

Efficacy of Cyclophosphamide treatment for immunoglobulin G4-related disease with addition of glucocorticoids

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Objective:

To evaluate the efficacy, relapse and safety of glucocorticoid monotherapy vs combination therapy of cyclophosphamide (CTX) for IgG4 related disease (IgG4-RD).

Methods:

To evaluate the efficacy, relapse and safety of glucocorticoid monotherapy vs combination therapy of cyclophosphamide (CTX) for IgG4 related disease (IgG4-RD).

Results:

Of 102 patients, 52 patients were enrolled in group 1 and 50 in group 2. At 1 month, both groups achieved obvious improvement; response rate was 98.08% and 100% respectively. Relapse rate during 1 year was 38.46% in Group 1, including 12 cases with clinical relapse and 8 patients manifesting only serological relapse; whereas there was 12.00% of recurrence in Group 2, in which only 1 patient had clinical relapse and other 5 patients only got serological relapse. The mean time to flare in Group 2 was significantly longer than that in Group 1 ($P=0.018$). All relapsing patients in Group 1 were sensitive to immunosuppressants. Most of patients having more than 6 organs involved in Group 1 had disease flare during 1 year, and IgG4 levels of relapse cases were significantly higher than non-relapsing patients at baseline. Bile duct, lacrimal glands, lymph nodes and pancreas etc. were commonly relapsed organs in Group 1, while only 1 case had organ recurrence in Group 2.

Conclusion:

Combined with CTX treatment had better effect and lower relapse rate than prednisone monotherapy. The patients with more than 6 organs involved should accept immunosuppressants therapy at beginning. CTX decreased organs relapse greatly; immunosuppressants were efficacious for most relapsing patients treated with glucocorticoid alone.

