其他單株免疫球蛋白增高血症的治療

Treatment for other paraproteinemia

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A paraproteinemia is a monoclonal immunoglobulin or light chain present in the blood produced by a clonal population of plasma cells. Except for plasma cell myeloma, there are several disorders that also induce paraproteinemia, such as amyloidosis, lymphoproliferative neoplasm (Waldenstrom macroglobulinemia), and POEMS syndrome (Polyneuropathy, organomegaly, endocrinopathy, M protein, skin changes). Most current strategies including systemic therapy to destroy the plasma cells responsible for the synthesis of paraproteinemia are derived from the treatment of multiple myeloma, such as proteasome inhibitors, Immunomodulatory drugs (IMiDs), anti-CD38 monoclonal antibodies, prednisolone, and chemotherapy. Besides, autologous stem cell rescue is also a treatment option for patients eligible for high-dose therapy. However, there are still some differences in the treatment strategies owing to distinct underlying pathophysiology between plasma cell myeloma and other paraproteinemia disorder. Taking amyloidosis for example, it is typically characterized by decreased numbers of monoclonal plasma cells in the bone marrow compared to plasma cell myeloma, but the protein produced by these plasma cells has an affinity for visceral organs (such as heart, kidney, liver, and spleen) inducing related end-organ dysfunction. As a result, though patients with this disease typically have a low burden of monoclonal plasma cells their survival is often poor due to the end organ damage by the amyloid protein. The goals of therapy include eliminating the misfolded amyloid light chains as promptly as possible, minimizing treatment toxicity, and supporting the function of the damaged organs.