Bilateral Recurrent Spontaneous Pneumothoraces in Sarcoidosis


A case of recurrent bilateral pneumothoraces is described in a young patient with sarcoidosis. An intercostal tube drainage was done for right sided pneumothorax. Patient was put on corticosteroid treatment and with this treatment there has been no recurrence of pneumothorax.

Key Words: Pneumothorax, Sarcoidosis.

Spontaneous pneumothorax is a relatively common complication in some of the interstitial pulmonary diseases like eosinophilic granuloma and lymphangioleiomyomatosis. The occurrence of spontaneous pneumothorax has also been reported in cryptogenic fibrosing alveolitis and sarcoidosis. However, bilateral recurrent spontaneous pneumothorax occurs rarely in patients with sarcoidosis.

CASE REPORT

A 38-year-old male complained of sudden onset of chest pain in April, 1985. A chest X-ray done at that time revealed bilateral pneumotorax (Fig. 1). Patient was treated with bed rest at this time and his chest X-ray revealed complete clearing of pneumothoraces after four weeks of conservative treatment. However, a week later patient again developed bilateral pneumothoraces. This time right sided pneumothorax was significant and required an intercostal tube drainage. There was complete expansion of underlying collapsed right lung after seven days. A review of his chest X-rays showed a pattern of diffuse interstitial lung disease (Fig. 2). Physical examination revealed a right supraclavicular lymph node measuring 1.5 x 1.5 cm. Rest of the systemic examination was normal. Biopsy of the right supraclavicular lymph node was done. The lymph node tissue was stained with hematoxylin eosin stain. The section showed nonconfluent discrete granulomas consisting of epitheloid cells without necrosis (Fig. 3). There was no lymphocyte cuffing.

Stains for acid-fast bacilli and fungi were negative. Arterial blood gas values during hospitalization were: paO₂ 59.5 mmHg, paCO₂ 48.8 mmHg, and repeat arterial blood gas analysis on follow up after three months of treatment revealed paO₂ 88.9 mmHg, paCO₂ 34.7 mmHg. Pulmonary function tests at the time of discharge from the hospital revealed: Forced vital capacity (FVC) 1.99 liters, FEV₁ (%FVC) 70%, Peak ex-
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Fig. 2. Chest X-ray showing bilateral diffuse interstitial lung pattern.

Fig. 3. Microphotograph showing discrete, non-confluent granulomas. There is no necrosis. The granulomas consist of epitheloid cells and occasional gaint cell (hematoxylin and eosin, x900).

Fig. 4. Chest X-ray showing improvement with prednisolone treatment.

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Table 1. Pulmonary Function Tests

<table>
<thead>
<tr>
<th>Test</th>
<th>Baseline</th>
<th>Follow-up</th>
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<tbody>
<tr>
<td>FEV1 (%)</td>
<td>72%</td>
<td>72%</td>
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<tr>
<td>FVC</td>
<td>2.87 L</td>
<td>2.87 L</td>
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<tr>
<td>PEFR</td>
<td>2.9 liters/sec</td>
<td>5.1 liters/sec</td>
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<tr>
<td>MMEFR</td>
<td>0.88 liters/sec</td>
<td>1 L/sec</td>
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<tr>
<td>DLCO</td>
<td>9.01 ml/min/mmHg</td>
<td>18.12 ml/min/mmHg</td>
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Repeat pulmonary function tests after two months of treatment revealed: FVC 2.87 L, FEV1 (%) 72%, PEFR 5.1 liters/sec, MMEFR 1 L/sec, DLCO 18.12 ml/min/mmHg. Bronchoalveolar lavage was done from right middle lobe, 50% of the infused normal saline was recovered, total cell count of the fluid was $8.2 \times 10^6$ cells/ml with alveolar macrophages 27%, lymphocytes 63%, polymorphs 10%. Liver function tests revealed serum proteins 6.9 g/dl with an albumin of 4.4 g/dl, serum alkaline phosphatase 215 IU (normal upto 270 IU), SGOT/SGPT 31/56 IU, serum Ca$^{++}$ 11.6 mg%. Mantoux test with 5 TU PPD was $0 \times 0$ mm after 72 hours. Kveim test was not done. Serum angiotensin—converting enzyme (SACE) activity was 28 units/ml (normal $8.7 \pm 1.3$ units/ml). Serum immunoglobulins were not done.

Course and follow up: Patient was put on 60 mg prednisolone per day for four weeks and later on the dose of prednisolone was tapered off gradually. He showed a remarkable improvement in his symptoms and chest X-ray (Fig. 4). During one year period of follow up there has been no recurrence of pneumothorax and he remains asymptomatic.

DISCUSSION

The diagnosis of sarcoidosis in our patient was based upon the negative PPD reaction, hypercalcaemia, restrictive lung function tests and the demonstration of multiple non-caseating granulomas in the supraclavicular lymph node. Elevation of serum angiotensin-converting enzyme (SACE) activity and bronchoalveolar lavage fluid (BALF) lymphocytosis further substantiated the diagnosis.

Pneumothorax is a rarer complication of sarcoidosis and occurs in less than 2% of patients with sarcoidosis. Their relationship has been debated. According to some this complication is not coincidental but is directly related to the pulmonary pathology and it occurs either due to rupture of subpleural bleb or necrosis of sub-
pleural granuloma. According to others their association may be simply furtuitous as both tend to occur in younger people. However, necrosis of subpleural tubercles appears to be an important aetiological factor as these patients show a remarkable response to prednisolone therapy. Hence an early and adequate treatment with corticosteroids may be beneficial in these patients.

Our patient has shown a good clinical response with corticosteroid treatment and there has been no recurrence of pneumothoraces during the follow-up period.

REFERENCES