Good Therapeutical Response of Goodpasture's Syndrome with Severe Renal Failure

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Sir,

The clinical course of anti-glomerular basement membrane (GBM) antibody-induced Goodpasture's syndrome is typically characterized by rapidly progressive glomerulonephritis, leading to end-stage renal failure, and by lung haemorrhages [1]. It is generally accepted that patients with anti-GBM nephritis whose serum creatinine exceeds 6.5 mg/dl or with severe crescent involvement, oliguria and advanced renal failure requiring haemodialytic treatment, do not respond to the appropriate treatment [2, 3].

We describe the case of a patient presenting with advanced renal failure whose conditions dramatically improved after plasma exchange and immunosuppressive therapy.

A 21-year-old woman was referred to our division for the development of rapidly progressive renal failure, with serum creatinine rising from 1.2 to 7.7 mg/dl in 12 days. Three months before admission, she had experienced recurrent episodes of haemoptysis followed by severe anaemia. She had a history of heavy smoking. On admission, physical examination revealed pallor and a systolic murmur, blood pressure was normal. Oliguria was not present. Laboratory data showed: haemoglobin 6.2 g/dl, haematocrit 18.8%, WBC 4,200/mm³, ESR 85 mm at the 1st hour, BUN 62 mg/dl, serum creatinine 7.7 mg/dl. Urine analysis showed microscopic haematuria, 24-hour protein excretion was 2.5 g. Protein electrophoresis, serum complement, cryoglobulins and antinuclear antibodies were normal. A chest X-ray was also normal. A renal biopsy immediately performed showed 20 glomeruli on light microscopy; all of them presented circumferential cellular crescents. Scarring and tubular alterations were not present. At immunofluorescence, heavy linear staining for IgG and C3 along glomerular basement membranes was present. An indirect immunofluorescence performed on normal renal tissue was positive for anti-GBM antibodies with a titer of 1:32.

The day following admission, serum creatinine rose to 8.5 mg/dl, and treatment was started with 3-liter plasma exchange every other day, pulse methylprednisolone 20 mg/kg/day for 3 days, then reduced to 1 mg/kg/day of oral prednisone, and cyclophosphamide 1.5 mg/kg/day. Three days later, serum creatinine was 9.6 mg/dl, and haemodialysis was started. After 3 weeks of treatment, serum creatinine decreased progressively to 4.2 mg/dl, and haemodialysis could be stopped. At the same time, circulating anti-GBM antibod-

ies were no longer detectable so that plasma exchange was interrupted after a total of 10 sessions. The patient was dismissed after 8 weeks of hospitalization: serum creatinine was 3 mg/dl, creatinine clearance 22 ml/min and circulating anti-GBM antibodies were absent. Cyclophosphamide treatment was interrupted, and corti-costeroid therapy was tapered to a maintenance dosage of 8 mg/day of prednisolone. After 12 months from the first manifestation of the disease, the patient maintains a stable renal function with absence of circulating anti-GBM antibodies. No other episodes of pulmonary haemorrhages have occurred.

It has been reported that in Goodpasture's syndrome treatment is successful only in patients with mild renal failure and moderate crescent formation [2]. The case described here, in agreement with another report [4], suggests that treatment with plasma exchange and immunosuppressive drugs is beneficial even in the presence of severe renal failure requiring haemodialysis, and extensive crescent formation. Early diagnosis and treatment are mandatory for these patients.

References

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