UNILATERAL CALCIFIED FIBROTHORAX WITH COR PULMONALE: FAILURE TO IMPROVE WITH DECORTICATION

SYNOPSIS

A 57 year old woman in cor pulmonale and cardiac failure secondary to a right calcified fibrothorax underwent decortication after the lung had been imprisoned for approximately 31 years. Although the operation was successful, it failed to arrest her gradual deterioration from cor pulmonale.

INTRODUCTION

Chronic pleural thickening is known to have a profound effect on cardio-pulmonary function (1). It is, however, rarely associated with pulmonary arterial hypertension and cor pulmonale (2); the result following decortication in such patients is not well documented. We report a patient with right calcified fibrothorax in cor pulmonale and cardiac failure who underwent decortication.

CASE REPORT

The patient was a 57 year old woman who was first seen in April 1977 because of dyspnoea and ankle oedema of one week’s duration. Five months earlier she had been admitted to another hospital for the same complaints. The significant findings on examination were signs of a right fibrothorax and congestive cardiac failure. Blood pressure was 120/80 mmHg. A loud pulmonary second sound was present. She had been in good health previously with no history of pulmonary tuberculosis or chest trauma. She never smoked. She noticed her right chest was 'sunken in' at the age of 18 years. The chest radiograph revealed cardiomegaly, congested lung fields and massive pleural calcification of the right hemithorax (Figure 1). The electrocardiogram showed evidence of right ventricular hypertrophy. She improved with treatment but was re-admitted in March 1978 for cardiac failure. The arterial oxygen tension (PaO₂) was 56 mmHg; arterial carbon dioxide tension (PaCO₂) 58 mmHg and arterial pH 7.38. Her hemoglobin was 14.6g%.

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Table 1. PULMONARY FUNCTION DATA

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Pre-operative</th>
<th>Post-operative</th>
<th>Normal Predicted Value</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Nov 78</td>
<td>May 79</td>
<td>Nov 79</td>
</tr>
<tr>
<td>VC, litre</td>
<td>0.61</td>
<td>0.78</td>
<td>0.79</td>
</tr>
<tr>
<td>FRC, litre</td>
<td>1.05</td>
<td>0.96</td>
<td>1.33</td>
</tr>
<tr>
<td>RV, litre</td>
<td>0.97</td>
<td>0.81</td>
<td>1.09</td>
</tr>
<tr>
<td>TLC, litre</td>
<td>1.58</td>
<td>1.45</td>
<td>1.68</td>
</tr>
<tr>
<td>FEV₁, litre</td>
<td>0.51</td>
<td>0.51</td>
<td>0.68</td>
</tr>
<tr>
<td>DLCO ml/min/mmHg</td>
<td>5.8</td>
<td>7.2</td>
<td>—</td>
</tr>
<tr>
<td>Arterial blood gases</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pH, units</td>
<td>7.35</td>
<td>7.33</td>
<td>7.31</td>
</tr>
<tr>
<td>PaO₂, mmHg</td>
<td>37</td>
<td>44</td>
<td>52</td>
</tr>
<tr>
<td>PaCO₂, mmHg</td>
<td>53</td>
<td>62</td>
<td>63</td>
</tr>
</tbody>
</table>

She improved with diuretic therapy and was maintained on this as an outpatient. Pulmonary function tests in November 1978 (Table 1) demonstrated severe restrictive impairment. Hypoxemia had increased compared to the results in March 1978. Her condition gradually deteriorated; she remained in cardiac failure. Her chest radiograph showed worsening cardiomegaly. In view of her deteriorating condition, decortication was performed in March 1979. At operation, a very heavily calcified rigid pleura completely imprisoning a macroscopically normal lung was found. Lung biopsy was not done. The patient tolerated the procedure well and the post-operative recovery was uneventful. She improved symptomatically. The lung function tests in November 1979 showed improvement in her ventilatory capacity and in the PaO₂. Her chest radiograph showed a reduction in the cardiac size. Cardiac catheterization in December 1979 revealed a pulmonary artery pressure of 52/25 mmHg, normal right atrial pressure and a cardiac index of 3.3 L/min/M².

She remained fairly well while on diuretics but gradually deteriorated again. Her haemoglobin rose to 17.7 g% and her hematocrit to 59%. She died in May 1980 from cor pulmonale and cardiac failure.

DISCUSSION

This case report documents the downhill course of a patient with right calcified fibrothorax in cor pulmonale and cardiac failure in spite of decortication. The fibrothorax had been present for 31 years. It would appear the prognosis was not altered significantly by the decortication.

Pulmonary decortication has been described since the 1890s. Decortication of the lung has been shown to improve pulmonary function (3, 4). The imprisoned lung is released from its limiting corset of thickened and/or calcified pleura, with consequent improvement in ventilatory capacity. Although improvement depends on the presence or absence of underlying parenchymal disease, the duration of the lung collapse did not adversely affect the outcome of the surgical procedure nor the degree of functional recovery (5, 6).

It is well known that severe pleural disease can produce striking abnormalities of pulmonary function (1). Courmand and Richards (7) described five patients with arrested pulmonary tuberculosis and unilateral fibrothorax. All demonstrated greater decreases in total lung capacity and vital capacity than would have been expected from the basis of parenchymal disease alone. On the ipsilateral side chronic pleuritis with or without constriction imposes a mechanical disadvantage on the lung by causing a loss of parietal elasticity due chiefly to pleural symphysis and deposition of fibrotic tissue. The loss of elasticity persists in some degree after even the most satisfactory decortication and probably accounts for the fact that pulmonary function seldom returns to the predicted normal postoperatively. Other irreversible changes such as overdistension of the good lung, shift of the mediastinum, elevation of the diaphragm and decrease in the size of the hemithorax may also contribute to incomplete functional restoration (6). Unilateral pleuritis has been shown not only to reduce ventilation and perfusion to the lung with pleural disease but also causes a limitation of alveolar expansion.

Fig 1 Chest radiograph showing calcific right fibrothorax and cardiomegaly
on both the involved and normal appearing sides (8).

The development of pulmonary hypertension and cor
pulmonale was described in four patients with unilateral
pleural constriction by Robin et al (9). Evidence of severe
pulmonary hypertension developed after many years of
apparently normal circulatory dynamics, similar to our pa-
tient. Pulmonary hypertension, in their view, seemed
relatively independent of hypoxemia and of the degree of
alveolar hypoventilation, and they therefore postulated a
humoral agent secreted by the poorly perfused lung as
causing the pulmonary hypertension (Goldblatt Lung). One
of the four patients had decorticication with some relief of
dyspnoea but died 21 months later from cardiac failure.

Cohen et al (10) described a patient with massive
unilateral pulmonary and pleural fibrosis associated with
severe hypoxemia and pulmonary hypertension who im-
proved following pneumonectomy. A second patient with
similar findings and improvement following pneumonec-
tomy was reported by Simons et al (11). Our patient had
severe calcific pleuritis with cor pulmonale and heart
failure. The pulmonary artery pressure nine months post-
operatively was 52/25 mmHg and it was most probably
higher pre-operatively. Pneumonectomy was not done as
the underlying lung was considered to be normal. The
severe and possibly irreversible pulmonary hypertension
and the persisting restrictive deformity of the chest wall
post-operatively with its accompanying effects on the
mechanical function of both lungs are likely to have con-
tributed to the unsuccessful outcome of the decorticication.

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