Features of systemic sclerosis (scleroderma) in South African goldminers

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Summary

Systemic sclerosis is more common among men exposed to silica-containing dust than in the general male population. The clinical features of systemic sclerosis in a group of 24 black goldminers are described. The better-known presenting features of systemic sclerosis, including Raynaud's phenomenon and dysphagia, were rare in this population. Initial presentation was usually with nonspecific features including swelling of the feet or hands, weakness, arthralgia or symptoms of respiratory or cardiac disease. Clinical evidence of pleural or pericardial involvement was more common than is usually described in non-occupational systemic sclerosis. Interstitial lung disease was frequently encountered and renal disease was rare.


An association between goldmining and systemic sclerosis was first described by Erasmus in 1957. A report from South Africa (published in 1985) described 79 cases, all in white goldminers. A more recent study documented a high incidence of the disease among black South African goldminers. Since that report was published, an increase in the incidence of the disease in that population has been observed, suggesting that the disease is either becoming more frequent or being recognised more readily.

We describe the symptoms and signs of systemic sclerosis (scleroderma) in 24 black goldminers to draw attention to the features of the disease in this occupational group.

Patients and methods

Patients with systemic sclerosis were detected when they presented to the medical service of the Ernest Oppenheimer Hospital during the 7-year period September 1981 - August 1988. All the patients were assessed clinically, chest radiographs and ECGs were taken, and routine biochemical and haematological blood tests and lung function tests were done. All the men were employed as goldminers at the time of presentation.

Results

A total of 24 men with systemic sclerosis were detected during the period of the study. Ten cases were diagnosed during the initial 5 years of the study period and 14 during the last 2 years.

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REFERENCES

The mean age of the men (± SD) was 40.7 ± 7.62 years, and they had worked underground in goldmines for a mean of 19.0 ± 7.44 years.

Presenting symptoms included swelling of the feet, swelling of the hands, loss of weight, dyspnœa, joint pains, chest pain and dysphagia (Table I). Four cases were detected after a routine chest radiograph showed basal lung opacification, pleural disease or cardiomegaly.

| TABLE I. PRESENTING SYMPTOMS IN 24 GOLDMINERS WITH SYSTEMIC SCLEROSIS (%) |
|-----------------------------------|------------------|------------------|------------------|
| Swelling of hands or feet         | 41               |
| Loss of weight                    | 18               |
| Dyspnœa                           | 18               |
| Cough                             | 14               |
| Chest pain                        | 14               |
| Polyarthritis                     | 14               |
| Dysphagia                         | 8                |
| Raynaud’s phenomenon              | 4                |

At the time of diagnosis all the patients fulfilled the American Rheumatism Association criteria for the diagnosis of systemic sclerosis. All had the one major criterion, the presence of scleroderma (thickening of the skin, which becomes indurated, tethered and difficult to pick up) proximal to the metacarpophalangeal (MCP) or metatarsophalangeal (MTP) joints. In addition, all had at least one of the minor criteria for the diagnosis; these include sclerodactyty (scleroderma distal to the MCP or MTP joints), loss of substance or pitting of the digital pulps, and fibrosis of the lung bases. Most of the patients had small mouths and tight, shiny nasal skin. Areas of depigmentation of the skin were commonly seen, but calcinosis and telangiectasia were not apparent.

Twenty-three of the 24 men had evidence of interstitial lung disease. In 15 cases basal opacification was apparent on the chest radiograph. In these and a further 8 cases reduction of the forced vital capacity (FVC) or of the lung diffusion measurement provided evidence of interstitial lung disease. The mean FVC of the patients, expressed as a percentage of the American Thoracic Society predicted values, was 64.1 ± 14.30%, and the mean single-breath diffusion for carbon monoxide (DLCO) was 49.0 ± 17.68%. The FVC exceeded 75% of the predicted value in 4 cases and the DLCO in 2. Pleural involvement was clinically or radiologically apparent in 4 cases. Twelve patients had an abnormal ECG and 11 of them also had clinical evidence of cardiac involvement. Pericardial disease was apparent at some time in 8 patients. Problematic ulceration of the toes or fingers occurred in 7 men. Oesophageal involvement was evident in 5. Renal disease was detected in only 1 patient, who was also anaemic and thrombocytopenic at the time of initial presentation. Generalised muscle weakness noted in 3 patients was associated with raised muscle enzyme values. These findings are summarised in Table II.

Twelve of the men had suffered from pulmonary tuberculosis before the diagnosis of systemic sclerosis. Antinuclear antibodies were detected in 10 of the 13 patients who were tested. In 3 cases the antinuclear antibodies had a homogenous staining pattern, in 1 the antibody was not characterised, and in the remaining 6 the pattern was speckled.

Five of the men are known to have died. Death was associated with respiratory failure in all cases except 1, in which death occurred suddenly 1 month after the development of signs of cardiac involvement. Another 7 patients were lost to follow-up after leaving mine service owing to disability.

