Raynaud's Disease with Sclerodactylyia

A Follow-Up Study of Seventy-one Patients

By Richard G. Farmer, M.D., Ray W. Gifford, Jr., M.D., and Edgar A. Hines, Jr., M.D.

During the course of previous studies of Raynaud's disease in women and girls, and of patients with systemic scleroderma, we encountered the records of 71 patients with Raynaud's disease and secondary sclerodermatous changes confined to the digits (sclerodactylia). These patients all had Raynaud's disease as defined by the criteria of Allen and Brown, namely: (1) episodes of Raynaud's phenomenon precipitated by cold or emotion, (2) bilaterality of the phenomenon, (3) absence of gangrene or the presence of only minimal degrees of cutaneous gangrene of the digits, (4) absence of any systemic disease that might account for the occurrence of Raynaud's phenomenon, and (5) duration of symptoms of 2 years or longer. In addition to the Raynaud's disease, all 71 patients had sclerodermatous changes in one or more digits when first examined at the Mayo Clinic, but none had any evidence of cutaneous gangrene in other sites or of systemic scleroderma.

Because of the confusion that has resulted from the various classifications of scleroderma, and in light of O'Leary and Waisman's contention that patients with Raynaud's disease and sclerodactylia should be considered to have systemic scleroderma of a type that they called "aerosclerosis," it was thought worth while to conduct a follow-up study of these patients. Follow-up information was obtained from all of the patients at re-examination at the clinic or by correspondence or both.

Preliminary Observations

The 71 verified cases of Raynaud's disease were divided into a medical group (40 patients) and a surgical group (31 patients), depending on whether or not a cervicothoracic sympathectomy had been performed for the relief of symptoms. The ages of the medically treated patients ranged from 22 to 68 years (average 39.3) at the time of the original diagnosis at the clinic. The ages of the patients treated surgically ranged from 18 to 56 years (average 35.5). The average age for the entire group was 37.6 years (table 1). Only 5 patients (7 per cent) were men; however, many of the cases were selected from a group that had included only women.

At the time of the original diagnosis, trophic changes of the digits (ulcerations, fissuring, chronic paronychia) had been observed in 26 of the 71 patients (36.6 per cent); 15 of the 26 were subsequently treated by means of cervicothoracic sympathectomy. Amputations of digits had been performed for 3 patients (4 per cent), involving amputation of single digits in 2 instances and of 2 digits in 1 case. These were performed primarily because of the presence of trophic changes; extensive gangrene was not observed.

Results of Follow-up Study

Except for 1 patient who died within a year, the time of follow-up after the original diagnosis at the clinic ranged from 2 to 31 years, the average being about 10 years for the entire group. Of the 40 patients treated medically, 10 reported that the condition had improved, 10 thought it worse, and 17 believed it was unchanged. Three patients were dead. Among the 31 patients treated surgically, 10 believed the condition to be better, 6 worse, and 12 about the same (table 2). Three were dead.

Trophic changes of the digits had developed subsequent to the time of initial examination in 3 of the patients treated medically and 5 of the patients treated surgically.
Table 1

Ages at Time of Original Diagnosis of Seventy-one Patients with Raynaud’s Disease and Sclerodactyly; Category of Treatment

<table>
<thead>
<tr>
<th>Age, years</th>
<th>Medical treatment</th>
<th>Surgical treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>9-20</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>21-30</td>
<td>9</td>
<td>11</td>
</tr>
<tr>
<td>31-40</td>
<td>12</td>
<td>6</td>
</tr>
<tr>
<td>41-50</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>51-60</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>61 or more</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>40</td>
<td>31</td>
</tr>
</tbody>
</table>

Average age 39.3 yr. 37.6 yr.

For whole group

Table 2

Follow-up Information Regarding Sixty-five Living Patients with Raynaud’s Disease and Sclerodactyly

<table>
<thead>
<tr>
<th>Condition of Raynaud’s phenomenon and sclerodactyly</th>
<th>Medical group</th>
<th>Surgical group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>Per cent</td>
</tr>
<tr>
<td>Better</td>
<td>10</td>
<td>27</td>
</tr>
<tr>
<td>Same</td>
<td>17</td>
<td>46</td>
</tr>
<tr>
<td>Worse</td>
<td>10</td>
<td>27</td>
</tr>
<tr>
<td>Total</td>
<td>37</td>
<td>100</td>
</tr>
</tbody>
</table>

Of the 6 patients who died (8.4 per cent), 2 died of cerebrovascular accidents, and 1 each of coronary arterial disease, cirrhosis of the liver, pneumonia, and ovarian carcinoma. None of the patients who died had systemic scleroderma. The average age at the time of death was 49.7 years, and death occurred on an average of 4.7 years after the diagnosis of Raynaud’s disease had been made at the clinic (table 3).

