DIFFUSE INTERSTITIAL PULMONARY FIBROSIS, DERMATOMYOSITIS AND LUNG CANCER — A CASE REPORT

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SYNOPSIS

A patient with diffuse pulmonary fibrosis, dermatomyositis and lung cancer is reported. It is recognised that there is more than a chance occurrence of diffuse interstitial fibrosis in dermatomyositis, lung cancer in dermatomyositis, and lung cancer in dermatomyositis, and lung cancer in diffuse interstitial fibrosis. We report a case with all three and a spinal schwannoma.

CASE REPORT

A 68 year old Chinese man was admitted to Tan Tock Seng Hospital on 12.9.77 for progressive dyspnoea, cough with small amounts of whitish sputum, and loss of appetite and weight for three weeks, and fever for one week. He had no significant past medical history. He had smoked 40 cigarettes a day for about 40 years. He denied smoking opium. He had worked as a building contractor previously. His temperature was 38 deg. C, pulse rate 80 per minute and blood pressure 120/70 mmHg. Fine crackling crepitations were heard over both lower zones anteriorly as well as posteriorly. There was no finger clubbing. Hemoglobin was 12.6g%, total white count 8,400 p.c. mm. and ESR 100 mm/Hr., RA factor, ANF and LE preparation were negative. Sputum cytology showed no malignant cells and smears for tubercle bacilli were negative. Arterial blood gases on ambient air showed pH 7.38, pCO_2, 32 mmHg. and PaO_2 62 mmHg. (predicted PaO_2 was 75). Pulmonary function tests showed a restrictive pattern (see Fig. 1). Chest x-ray showed streaky opacities over both lower zones with both diaphragms at the 9th rib position posteriorly (Fig. 2). He was treated with antibiotics and bronchodilators.

On 29.12.77, he was admitted again, but this time he complained of generalised weakness and difficulty in swallowing. Clinical examination revealed he had severe proximal muscle weakness of all four limbs and weakness of his neck flexors. He also had a violaceous rash over his forehead and malar regions. His liver was palpable at 2 cm. below the right costal margin. Serum potassium was 4.3mEq/L, LE preparation and ANF were negative. RA was positive. Serum glutamate-pyruvate transferase was 188U/L (N 9-36), serum alkaline phosphatase 143U/dl (N 32-105), serum bilirubin 0.6mg%, serum albumin 1.8g%, serum globulin 4.3g%, serum creatine phosphokinase was more than 20U/ml (N $<$ 4.5) and serum aldolase 26I.U./L (N 3-12). Liver biopsy showed "reactive hepatitis". Barium swallow showed no abnormalities. Electromyography showed a "myopathic" pattern consistent with polymyositis. Prednisolone 60 mg/day was started

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on 12.1.78 and within one week, he began to show improvement of muscle power. On 22.2.78 he developed klebsiella pneumonia in his right upper lobe. This responded to Gentamycin, but there were residual scars in the right upper lobe on the chest x-ray. The patient required 30-60 mg. prednisolone daily to maintain adequate function.

On 20.9.79 he was admitted to hospital for right sided weakness. Over the next few days, it gradually became a quadriparesis with a sensory level at T-4 anteriorly. Chest x-ray revealed an opacity in the right upper lobe (Fig. 3). Sputum cytology was now positive for malignant cells. A myelogram done showed an extradural block at C6-7 vertebral level, and a laminectomy revealed an extradural benign non-invasive tumour in that area. The tumour was removed and the histology showed a spinal schwannoma. The patient perished on 18.11.79 from post operative complications.

Pulmonary Function Tests

	Predicted	Result
VC (litres)	3.07	1.44
FRC (litres)	3.00	2.32
RV (litres)	2.29	1.54
TLC (litres)	5.11	2.98
MMFR L/sec	2.63	1.89
DCOml/min/mmHg	10.8	6.0

Fig. 1.

