Clinical and X-ray Features of Sarcoidosis

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Introduction

The etiology of sarcoidosis remains unknown. It is, however, well known that the disease begins as a mononuclear alveolitis proceeding to a multinucleated giant cell granulomatous disease. Bronchoalveolar lavage has shown clear distinctions in cell populations among normals, sarcoidosis and other interstitial lung disease. Normal individuals show less than 10% lymphocytes and less than 1% polymorphonuclear cells whereas sarcoid patients have 33% ± 6% lymphocytes which can go as high as 55% ± 8% in very active disease. Alveolar macrophages are also increased and include a population of activated cells. In far advanced disease increased numbers of polys are seen.

The various subpopulations of lymphocytes also show characteristic differences. The study population of sarcoid patients show an average of 90% ± 4% of all lung lymphocytes as T-lymphocytes compared to the expected 65%—80% in normals. In parallel with the increase in T-lymphocytes is an increase in number of "activated" T-lymphocytes, cells that release a monocytic chemotactic factor in addition to numerous other factors.

The alveolitis in any given patient can be unstable with spontaneous change from high intensity to low intensity (75% of cases) and vice versa (12% of cases). When high intensity alveolitis is present, deterioration in at least one lung function occurred in 87% of cases within 6 months while low intensity alveolitis indicated stability or improvement. The intensity is best gauged by lavage and/or gallium scan. If one study indicates low intensity, a second is not necessary.
The disease is usually staged roentgenographically:

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>0</td>
<td>Normal x-ray</td>
</tr>
<tr>
<td>I</td>
<td>Hilar adenopathy</td>
</tr>
<tr>
<td>II</td>
<td>Hilar adenopathy and parenchymal infiltrates</td>
</tr>
<tr>
<td>III</td>
<td>Infiltrates alone</td>
</tr>
</tbody>
</table>

Among 303 patients with sarcoid the pattern of x-ray presentation was:

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Percentage</th>
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</thead>
<tbody>
<tr>
<td>Normal</td>
<td>8%</td>
</tr>
<tr>
<td>Unilateral hilar adenopathy</td>
<td>4%</td>
</tr>
<tr>
<td>Bilateral hilar adenopathy</td>
<td>33%</td>
</tr>
<tr>
<td>Mediastinal adenopathy</td>
<td>1%</td>
</tr>
<tr>
<td>Lung infiltrates</td>
<td>21%</td>
</tr>
<tr>
<td>Adenopathy and infiltrates</td>
<td>33%</td>
</tr>
</tbody>
</table>

The course of untreated disease in these same 303 patients was:

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Progression</td>
<td>56%</td>
</tr>
<tr>
<td>Stable</td>
<td>25%</td>
</tr>
<tr>
<td>Remission</td>
<td>16%</td>
</tr>
<tr>
<td>Died</td>
<td>3.6%</td>
</tr>
</tbody>
</table>

Other studies have reported overall mortality of 5% to 10%. However, the prognosis of sarcoidosis in patients whose involvement is primarily in skin or lymph nodes, or a combination of these sites (Stages 0 and I) is similar to that of men in the general population.

The diagnosis is usually made on the basis of clinical presentation, x-ray appearance, and confirmed by biopsy. Ancillary testing including gallium scanning, angiotensin converting enzyme (ACE) and bronchoalveolar lavage can play a part, though perhaps their greatest value may be in following the course of disease. Klech et al. have reported on the overall sensitivity and specificity of these tests in assessing the activity of sarcoidosis.

Figure 2
Activity was defined by clinical symptoms together with x-ray appearance.

**In this study, T-lymphocytes were measured in peripheral blood taking advantage of the observation that in sarcoid as the alveolar T-lymphocytes show a rise, the peripheral T-lymphocytes show a fall.

The Kviem-Siltzbach test, although it has a reported accuracy of 65%-90%, is used less because of the difficulty in obtaining a potent validated antigen. Serum lysozyme activity and measurement of transcobalamin II are as yet of uncertain value.

**X-Ray Features**

Intrathoracic lymphadenopathy occurs in 75—85% of patients with sarcoidosis (Boeck sarcoid) at some time during the course of their disease. Parenchymal lung lesions are seen in more than 60% of patients.

The classic distribution consists of bilateral hilar and right paratracheal adenopathy (Fig. 1). Left paratracheal adenopathy occurs more frequently than originally appreciated but may be more difficult to detect.

Bilateral paratracheal adenopathy (alone), unilateral hilar adenopathy (Fig. 2), and anterior mediastinal adenopathy are sufficiently rare that a diagnosis other than sarcoidosis should be considered.

The most common parenchymal abnormality is a reticulonodular lesion (Figs. 3&4). Less common pulmonary manifestations include a miliary pattern (simulating miliary tuberculosis), and large nodular densities resembling pulmonary metastases (Fig. 5). Pulmonary fibrosis is reported to occur in 10—25% of cases.

Other intrathoracic findings include cardiomegaly due to cor pulmonale or direct cardiac involvement (4—8%), spontaneous pneumothorax (2—3%), pleural effusion (1—3%) and segmental or lobar atelectasis (1%).
REFERENCES


Report from Europe

R.D.C. Brackenridge, M.D.
Regional Editor

The Assurance Medical Society held its third regional scientific meeting, this time in Edinburgh, on October 14, 1983. The meeting, which took place in the buildings of the Royal College of Physicians of Edinburgh was chaired by the President of the British Medical Association, Dr. R.F. Robertson, and included papers by the President of the College, emeritus Professor R.H. Girdwood and other eminent Edinburgh physicians, some of whom are also advisers to various Scottish life companies.

In the evening delegates and their wives were entertained to a reception and tour of the historic home of the Marquis of Linlithgow, Hopetoun House, near Edinburgh, followed by dinner in the magnificent ballroom.

A changing scene in Britain — an underwriting colleague gives a warning:

"It has been said that British term assurance portfolios are written on the back of the enormous amounts of 'thick premium' endowment business in our traditional life funds, and to some extent the recent phenomenon of guaranteed acceptance is, it seems to me, an acknowledgement of that. To my mind British underwriters have had an easy ride relative to their American counterparts, because the vast quantities of savings money in British funds tend to over-ride the need to get the mortality absolutely right. There is some evidence that the British life market is becoming more protection orientated; if this trend continues we shall be faced with the sterner task of underwriting mainly thin-premium business, and getting it right!"

Of Interest

A paper entitled "Insurance Premium Reductions—A Motivating Factor in Long Term Hypertensive Management," written by Marvin Moser, M.D., John Rafter, M.D., and Jerzy Gajewski, M.D., has been accepted for publication in the Journal of the American Medical Association.