CLINICAL MANIFESTATIONS OF THE EARLY DEAD CASES IN PATIENTS WITH SYSTEMIC SCLEROSIS (SS)

Junichi Kaburaki¹), Masataka Kuwana²), Takeshi Tojo²), Makoto Takano¹), Yuzo Funatsu¹)
¹) Department of Medicine, Tokyo Electric Power Company Hospital, Tokyo, Japan.
²) Department of Medicine, Keio University, Tokyo, Japan.

Patients with Systemic Sclerosis (SS) have a wide variety of clinical manifestations. Some patients are found to have no progressive involvement of the skin and internal organs. Although the reason for a subset of the severe forms in patients with SS is still not clear, it is generally accepted that the insidious involvement of the cardiopulmonary system is the main cause of the decreased survival in patients with SS¹). In this study, the clinical manifestations of SS patients who died within 5 years of the first visit to Keio University hospital are retrospectively analyzed to elucidate a severe form in patients with SS.

PATIENTS AND METHODS
One hundred fifty-six patients with SS were studied. They did not have any overlapping features suggestive of systemic lupus erythematosus, polymyositis or dermatomyositis. Patients who died within 5 years of their first visit were chosen, and the cause of death was clarified in each case. The clinical manifestations of these patients who died because of cardiopulmonary involvement were retrospectively analyzed.

RESULTS AND DISCUSSION
Thirty-two of our SS patients were found to have died. The cause of death could be determined in 21 cases. Fourteen of them died within 5 years of their first visit. Five patients died of heart failure, and 5 patients died of respiratory failure. These data confirmed the previous report which showed that the cardiopulmonary system plays an important role in the survival of patients with SS¹). However, only one patient died of renal failure. This may be due to the development of new drugs, angiotensin converting enzyme inhibitors, and hemodialysis. As for other causes of death, 2 patients had cerebrovascular diseases, and one patient had gastric adenocarcinoma. Clinical manifestations in the 10 patients who died of complications in the cardiopulmonary system were compared with those in 62 patients who survived for more than 5 years after
Kaburaki J et al.; Severe form in SS
their first visit. The frequency of Sjögren's syndrome was significantly lower in the patients who died early than in the surviving patients (Table). In addition, it was suggested that the frequency of proteinuria was higher in those who died early than in the surviving patients. Oddis et al. found a correlation between Sjögren's syndrome and vasculitis in patients with SS2). However, their mean observation period was 12.7 years, which was longer than our observation period of 5 years. Therefore, it is necessary to follow up the clinical course of these 5-year survival patients. In this study, it is suggested that the clinical manifestations, such as Sjögren's syndrome and proteinuria, may be valuable in finding one of the severe forms in patients with SS, especially those who had the involvement of the cardiopulmonary system.

REFERENCES

Table. Clinical manifestations in the patients who died early and the surviving patients

<table>
<thead>
<tr>
<th>Clinical manifestations</th>
<th>Dead</th>
<th>Surviving</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sjögren's syndrome</td>
<td>10%(1/10)</td>
<td>50%(31/62)</td>
<td>p&lt;0.05</td>
</tr>
<tr>
<td>Proteinuria</td>
<td>20%(2/10)</td>
<td>2%(1/62)</td>
<td>p&lt;0.10</td>
</tr>
</tbody>
</table>