Amyloidosis with IgM Monoclonal Gammopathies

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Purpose:

To review outcomes of patients who have immunoglobulin light chain amyloidosis of the IgM type.

Patients and Methods:

Fifty patients with a serum IgM monoclonal protein and biopsy-proven amyloid were evaluated.

Results:

The percentages of patients presenting with cardiac, renal (2), hepatic, and pulmonary amyloid were 44%, 32%, 14%, and 10%, respectively. Forty-two percent of the patients had an M-protein spike in the serum greater than 1.5 gm/dL, and 12% had an IgM peak greater than 3 gm/dL. Amyloidosis was easily diagnosed using either biopsy of the subcutaneous fat, rectum, and bone marrow demonstrating deposits in 84%, 72%, and 50% respectively. The median survival of all patients was 24.6 months. Fifty-three percent of all deaths were due to cardiac amyloidosis. Twelve percent succumbed to respiratory failure, and 7% each succumbed to macroglobulinemia, hepatic failure, and renal failure.

Conclusion:

The presence of amyloid cardiomyopathy and an elevated creatinine had the greatest impact on survival. Of the 22 patients with amyloid cardiomyopathy, the median survival was 11.1 months, and less than 10% survived five years. Of the 28 patients without amyloid cardiomyopathy at the time of diagnosis, the median survival was 27 months with nearly 30% surviving five years.