

Heart transplantation for Churg-Strauss syndrome

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SUMMARY A patient with heart failure caused by Churg-Strauss syndrome was successfully treated with transplantation. The case was unusual because there was little evidence of Churg-Strauss syndrome in the lung. The patient remains well on standard transplant immunotherapy.

Churg-Strauss syndrome was first described in 1951.¹ It is characterised by blood and tissue eosinophilia and disseminated necrotising vasculitis in asthmatic patients. Heart failure occurs in 47% of patients and accounts for nearly half the deaths.² We report a 22 year old man with Churg-Strauss syndrome in whom heart failure developed necessitating heart transplantation. To our knowledge, this is the first report to describe heart transplantation in a patient with this syndrome.

Case report

A 22 year old man was first seen in July 1986 complaining of breathlessness and wheezing. The chest radiograph showed bilateral interstitial lung opacities and he had pronounced eosinophilia ($7.7 \times 10^9/l$). Pulmonary eosinophilia was diagnosed and resolved when he was treated with oral steroids and bronchodilators. He remained well on 10 mg/day of prednisolone until July 1987 when he was admitted to hospital with a sudden onset of cough and breathlessness. The radiograph (figure) showed considerable cardiomegaly and pulmonary oedema and the electrocardiogram showed low voltage complexes. Echocardiography showed biventricular dilatation with a small pericardial effusion. The ejection fraction was estimated to be 15%. The eosinophil count was $3 \times 10^9/l$. The dose of steroids was increased and the eosinophilia resolved within three days. Cardiac catheterisation showed a pulmonary capillary wedge pressure of 18 mm Hg and a mean pulmonary artery pressure of 24 mm Hg. Myocardial muscle biopsy showed prominent foci of young fibroblastic tissue with necrotic and hypertrophied myocytes and an infiltrate of lymphocytes, plasma cells, and eosinophils. No vasculitis was seen. Two months later the

heart failure worsened. Echocardiography showed further dilatation of the heart chambers and impairment of left ventricular function. Serum concentrations of urea and creatinine were 7.1 mmol/l and 0.11 mmol/l respectively, with a creatinine clearance of 113 ml/min. Microscopical examination of urine did not show haematuria, casts, or proteinuria. In October 1987, he was transferred to Wythenshawe Hospital. He was anuric and had a mean blood pressure of 30 mm Hg, that improved during treatment with adrenaline, intra-aortic balloon counterpulsation, and ventilation. Two days later, orthotopic cardiac transplantation was performed.

The excised heart weighed 340 g and there was dilatation of all chambers. The right ventricle

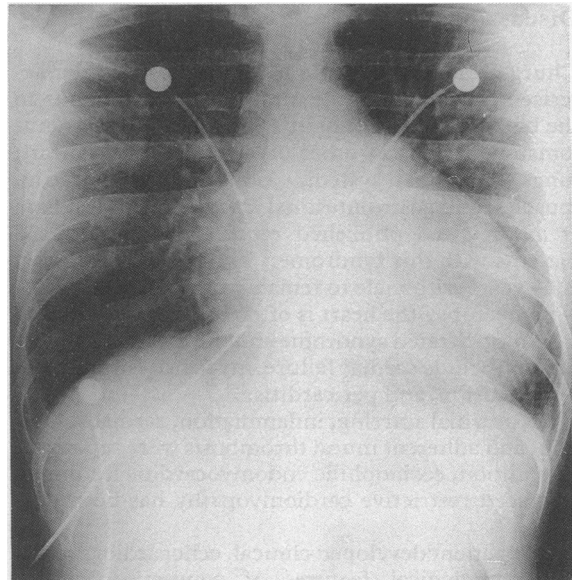


Figure Chest x ray showing considerable cardiomegaly and pulmonary oedema.

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weighed 83 g and the left 213 g. The myocardium had mottled yellow/white foci. The coronary arteries were macroscopically normal.

Fibrinoid necrosis with some giant cells was seen in some intramyocardial vessels. There was arteritis in the right coronary artery. All the cardiac valves were histologically normal. Subendocardial fibrinoid necrotic foci were seen especially in the left atrium and ventricle, as well as foci of fibrosis and associated hypertrophy. A mild eosinophilic infiltrate was present. A lung biopsy specimen showed an interstitial infiltrate of lymphocytes, plasma cells, and occasional neutrophils. No vasculitis was identified.

Postoperatively, after perioperative antithymocyte globulin, he was treated with cyclosporin, azathioprine, and steroids. Three weeks after transplantation, a fever and diffuse pulmonary shadowing developed. The donor heart came from a cytomegalovirus seropositive person and so we treated our patient for cytomegalovirus pneumonitis. He recovered after treatment with gancyclovir and cytomegalovirus immunoglobulin. Two months after discharge from hospital he was readmitted with cough and breathlessness and eosinophilia ($2 \times 10^9/l$). A cardiac biopsy specimen did not show rejection and we thought he had had a relapse of Churg-Strauss syndrome. The maintenance dose of steroids was increased; his condition improved and the eosinophil count returned to normal. The patient remains well twelve months after transplantation.

Discussion

Churg-Strauss syndrome is a rare disorder characterised by asthma, large numbers of eosinophils in the blood and tissue, angiitis, and necrotising granulomatous lesions in other organs such as the heart, lungs, nervous system, kidneys, spleen, skin, muscles, and gastrointestinal tract.¹⁻³ When Lanham *et al* reviewed published reports they found 138 patients with this syndrome.² The mean age of onset is 44 years with male to female ratio of 1.3:1.

At necropsy the heart is often found to be affected in Churg-Strauss syndrome and the cardiac manifestations include cardiac failure, myocardial infarction, hypertension, and pericarditis.¹⁻⁴

Myocardial scarring, inflammation, cardiac dilatation, and adherent mural thrombosis were reported.¹ In addition, eosinophilic endomyocarditis leading to advanced restrictive cardiomyopathy has been described.⁵

Our patient developed clinical, echocardiographic, and pathological features of congestive cardiomyopathy with multiple ventricular thromboses. The echocardiographic diagnosis of multiple ven-

tricular thrombi in a patient with Churg-Strauss syndrome has not previously been reported.

If untreated, the prognosis is poor with a five year survival rate of 25%. With immunosuppressive treatment a five year survival rate of 62% was reported.³ In our patient intractable heart failure developed despite conventional treatment. Cardiac transplantation was the only remaining option.

The fact that a "relapse" after transplant was felt to be due to resurgence of Churg-Strauss syndrome gives rise to the question of whether standard triple immunosuppressive treatment can be adequately targeted against such a syndrome. Because Churg-Strauss syndrome is basically caused by a humorally mediated response, we would not expect that cyclosporin with its specific action against T lymphocytes would have much effect on the suppression of Churg-Strauss syndrome. The single episode of relapse was successfully treated by increasing the dose of steroids and it may be that the best combination is azathioprine and cyclosporin, with lower doses of cyclosporin than usual, together with indefinite treatment with steroids, which currently we stop after a year.

As yet there is no evidence that the transplanted heart has been affected by the Churg-Strauss syndrome.

D T is Wellcome Transplant Fellow at Wythenshawe Hospital. We thank the following for their help and for permission to report this case. A N Rahman (consultant surgeon, Wythenshawe Hospital), N H Brooks (consultant cardiologist, Wythenshawe Hospital), A Holmes (renal physician, Hope Hospital), W P Stephen (consultant physician, Park Hospital).

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