

Light chain amyloidosis associated with Waldenström macroglobulinemia: treatment and survival outcomes

Joshua N. Gustine, Raphael E. Szalat, Andrew Staron, Tracy Joshi, Lisa Mendelson, J. Mark Sloan and Vaishali Sanchorawala

Amyloidosis Center and Section of Hematology and Medical Oncology, Boston University School of Medicine and Boston Medical Center, Boston, MA, USA

Correspondence: V. SANCHORAWALA - vaishali.sanchorawala@bmc.org

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SUPPLEMENTAL APPENDIX

Table S1. Hazard regression analysis for event-free survival and overall survival to frontline treatment regimen in patients with WM-AL amyloidosis.

Event-Free Survival	Univariate		Multivariate	
Variables	HR (95% CI)	P-value	HR (95% CI)	P-value
Age >65 years	0.81 (0.33-1.96)	0.64		
Male sex	1.43 (0.59-3.45)	0.43		
Lambda light chain isotype	1.21 (0.48-3.05)	0.67		
Hemoglobin ≤11.5 g/dL	1.09 (0.41-2.89)	0.86		
Platelet count ≤100 K/uL	UTC	UTC		
Beta2-microglobulin >3 mg/L	1.97 (0.74-5.23)	0.18		
Serum IgM >4000 mg/dL	0.33 (0.04-2.49)	0.28		
dFLC >180 mg/L	1.88 (0.68-5.23)	0.23		
BM involvement >10% by LPL	0.42 (0.15-1.18)	0.10		
MYD88 mutation	0.81 (0.08-7.84)	0.86		
Serum creatinine >2.0 mg/dL	4.20 (1.52-11.7)	0.006		
Urine protein >5000 mg/24hr	1.50 (0.61-3.73)	0.39		
ALP >150 IU/L	1.14 (0.33-3.94)	0.84		
BNP >81 pg/mL	1.56 (0.63-3.85)	0.33		
Troponin I >0.1 ng/mL	2.10 (0.47-9.29)	0.33		
Previously treated	0.91 (0.26-3.12)	0.88		
Overall Survival	Univariate		Multivariate	
Variables	HR (95% CI)	P-value	HR (95% CI)	P-value
Age >65 years	1.07 (0.44-2.59)	0.88		
Male sex	2.27 (0.90-5.72)	0.08		
Lambda light chain isotype	1.75 (0.66-4.52)	0.25		
Hemoglobin ≤11.5 g/dL	1.08 (0.41-2.83)	0.88		
Platelet count ≤100 K/uL	UTC	UTC		
Beta2-microglobulin >3 mg/L	2.58 (0.98-6.78)	0.06		
Serum IgM >4000 mg/dL	0.46 (0.06-3.45)	0.45		
dFLC >180 mg/L	2.06 (0.78-5.43)	0.15		
BM involvement >10% by LPL	0.66 (0.22-2.01)	0.47		
MYD88 mutation	0.51 (0.08-3.14)	0.47		
Serum creatinine >2.0 mg/dL	4.46 (1.63-12.2)	0.004	3.91 (1.29-11.8)	0.02
Urine protein >5000 mg/24hr	0.94 (0.39-2.30)	0.90		
ALP >150 IU/L	3.60 (1.19-10.9)	0.02	2.59 (0.79-8.48)	0.11
BNP >81 pg/mL	2.47 (1.03-5.94)	0.04	2.31 (0.93-5.77)	0.07
Troponin I >0.1 ng/mL	2.16 (0.62-7.53)	0.23		
Previously treated	1.23 (0.42-3.93)	0.65		

Previously treated patients (n=8) received a WM-directed therapy before the diagnosis of AL amyloidosis. dFLC, difference between the involved and uninvolved serum free light chain; BM, bone marrow; LPL, lymphoplasmacytic lymphoma; ALP, alkaline phosphatase; BNP, brain natriuretic peptide; UTC, unable to calculate; OR, odds ratio; CI, confidence interval.

Table S2. Salvage treatment regimens utilized in patients with WM-AL amyloidosis.

Treatment Regimen	Number of Patients
BDR	4 (36%)
Benda-R	3 (27%)
CyBorD-R	2 (18%)
Ibrutinib	2 (18%)
CaRD	1 (9%)
Idelalisib	1 (9%)
Ven-O	1 (9%)

Eleven patients with WM-AL amyloidosis received salvage therapy. BDR, bortezomib, dexamethasone, and rituximab; Benda-R, bendamustine and rituximab; CyBorD-R, cyclophosphamide, bortezomib, dexamethasone, and rituximab; CaRD, carfilzomib, rituximab, and dexamethasone; Ven-O, venetoclax and obinutuzumab.

Figure S1. Survival in patients with WM-AL amyloidosis. Kaplan-Meier survival curves for event-free survival (EFS) and overall survival (OS) stratified by serum creatinine (A-B); OS stratified by BNP (C); OS stratified by BU cardiac staging system (D); and EFS and OS stratified by maintenance rituximab among patients who achieved a partial response or better to a rituximab-containing frontline regimen (E-F). BNP, brain natriuretic peptide.

