Case report

Pulmonary hypertension, systemic lupus erythematosus, and the contraceptive pill

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SUMMARY Pulmonary hypertension in systemic lupus erythematosus (SLE) in the absence of chronic parenchymal lung disease or pulmonary emboli is rare. We report such a case with an acute and rapidly progressive onset of symptoms in a patient who had started taking the contraceptive pill eight months previously.

A 22-year-old woman presented with a three-week history of increasing dyspnoea, pleuritic pain, and a skin rash. Three years previously she had an isolated episode of acute arthritis affecting some of the small joints of the hands and feet, which terminated spontaneously after one week. She remained well and had a normal first pregnancy, after which she started taking the contraceptive pill Eugynon 30 (ethinyloestradiol 30 µg, levonorgestrel 250 µg) for the first time.

Nine months later she was admitted to hospital looking extremely ill, dyspnoeic, centrally cyanosed, pyrexia, and she had a widespread vasculitic rash. Marked right ventricular hypertrophy and failure were evident, and on auscultation there was a loud pericardial rub.

Haematological investigations showed: Hb 13.6 g/dl, leucocytes $16 \times 10^9/l$, platelets $190 \times 10^9/l$, ESR 76 mm in the first hour. Numerous LE cells were present on three separate occasions.

Antinuclear antibody was found to a titre of 1:80 in IgG class, but antibody to dsDNA was not detected by radioimmunoassay nor was specificity of this antibody to extractable nuclear antigens detected. Lupus anticoagulant and anticardiolipin antibodies¹ were not found. Renal function tests and serum complement levels have remained normal throughout the illness to date. Biopsy of non-light-exposed skin showed deposits of IgG. Chest x-ray showed an acute left basal pneumonitis. Electrocardiogram and echocardiogram confirmed right ven-

tricular hypertrophy. A ventilation/perfusion lung scan and pulmonary arteriograms showed no existence of pulmonary emboli.

Within 48 hours of presentation, prednisone therapy (80 mg per day) was started. There was rapid clinical improvement, and in less than 12 hours pyrexia, leucocytosis, and tachycardia resolved and LE cells disappeared.

Right heart catheterisation 4 days later confirmed raised pulmonary artery (PA) pressure at 40/20 mmHg, pulmonary resistance (PR) was 456 dynes cm^{-5} (normal range 67, SD 23), while a repeat procedure after a further three weeks showed reduced PA pressure at 32/13 mmHg, and PR was 120 dynes $s\ cm^{-5}$. Repeat measurements after four months showed no further reduction.

Pulmonary function tests initially showed a mild restrictive pattern and decreased transfer factor, but after six months these have returned to normal. Attempted reduction of prednisone therapy over several months led to a relapse of pleuritis and pericarditis at a dosage of 10 mg per day. Prednisone was therefore increased to 40 mg per day and azathioprine introduced at a dosage of 150 mg per day. The patient remains well and has few symptoms one year later with maintenance therapy of prednisone 10 mg and azathioprine 150 mg per day.

Discussion

The mechanism of this type of pulmonary hypertension in SLE is uncertain and has been discussed recently by Asherson *et al.*² It may be caused by sustained vasoconstriction from an unknown source

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which might explain the frequency of occurrence of Raynaud's phenomenon in reported cases, though this feature was absent in our patient.

What role did the contraceptive pill play in this case? Our patient was well throughout a normal pregnancy and is therefore unlikely to have had significant pulmonary hypertension before starting the contraceptive pill. To account, though, for the degree of right ventricular hypertrophy at presentation, the onset of pulmonary hypertension must have occurred shortly after starting the contraceptive pill. This is consistent with the findings of Jungers *et al.*³ who showed a flare up incidence of 43% in patients with SLE within three months of starting oestrogens.

Thus it seems probable that the contraceptive pill

has initiated this episode of pulmonary hypertension in a patient with SLE. There are no similar previously reported cases.

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References

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- 3 Jungers P, Dougados M, Pelissier C, *et al.* Influence of oral contraceptive therapy on the activity of systemic lupus erythematosus. *Arthritis Rheum* 1982; **25**: 616-24.