Table II. System involvement in 24 goldminers with systemic sclerosis.

<table>
<thead>
<tr>
<th>System Involvement</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proximal scleroderma</td>
<td>100</td>
</tr>
<tr>
<td>Interstitial lung disease</td>
<td>96</td>
</tr>
<tr>
<td>Previous tuberculosis</td>
<td>60</td>
</tr>
<tr>
<td>Myocardial disease</td>
<td>46</td>
</tr>
<tr>
<td>Pericardial disease</td>
<td>33</td>
</tr>
<tr>
<td>Finger or toe ulceration</td>
<td>29</td>
</tr>
<tr>
<td>Oesophageal disease</td>
<td>21</td>
</tr>
<tr>
<td>Pleurisy</td>
<td>17</td>
</tr>
<tr>
<td>Polyarthritis</td>
<td>17</td>
</tr>
<tr>
<td>Myopathy</td>
<td>13*</td>
</tr>
<tr>
<td>Renal disease</td>
<td>4</td>
</tr>
</tbody>
</table>

*Evidence for myopathy was sought in only the last 3 subjects included in this study. All had elevated muscle enzymes.

Discussion

Systemic sclerosis is considered to be an occupational disorder in goldminers. Although the features of the disease are characteristic, they may readily be overlooked in a patient with less specific manifestations including respiratory or cardiac disease, loss of weight, joint pains, weakness or isolated leg oedema.

Only 2 of the patients in this study complained of dysphagia. It is possible that cases with oesophageal involvement have been missed, since only the men with dysphagia and a few of the early cases were investigated; there is evidence from studies of carcinoma of the oesophagus in black South Africans that all but extreme dysphagia is tolerated in this population.

Only 1 patient presented with Raynaud’s phenomenon, and men in this working population may have a particular resistance to its development. No other case of that disorder has been seen over a 10-year period at the medical facility provided for this population, notwithstanding the fact that approximately 11 000 men work daily with pneumatic drills, an occupation in which as many as 50% of workers have been reported to suffer from Raynaud’s phenomenon.

The subjects in this report are similar to those described by others with regard to the frequency with which cardiac abnormalities were detected. Pericardial involvement and clinically apparent pleural disease were more common in the present study than in other reports. Erasmus was also of the opinion that pericardial and pleural disease were more common among goldminers with systemic sclerosis than among subjects with the non-occupational form of the disease.

The rarity of renal disease in the present study contrasts with Le Roy’s clinical series, in which renal disease was apparent in 35% of cases. Renal disease is said to be the cause of death in 50% of cases of systemic sclerosis but did not account for any of the deaths in the present series.

Interstitial lung disease was apparent in all but 1 of the patients in the present series (96%). Hunnigkake and Faucistate that interstitial fibrosis is found in nearly all cases post mortem. In Le Roy’s clinical series, however, only 43% of the patients had evidence of lung disease, and a recent Italian study designed specifically to detect lung disease in systemic sclerosis found that 62% of patients had interstitial lung disease (defined according to the same criteria as in the present series).

Systemic sclerosis is associated with silica dust exposure and not necessarily with silicosis. Silicosis has been reported in 60% of black South African goldminers with systemic sclerosis, but was apparent in only 13% of white South African goldminers with the disease. The incidence of systemic sclerosis was found to be 24 times higher among black men exposed to...
silica dust than in the general population. Haustein et al. found systemic sclerosis to be 25 times more common among men exposed to silica dust than among male workers without silica dust exposure. Sluis-Cremer et al. showed a direct association between systemic sclerosis and cumulative, lifetime silica exposure.

A strong association between systemic sclerosis and pulmonary tuberculosis in the present series was noted, but no explanation is apparent. The association cannot be attributed to the prevalence of silicosis alone; of a group of 859 men with silicosis from the same working population, 12% had had pulmonary tuberculosis in the past.

The increase in the incidence of systemic sclerosis among miners working in the Orange Free State goldfields might be attributable to increased awareness of the disease on the part of the medical personnel. However, the medical unit to which these men presented has been interested in and aware of systemic sclerosis as an occupational disease for the last 12 years. There is therefore a distinct possibility that the disease has become more common; this could be associated with an increase in the age of this working population or with an increase in silica-dust exposure.

Rodnam et al. and Haustein and Ziegler consider systemic sclerosis associated with silica-dust exposure to be the same as the non-occupational disease. Erasmus, writing mainly about white South African goldminers, believed the disease to be different with regard to its acute presentation, its association with marked disability, and the frequency of pleural and pericardial involvement. In the present study the major differences in presentation were the rarity of Raynaud's phenomenon and of dysphagia; both these features could reflect characteristics of black South Africans rather than the nature of the disease. On the other hand, the almost invariable finding of interstitial lung disease and the rarity of renal disease suggest that systemic sclerosis in this occupational group does differ from the non-occupational form of the disease.

REFERENCES