Short Communication

Polyarthralgia Suggests Poor Prognosis in Systemic Sclerosis with Both Pulmonary Fibrosis and Anti-DNA Topoisomerase I Antibodies

Junichi KABURAKI¹, Masataka KUWANA², Yutaka OKANO³, Michito HIRAKATA², Takeshi TOJO¹ and Yasuo IKEDA²

Departments of Internal Medicine, ¹ Tokyo Electric Power Company Hospital, ² Keio University School of Medicine, ³ Nippon Kokan Hospital and ⁴National Tokyo Dai1 Hospital

Abstract: Clinical features were studied in patients with systemic sclerosis (SSc) who had both pulmonary fibrosis and anti-DNA topoisomerase I antibodies and who ultimately died of this organ involvement. The subjects consisted of 62 patients with SSc. Medical records were retrospectively reviewed. The frequency of polyarthralgia was significantly higher in these patients than in other SSc patients. However, the frequency of positive reaction of rheumatoid factor was not significantly different. These results suggested that polyarthralgia as fibrotic change in the skin and contracture shows a poor prognosis in these SSc patients.

Keywords: systemic sclerosis, pulmonary fibrosis, anti-DNA topoisomerase I antibodies, polyarthralgia

Pulmonary fibrosis is one of the important internal organ involvements on which the diagnosis and prognosis of patients with systemic sclerosis (SSc) are based. We have already reported that anti-DNA topoisomerase I antibodies in sera from patients with SSc are associated with pulmonary fibrosis. Moreover, reaction to the specific epitope region 4 (ER4), amino acid residues 658-700 on the DNA topoisomerase I molecule, predicts the progression of abnormal pulmonary function tests. However, other clinical manifestations which are related to poor prognosis in SSc patients with both pulmonary fibrosis and anti-DNA topoisomerase I antibodies remain unknown. Our objective in this study is to clarify the clinical features which suggest poor prognosis in these patients.

Sixty-two Japanese patients with SSc were studied. They were diagnosed as having SSc according to the preliminary criteria by the American Rheumatism Association. All patients had both pulmonary fibrosis and anti-DNA topoisomerase I antibodies. Pulmonary fibrosis was defined as bibasilar interstitial fibrosis on chest radiography. Anti-DNA topoisomerase I antibodies were detected by the double immunodiffusion method. Medical records were retrospectively reviewed. Statistical analyses were performed by previously described methods.

The cumulative survival rate at 5 years from the first visit to Keio University Hospital was 84%. Seventeen patients died of respiratory and/or cardiac failure. Group A included SSc patients with both pulmonary fibrosis and anti-DNA topoisomerase I antibodies who died of respiratory and/or cardiac failure. Group B consisted of other SSc patients.

Table I Clinical features of systemic sclerosis (SSc) patients who had both pulmonary fibrosis and anti-DNA topoisomerase I antibodies

<table>
<thead>
<tr>
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<th>Group A (n=17)</th>
<th>Group B (n=45)</th>
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<tr>
<td>Female to male ratio</td>
<td>16:1</td>
<td>39:6</td>
<td>n. s.</td>
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<td>Age at the disease onset (years)</td>
<td>47.2±12.2</td>
<td>40.9±13.1</td>
<td>n. s.</td>
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<td>Age at the first visit (years)</td>
<td>51.0±11.5</td>
<td>44.1±12.9</td>
<td>n. s.</td>
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<tr>
<td>Observation period (years)</td>
<td>7.8±5.3</td>
<td>8.7±6.3</td>
<td>n. s.</td>
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<tr>
<td>Polyarthralgia</td>
<td>10 (59%)</td>
<td>14 (31%)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Rheumatoid factor</td>
<td>6 (35%)</td>
<td>11 (24%)</td>
<td>n. s.</td>
</tr>
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</table>

Results are expressed as the mean value ± standard deviation.

Group A included SSc patients with both pulmonary fibrosis and anti-DNA topoisomerase I antibodies who died of respiratory and/or cardiac failure. Group B consisted of other SSc patients.
or cardiac failure. The clinical manifestations in these 17 patients (designated as group A) and 45 other patients (designated as group B) were compared (Table 1). The gender, age at the disease onset, age at the first visit, and observation period were similar in these two groups. The frequency of polyarthralgia was significantly higher in group A than in group B, but the frequency of positive reaction of rheumatoid factor in the latex agglutination test was not significantly different.

We observed that polyarthralgia is found in SSc patients with both pulmonary fibrosis and anti-DNA topoisomerase I antibodies, and who ultimately died of this organ involvement. However, the frequency of a positive reaction of rheumatoid factor in these patients was similar to that in other patients. Therefore, these results suggested that fibrotic change in the skin and contracture as well as active synovitis cause polyarthralgia. Further prospective studies are necessary to clarify this point. In conclusion, polyarthralgia indicates a poor prognosis in SSc patients with pulmonary fibrosis and anti-DNA topoisomerase I antibodies.

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**REFERENCES**


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*Reprint request to* : Dr. Junichi Kaburaki. M. D. Department of Internal Medicine, Tokyo Electric Power Company Hospital, 9–2 Shinunomachi, Shinjuku-ku, Tokyo 160, Japan Tel. 03–3341–7121 Fax. 03–3341–9787