Management of Plasma Cell Disorders

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INTRODUCTION

The plasma cell disorders are a spectrum of conditions that include asymptomatic precursor conditions—monoclonal gammopathy of undetermined significance (MGUS) and smoldering multiple myeloma (SMM)—as well as symptomatic multiple myeloma (MM) and solitary plasmacytoma. Other plasma cell disorders include immunoglobulin light chain amyloidosis and POEMS syndrome, which are characterized by a unique set of end-organ manifestations. There are other related plasma cell and B-cell proliferations, such as light chain deposition disease and cryoglobulinemia, that are beyond the scope of this review but are relevant to the hematologist/oncologist and have been reviewed in detail elsewhere.

MM is the second most common hematologic malignancy, with approximately 20,000 patients diagnosed annually in the United States. The median age at presentation is 72 years, and it is more common in men and in African Americans. In fact, MM is the most common hematologic malignancy in African Americans.

The precursor states MGUS and SMM are asymptomatic without end-organ manifestations. MM is characterized by the accumulation of clonal bone marrow plasma cells and production of monoclonal immunoglobulins leading to the cardinal end-organ manifestations known by the acronym CRAB: hypercalcemia, renal failure, anemia, and bone disease. Hypercalcemia occurs as a result of bone destruction from increased osteoclast activity stimulated by malignant plasma cells. Renal failure may be associated with hypercalcemia, light chain cast nephropathy, or light chain deposition. Anemia is generally due to the expansion of plasma cells in the bone marrow, which may also lead to leukopenia and thrombocytopenia. Myeloma-related bone disease includes osteoporosis, osteolytic bone lesions, fractures, and bone pain. Additional, less common manifestations of symptomatic MM include hypogammaglobulinemia with frequent infections, susceptibility to bleeding, plasmacytomas either extending from bone or in soft tissue sites, and amyloidosis.

Immunoglobulin light chain amyloidosis (AL) may be a primary disorder or may be seen in association with frank MM. Features of amyloidosis are rare in newly diagnosed MM, but may occur in up to 10% of patients in the relapsed and refrac-