Treatment Options for Amyloid related Disease in WM

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IgM Amyloidosis Immunoglobulin M associated AL is a rare but distinct entity. It occurs in 4.7% of patients with AL. Previous reports suggest that lymph node involvement is more common and urinary protein loss is less prevalent. Among 434 AL patients, 5% or 22 had an IgM monoclonal protein. The 5% with IgM amyloidosis compares with 1% IgM myeloma and 15% with IgM monoclonal gammopathy of undetermined significance. Eleven of the AL patients had IgM λ monoclonal proteins, 5 had IgM κ monoclonal proteins, 2 had an IgM κ and λ, 2 had biclonal IgM- κ/IgG- λ, 1 had biclonal IgM- λ /IgA- λ, and 1 had biclonal IgM- λ /IgG- λ. The patients with IgM amyloidosis were a median of six years older than non-IgM patients ($p < 0.001$) and had a 32% incidence of neuropathy compared with non-IgM 11% ($p < 0.01$). Patients with IgM amyloidosis had a statistically significantly lower troponin T ($p < 0.03$), NT-Pro BNP ($p < 0.01$), and intraventricular septal thickness ($p = 0.07$). The survival of patients with IgM amyloidosis was 77.6 months, and this is not different from those with non-IgM amyloidosis whose median overall survival is 96.5 months ($p = 0.3$). There continues to be a λ preponderance in IgM amyloidosis. We speculate that IgM amyloid proteins have a greater affinity for peripheral nervous system tissues and a lower affinity for cardiac tissues. Response analysis showed no difference in response rates to high-dose therapy, 86 versus 75% ($p = 0.27$). Patients with IgM amyloidosis tended to have a lower level of the involved free light chain compared with those with non-IgM amyloidosis, 4.1 versus 15.5 mg/dL ($p = 0.0002$) suggesting IgM-associated amyloidosis has a lower number of circulating unbound light chains.