Extramedullary disease in plasma cell disorders can occur as a solitary plasma cell tumor of the bone, in the absence of systemic features of multiple myeloma such as anemia, hypercalcemia, renal insufficiency, or multiple lytic bone lesions. The primary treatment for patients with SPB is similar to that for SEP—localized radiation therapy at curative doses of 40 to 50 Gy. Surgery may be required if there is structural instability of the bone or rapidly progressive symptoms from cord compression. The local response rate to RT is about 90%. The median overall survival (OS) of patients with SPB is approximately 10 years, but with 85% of patients experiencing disease progression at 10 years.

Multiple myeloma with extramedullary plasmacytomas (MM/EP): As opposed to SPB and SEP, the outcomes of patients with extramedullary plasmacytoma involving multiple sites are dismal, with OS of only about 15 months. Some regularly involved sites are liver, gastrointestinal tract, lung, pleura, lymph nodes, skin, and central nervous system (CNS). Patients with CNS involvement rarely live past five months. In a recent review by Varettoni et al, the overall incidence of extramedullary disease was noted to be about 13% in MM, with 7% at diagnosis and 6% at relapse. Extramedullary disease was associated with shorter overall survival (hazard ratio [HR] 3.26, P < .0001) and progression-free survival (HR 1.46, P = .04).

In SEP and SPB, if the first-line therapy of radiation or surgery does not achieve complete remission (CR), then chemotherapy regimens commonly used for treatment of multiple myeloma should be used as second-line therapy. After attaining CR, it is necessary to continue monitoring for local as well as systemic evidence of progression with history and physical, complete blood count, serum creatinine, serum calcium, beta 2-microglobulin, albumin, lactate dehydrogenase, and serum and urine protein electrophoresis with immunofixation. Initially, monitoring should be monthly and then every three months for two to three years, followed by six monthly evaluations for another three years and then annually afterwards. Bone marrow exam and myeloma survey should be considered once a year. It is also useful to do PET-CT and MRI every 6 to 12 months. Investigations aimed at evaluating local recurrence, such as fiberoptic endoscopy for patients with head and neck involvement and cystoscopy for bladder involvement, should be performed at regular intervals and upon onset of relevant symptoms. The pathogenesis of SEP, SPB, MM/EPP is poorly understood and it's likely that adhesion factors on the plasma cell surface play a significant role in the evolution of these entities. Genomic and proteomic studies may help us in better understanding the mechanism behind extramedullary manifestations of myeloma.

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