Short reports

Pleurectomy in the management of massive pleural effusion associated with primary lymphoedema: demonstration of abnormal pleural lymphatics

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Pleural effusions occur in patients with primary lymphoedema. Twenty such cases from published reports have been reviewed by Beer *et al.*¹ Although these effusions may be massive with rapid reaccumulation after thoracocentesis, we have been able to find only two brief reports of cases in which obliteration of the pleural space by pleurectomy^{2,3} was considered necessary. We have found no histological documentation of the abnormality of the pleural lymphatics in this condition. We report a case of primary lymphoedema with recurrent massive pleural effusions successfully treated by pleurectomy.

Case report

A 46-year-old man first presented to the Johannesburg Hospital in 1978 with generalised primary lymphoedema associated with bilateral pleural effusions. The case was reported by Warman *et al.*⁴ Pulmonary function studies showed an appreciable restrictive defect and hypoxaemia (table).

The patient was well until June 1980, when he began to experience exertional dyspnoea. He was a life-long non-smoker. Examination showed an obese man with generalised lymphoedema of the limbs and trunk. The nails were normal. The respiratory rate was 24/min at rest and examination of the chest indicated bilateral pleural effusions. These were confirmed radiologically. Examination of the cardiovascular system showed nothing abnormal. Pulmonary function tests (table) showed a considerable deterioration in lung volumes and flow rates since 1978. Thoracocentesis yielded straw-coloured fluid (protein content 4.5 g/dl; specific gravity 1032; microbiological culture negative; no malignant cells). Six litres of fluid were removed from the pleural cavities with appreciable symptomatic improvement.

Five weeks later the patient was readmitted with severe dyspnoea due to bilateral reaccumulation of pleural fluid. Five litres were removed, again resulting in symptomatic improvement. Pulmonary function tests after thoracocen-

tesis (table) showed an improvement in lung volumes and flow rates with persistent hypoxaemia.

From August to December 1980 the patient was readmitted on three further occasions for therapeutic thoracocentesis. It was decided that a left parietal pleurectomy should be performed, the left pleural effusion being consistently larger. Thoracotomy revealed a moderate amount of serous fluid in the left pleural space with a dense layer of fibrous tissue encasing the lung. Decortication was performed and the considerably thickened parietal pleura was removed; some pleura was left adherent to the mediastinum and diaphragm.

Histological sections of the pleura (fig) showed appreciable fibrous thickening distinguishable from that associated with chronic empyema or haemothorax by the presence of several considerably dilated lymphatic channels lined by flattened cells. Some newly formed lymphatic channels were seen in the fibrous tissue, which in some areas appeared oedematous.

The patient was discharged on the 10th postoperative day with a completely expanded left lung, but with pulmonary function tests (table) still showing severe restriction and hypoxaemia. Subsequent pulmonary function tests showed progressive improvement in lung volumes, flow rates, and arterial oxygen tension (table). Serial chest radiographs show only thickening of the residual pleura on the left. The right pleural effusion has not reaccumulated but pleural thickening is present.

Discussion

By means of lymphangiography Kinmonth *et al.*⁵ showed various lymphatic abnormalities in 107 cases of primary lymphoedema affecting the lower limbs, including aplasia, hypoplasia, and dilatation. While it has been suggested that the pleural effusion in primary lymphoedema is related to hypoplasia of the pleural lymphatics,² with consequent defective lymphatic drainage,⁶ we have been unable to find previous histological documentation of the abnormality of the pleural lymphatics in this condition. Histological sections of the parietal pleura in our patient clearly show abnormal dilated lymphatics, neogenesis of lymphatic channels, and oedematous tissues in some areas, suggesting some defect in lymphatic drainage (fig). As with the limbs,

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Accepted 7 December 1982

Relationship of pulmonary function to thoracocentesis and pleurectomy

	<i>Predicted values</i>	<i>1978</i>	<i>1980 June</i>	<i>1980 August, after thoracocentesis</i>	<i>1980 10 days after thoracotomy and left pleurectomy</i>	<i>1981 3 months after surgery</i>	<i>1981 9 months after surgery</i>	<i>1982 18 months after surgery</i>
FVC (l)	5.20	1.80	1.50	2.40	1.80	2.30	3.10	2.90
TLC (l)	7.55	4.25	2.90	4.10	3.15	3.60	4.45	4.85
FEV ₁ (l)	4.00	1.55	1.20	2.00	1.55	1.95	2.70	2.40
MEF _{50%} (l/s)	5.80	3.20	1.75	2.40	2.20	2.20	4.00	3.60
Pao ₂ (kPa)	9.2*	7.5	8.0	7.5	7.5	8.5	8.4	8.4

*Normal value for Johannesburg (altitude 1763 m): 10 kPa.

FVC—forced vital capacity; TLC—total lung capacity; FEV₁—forced expiratory volume in one second; MEF_{50%}—maximum expiratory flow after exhalation of 50% of FVC; Pao₂—arterial oxygen tension.

Conversion: SI to traditional units—Pao₂: 1 kPa = 7.5 mm Hg.

(a) Fibrous thickening of parietal pleura with lymphocytic infiltrate along costal surface (arrow head), dilated new lymphatic channels within the fibrous tissue (solid arrow), and oedematous tissue on free surface (open arrows). (Haematoxylin and eosin $\times 50$.) (b) Higher magnification ($\times 150$) of the centre section of (a) to show new lymphatic channels with endothelial lining.

probably various abnormalities of the pleural lymphatics exist.

Successful treatment of recurrent large pleural effusions in primary lymphoedema by pleurectomy has been reported in only two cases.^{2,3} In neither of these reports are the results of comprehensive serial pulmonary function studies available.

On initial presentation in 1978 our patient had an appreciable restrictive ventilatory defect and hypoxaemia, and by June 1980 considerable deterioration in static lung volumes and flow rates had occurred (table). These abnormalities are compatible with the presence of large pleural effusions.⁷ Obesity, which may be associated with a reduction in functional residual capacity and hypoxaemia, may have been an aggravating factor.⁸ While the reduction in flow rates may be related to pulmonary restriction with airway narrowing lymphoedema of the airways could also perhaps have been a contributing factor.

Serial pulmonary function studies after pleurectomy showed progressive improvement, stabilising nine months after operation. The slowness of the improvement may be related to the after-effects of thoracotomy,^{9,10} which have been shown to include impairment of lung volumes and diffusion for up to eight months and hypoxia for up to four months. Eighteen months after pleurectomy our patient still had an appreciable restrictive ventilatory defect (table), which could be due to several factors, including lymphoedema of the chest wall and lungs, obesity, and pleural thickening. Despite this he became symptom free and able to pursue a normal life. The right pleural effusion has never reaccumulated since the last thoracentesis just

before surgery and we have no explanation for this phenomenon.

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