Systemic scleroderma apparently had developed in 3 of the 71 patients (4 per cent). Brief reports of their cases follow.

Report of Cases

Case 1

A 48-year-old woman, first seen at the clinic in 1940, had had Raynaud’s phenomenon, involving both hands, for 15 years; trophic changes had been present for 5 years. Scleroderma of the fingers was observed. All peripheral arteries were pulsating normally, and no primary systemic disease or organic arterial disorder could be found. A letter from the patient in 1948 indicated that the Raynaud’s phenomenon had involved the feet also and that the scleroderma had extended to the arms, chest, and face. A report in 1951 stated that the scleroderma had progressed and that the patient had a “heart ailment.” According to a letter received in January 1960, the sclerosis has remained about the same, but the patient now has dysphagia.

Case 2

A 21-year-old woman was seen at the clinic in 1942 with Raynaud’s phenomenon, involving both hands, which had been present for 5 years. No underlying cause could be determined, and no trophic changes were found, but sclerodactyly was noted. All peripheral arteries were pulsating normally. A cervicothoracic sympathectomy was performed. A follow-up letter in 1951 revealed that the sclerosis had increased and involved both arms as well as the hands; trophic changes of the digits had developed. In addition, progressive dysphagia had been present for 6 years.

Case 3

A 23-year-old woman was first seen at the clinic in 1926 with Raynaud’s phenomenon, involving the hands, of 6 years’ duration. Trophic changes and sclerodactyly were present. The peripheral arteries were pulsating normally, and no primary systemic disease or organic arterial disorder was found. A right cervical sympathetic ramisection was performed at that time. The patient returned in 1932, and because of a poor result from the first operation a bilateral cervicothoracic sympathectomy ganglienection was performed. A letter from her physician in 1943 described changes involving the arms, face, and hands that were consistent with a diagnosis of scleroderma. The patient was seen again at the clinic in 1957, and a diagnosis of scleroderma was made. At this time the Raynaud’s phenomenon was unchanged, but sclerodermatous changes of
the skin on the hands, arms, face, and trunk were noted. Calciosis cutis was present in the hands, and there were trophic changes of the digits. A letter received in January 1960 indicated that the patient’s condition has remained about the same.

In the first 2 cases the subsequent diagnosis of systemic scleroderma is only presumptive; however, the subjective complaints by the patients make this diagnosis likely. The third patient was re-examined at the clinic and the diagnosis of systemic scleroderma was confirmed.

Discussion

The relationship between systemic scleroderma and sclerodermatous changes confined to the digits (sclerodactytya) occurring secondary to long-standing Raynaud’s disease has not been clear. However, in this study only 4 per cent of the patients who originally had Raynaud’s disease and sclerodactytya were found to have developed systemic scleroderma, and there seems to be no justification for the belief that the two are the same disease. The incidence of sclerodactytya has been reported as being 10 to 12 per cent among patients with Raynaud’s disease. It should be emphasized that sclerodactytya is often a late development and, as the current study bears out, is frequently associated with trophic changes. The total number of patients with trophic changes, including those in whom these changes developed during the period of follow-up, was 34 (48 per cent).

As mentioned, only patients who had Raynaud’s phenomenon for at least 2 years prior to the diagnosis of Raynaud’s disease were included in this study. Sclerodermatous changes in the digits among patients who have had Raynaud’s phenomenon for less than 2 years often represent an early sign of systemic scleroderma. However, after the diagnosis of Raynaud’s disease has been established, the development of sclerodactytya will be followed by systemic involvement only rarely (4 per cent in this study). The use of strict criteria for the diagnosis of Raynaud’s disease will help to prevent mistakes in distinguishing between systemic scleroderma with secondary Raynaud’s phenomenon and Raynaud’s disease with sclerodactytya. The correct diagnosis is important because of the vastly different prognosis of these two conditions.

A recent study of 271 patients with systemic scleroderma revealed that about half of those who were traced died during a follow-up period which averaged 8.5 years. However, in the current series of 71 patients with Raynaud’s disease and sclerodactytya, only 8.4 per cent died during the follow-up period, and none of these had systemic scleroderma.

Summary and Conclusions

The records of 71 patients with Raynaud’s disease and sclerodactytya seen at the Mayo Clinic were studied. Follow-up information was obtained for all, with periods varying from 1 to 31 years. Systemic scleroderma seemed to have developed in only 3 patients. Six patients had died but none from systemic scleroderma. This study does not support the belief that systemic scleroderma and sclerodactytya secondary to Raynaud’s disease are the same disease.

References

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