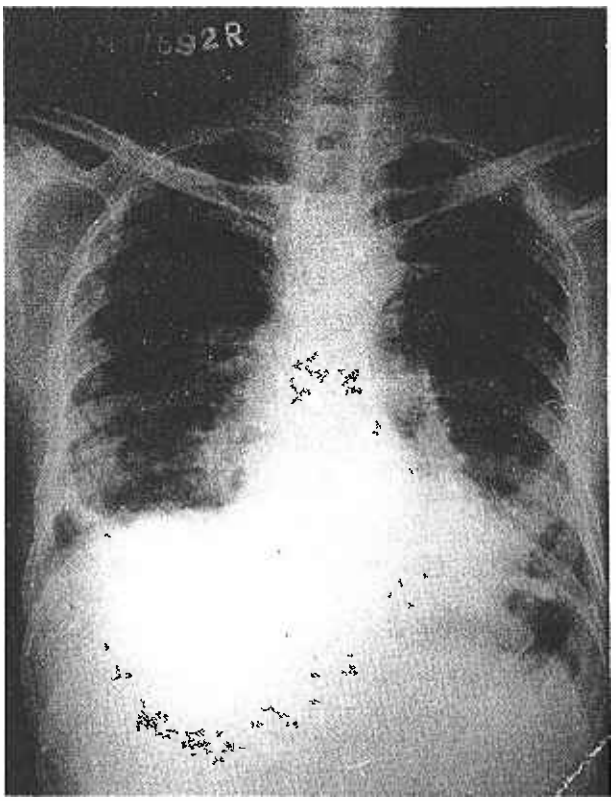

Fig. 2 CXR

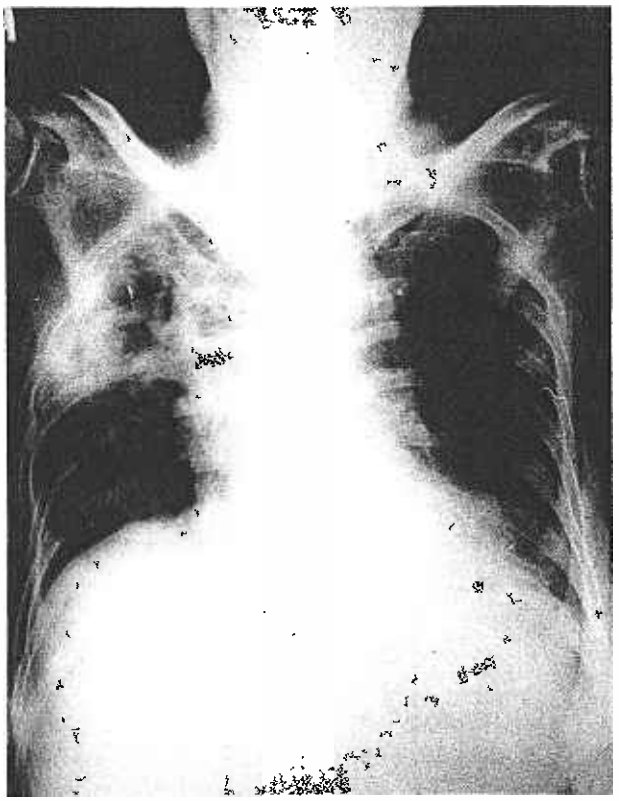

Fig. 3 CXR

CASE COMMENTS

The diagnosis of diffuse interstitial fibrosis in this patient was not proved by histology, but the clinical presentation, radiologic features, and the pulmonary function tests provided sufficient grounds for the diagnosis to be made (6). The diagnosis of dermatomyositis was made on the basis of the clinical findings viz. the proximal pattern of weakness and the violaceous rash, the raised serum enzymes, the electromyographic pattern, and the response to steroids. Lung cancer was suspected radiologically and confirmed by sputum cytology.

DISCUSSION

Diffuse Interstitial Lung Disease And Dermatomyositis.

Fibrosing alveolitis in polymyositis has been a reported association (3, 7 & 10). The pulmonary disease may precede, present simultaneously with, or appear after the muscle disorder. The interval between the onset of the two diseases may range from a few months to 3 years. Finger clubbing was not seen or mentioned in the reports, in contrast to its usual frequent occurrence in fibrosing alveolitis. In most instances, the polymyositis improved with steroids. There have been attempts to explain this association. As both of these disorders are of a presumed immunological basis, the reason for their association may be immunological too. Perhaps in polymyositis there is an autoimmune humoral or cytotoxic response directed against bronchiolar smooth muscle thus triggering off the pulmonary disease. However, a search for histological and immunological evidence (viz. immune complexes, immunoglobulins) has failed to produce any definite proof to confirm the abovementioned suspicions.

Lung Cancer In Fibrosing Alveolitis

Lung cancer has been reported to occur in fibrosing alveolitis (1, 5, 6 & 9). Turner-Warwick (11) has concluded from a retrospective study of 220 cases of cryptogenic fibrosing alveolitis that there is an increased incidence of lung cancer in fibrosing alveolitis. It has been postulated that chronic inflammation and fibrosis predispose to the development of malignant change. Lung cancer has been reported in pulmonary tuberculosis (8) and bronchiectasis (2). Analogous situations in other organs are the occurrence of malignancy in cirrhosis, in burn scars, in the colon of chronic ulcerative colitis, and in the oesophagus in achalasia. Womack and Graham (12) suggested the epithelial metaplasia in congenital cystic disease could be related to the development of bronchogenic carcinoma.

Lung Cancer And Dermatomyositis

Dermatomyositis occurring in an elderly man always raises the possibility of internal malignancy especially lung cancer. In our patient, the dermatomyositis antedated the tumour by 2 years.

Spinal Schwannoma

Pulmonary interstitial involvement may occur in neurofibromatosis (4). This patient only had a spinal schwannoma and no evidence of widespread neurofibromas.

Dermatomyositis, diffuse interstitial fibrosis and lung cancer are related to one another (Fig. 4) perhaps even in a causal manner. We report a patient with all the three diseases and in addition, a spinal schwannoma.

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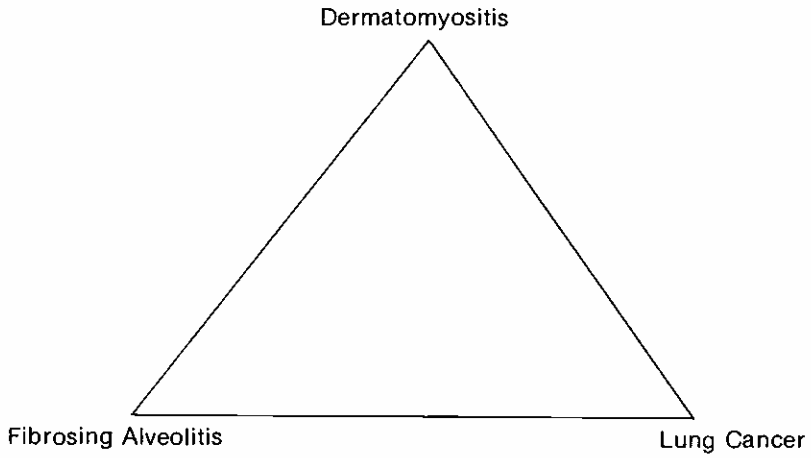

Fig. 4