laboratory findings between the patients with 4 or more and those with 3 or less items of ARA preliminary criteria.

The cumulative mortality rate of 325 patients with SLE in the Kinki district during intervals of observation of half a year, one year, 3 years and over 3 years was 12%, 15%, 25% and 30% respectively, and that of 60 patients with PSS, 8%, 10%, 17% and 20% respectively (Fig. 5).

Fig. 5. Cumulative mortality rate in Kinki district.