

Treatment of refractory Wegener's granulomatosis with antithymocyte globulin (ATG): an open study in 15 patients.

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BACKGROUND: A subset of patients with Wegener's granulomatosis does not respond sufficiently to cyclophosphamide and glucocorticosteroids or suffers of intolerable side effects. Anecdotal data suggest that antithymocyte globulin (ATG) may be a treatment option for these patients. We now describe 15 patients treated with ATG for refractory Wegener's granulomatosis.

METHODS: Fifteen patients with histologically proven active refractory Wegener's granulomatosis (seven unresponsive to cyclophosphamide, eight intolerant) were treated with ATG by a protocol (SOLUTION protocol) designed by the European Vasculitis Study (EUVAS) Group.

RESULTS: Before ATG administration, patients had received a mean of 5.2 (range 2 to 7) different therapeutic approaches including glucocorticosteroids and cyclophosphamide in all and experimental therapies in six, without control of disease activity [2.8 (range 1 to 7) relapses during a disease duration of 63.2 (range 18 to 180) months]. Thirteen of 15 patients showed a favorable response to ATG with partial (N= 9) or complete (N= 4) remission. During a follow-up of 21.8 (range 6 to 68) months, seven patients relapsed after a mean of 8.4 (range 2 to 24) months (five minor and two major relapses). Six patients are free of relapse for 22.3 (range 7 to 64) months. Two patients died, 1 and 3 days following the first dose of ATG, due to pulmonary hemorrhage and infection (one each). Although further immunosuppressive treatment was required in all surviving patients, a less intensive regimen could be applied in 12. Beside fever and chills associated with the first gift of ATG, ATG was well tolerated, with infections being observed in five cases and serum sickness in two.

CONCLUSION: Anti-T-cell-directed treatment with ATG may be a therapeutic option for severe refractory Wegener's granulomatosis if simultaneous infections and fluid overload have been ruled out. In patients with alveolar hemorrhage, ATG should only be used under special caution.