W29: Long term follow-up of Waldenström Macroglobulinemia patients primary treated with Dexamethasone, Rituximab, and Cyclophosphamide : a single centre experience

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Background

Waldenström's macroglobulinemia (WM) is a rare indolent B-cell lymphoproliferative disorder resulting from the accumulation of monoclonal lymphoplasmacytic cells that secret a monoclonal IgM protein. Current guidelines recommend to treat WM patients not based on the concentration of the paraprotein, but on the presence of signs and/or symptoms of disease. Although the rarity of this disease and the paucity of randomized clinical trials, a precise algorithm of therapy is missing and immunochemotherapy is considered a standard of care for WM patients. The Eighth International Workshop on WM consider DRC regimen (dexamethasone, rituximab, and oral cyclophosphamide) a suitable first line treatment because it's safe, manageable and exerts a high rate of responses, at least a partial response in 30% to 50% of patients.

Aim

To date, only two studies describe the use of DRC regimen in previously untreated and in relapsed WD patients (DIMOPOULOS, JCO 2007; PALUDO, BJH 2017). We therefore described 20 previously untreated symptomatic patients with WM who received DRC outside of clinical trials to provide further insights on efficacy, tolerability and safety of this regimen after a long term follow-up.

Patients and Methods

20 patients were treated with dexamethasone 20 mg intravenously followed by rituximab 375 mg/m^2 intravenously on day 1 and cyclophosphamide 100 mg/m^2 orally bid on days 1 to 5 (total dose, 1.000 mg/m^2). This regimen was repeated every 21/28 (depending on frailty of patients) days for 6 cycles. Rituximab 375 mg/m^2 intravenously was administered for 2 more cycle as maintenance. Several patients had features of advanced disease such as anemia (75%), hypoalbuminemia (30%), and elevated serum beta2-microglobulin (45%). They were 6 (30%) low, 8 (40%) intermediate and 6 (30%) high risk patients according to International Prognostic Macroglobulinemia Scoring System for Waldenstrom Macroglobulinemia (GERTZ, AJH 2017).

Results

Mean age of patients when they started DRC was 67 years old (range 46-82). On an intent-to-treat basis, 85% of patients achieved a response, including 5% complete, 75% partial responses; 5% of patients had a stable disease and 10% a progressive disease (Figure 1). The 4-year progression-free survival rate for all patients was 56%; for patients who responded to DRC, it was 67%. The 4-year disease-specific survival rate was 74% (Figure 2 and 3).

Conclusion

Our data showed that DRC regimen is an active, well-tolerated treatment for symptomatic patients with WM. Long term follow up of patients treated with DRC showed a good OS.

Figure 1: response to treatment.

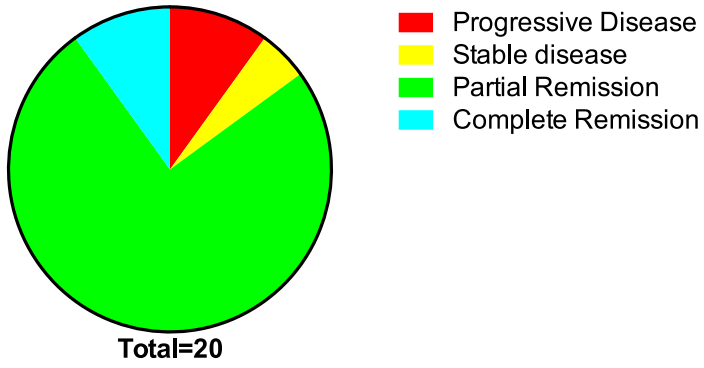

Figure 2: PFS

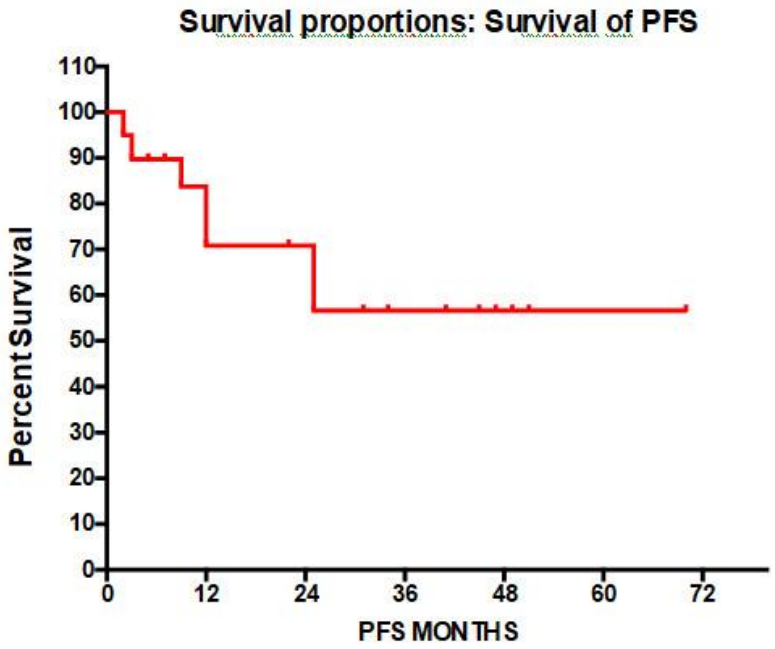

Figure 3: OS