Short reports

Adverse effect of pregnancy on familial fibrosing alveolitis

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Familial fibrosing alveolitis (pulmonary fibrosis) is a rare, chronic interstitial lung disease which usually presents in the fourth to sixth decades,¹ although clinical features have been described during childhood.² Since it is inherited as an autosomal dominant trait with variable penetrance¹ the two sexes are equally affected. Hence the disease can affect women of childbearing age. There are no previous reports of the effect of pregnancy on the disease or of the disease on pregnancy, and we report what we believe to be the first case of a pregnancy in a patient with familial fibrosing alveolitis.

Case report

A 25 year old symptom free woman was discovered to have diffuse, bilateral, basal pulmonary infiltrates on a routine pre-employment chest radiograph. She had digital clubbing and fine late inspiratory crackles in both lung bases. Measurement of pulmonary function in May 1982 showed a reduction in lung volumes and single breath carbon monoxide transfer factor. Spirometry showed increased maximum expiratory flow rates. Mild arterial oxygen desaturation was noted on exercise testing (fig).

The maximum work load achieved during exercise was 800 kpm/min (131 watts) (predicted 870 kpm/min (142 watts)*). The maximum oxygen uptake achieved was 1590 ml min⁻¹ (71 mmol min⁻¹) (predicted 2410 ml min⁻¹ (108 mmol min⁻¹)*). The maximum static elastic recoil pressure was increased at 55 cm H₂O (predicted 35.1 cm H₂O⁵); the exponential constant, k, of the volume pressure relationship of the lungs was 0.057 (predicted 0.120⁵) and the lung volume at 10 cm static elastic recoil pressure was 2.38 l (predicted 3.62 l⁵).

The patient's brother, aged 21, who had similar clinical and radiological features, had previously had an open lung biopsy confirming the presence of fibrosing alveolitis. Their maternal grandmother died at the age of 55 years from idiopathic pulmonary fibrosis, which had been confirmed at necropsy.

The patient remained symptom free with no appreciable change in lung volumes, gas transfer, or exercise performance until she had been pregnant for 26 weeks. From this

time she noticed increasing exertional dyspnoea and her pulmonary function deteriorated until the 34th week of gestation (fig). Treatment with prednisolone was commenced at a dose of 40 mg a day. At 38 weeks a normal boy was born by an induced vaginal delivery. Despite an increase in the dose of prednisolone to 80 mg a day and the addition of azathioprine, pulmonary function failed to return to the initial values and the patient remained incapacitated by breathlessness three months later.

Discussion

During the course of a normal pregnancy total lung capacity falls slightly close to term and vital capacity remains unchanged or even increases.^{6,7} Gas transfer increases or remains unchanged until the middle trimester, then falls to a value around or slightly below the pre-pregnant level.⁸ Thus the steady decline in lung volumes and gas transfer during pregnancy in this patient represents a non-physiological change and suggests an increase in disease activity in the lungs. This is supported by the post partum course of pulmonary function.

Sarcoidosis is the only diffuse interstitial lung disease to have been studied systematically during pregnancy: often pregnancy results in reduced disease activity although exacerbation may occur after delivery.⁹ Rheumatoid arthritis, a disease which may be associated with pulmonary fibrosis, frequently improves during pregnancy⁹ and systemic lupus erythematosus may also improve.^{10,11} Systemic lupus erythematosus may also develop for the first time during pregnancy and a fatal outcome of lupus pneumonitis has been reported in this setting.¹¹ The present case illustrates that it is not possible to infer an interaction between pregnancy and a lung disease on the basis of experience with similar lung diseases. The paucity of publications on this subject also reflects the lack of knowledge of the basic disease mechanism in fibrosing alveolitis.

Although the present patient did not have circulating immune complexes estimated during pregnancy, several authors have noted their presence in normal and pre-eclamptic pregnancy.¹² Since immune complexes are also found in patients with active idiopathic (non-familial) fibrosing alveolitis and may be important in the pathogenesis of this disease,¹³ possibly the pregnancy related decline of pulmonary function in this patient was mediated by this mechanism. Other immunological changes have been described in pregnancy, including enhanced (fetal) suppressor cell activity,¹⁴ although it is not

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Progress of pulmonary function in relation to pregnancy. Total lung capacity (■), vital capacity (□), transfer factor (◆), transfer factor/effective alveolar volume (◇, ml/min/mm Hg), and exercise desaturation () were plotted against time in months. All the mean predicted values indicated are derived from Cotes³ except for exercise desaturation value, which is from Jones.⁴ Conversion: Traditional to SI units—Transfer factor (TLCO): 1 ml/min/mm Hg = 0.33 mmol min⁻¹ kPa⁻¹ l⁻¹; transfer factor/effective alveolar volume (KCO): 1 ml/min/mm Hg = 0.33 mmol min⁻¹ kPa⁻¹ l⁻¹.*

Addendum

At the last review on 6 January 1984 there had been no appreciable change in lung volumes or gas transfer since September 1983. The dose of prednisolone had been reduced to 10 mg/day, azathioprine was being continued, and penicillamine had been started.

clear what effect this might have on the inflammatory process in fibrosing alveolitis.

As a result of the deterioration of pulmonary function during pregnancy this patient's prognosis is now worse than it was before she became pregnant. There has been no recovery in gas transfer during the three months since delivery. For this reason she has been advised against becoming pregnant again.

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