similar to therapy for myeloma. The combination of bortezomib, cyclophosphamide, and dexamethasone is effective in some patients. Selected patients may respond well to high-dose chemotherapy and autologous stem cell support, but there are increased risks of morbidity and mortality if significant end-organ dysfunction such as cardiomyopathy occurs.

POEMS syndrome is a rare disorder characterized by polyneuropathy, sclerotic bone lesions, endocrinopathy, monoclonal gammopathy, and skin lesions. The cause of POEMS syndrome is unknown, but the disease may be progressive, causing severe disability, third spacing of fluid, and elevated vascular endothelial growth factor (VEGF) levels. Monoclonal λ light chains are typically elevated. Limited bone disease may be treated with radiotherapy. High-dose therapy and autologous stem cell transplantation is effective in patients with extensive disease.

For a deeper discussion of these topics, please see Chapter 187, "Plasma Cell Disorders," and Chapter 188, "Amyloidosis," in Goldman-Cecil Medicine, *25th Edition.*

CONGENITAL AND ACQUIRED DISORDERS OF LYMPHOCYTE FUNCTION

Several congenital disorders affect lymphocyte maturation or function, resulting in immunodeficiency disorders. Acquired disorders of lymphocyte function are far more common than congenital disorders. HIV infection is the most important infectious cause of acquired immunodeficiency (see Chapter 109). Patients with HIV infection are at increased risk for NHL. NHLs that occur in the setting of HIV have the diffuse, aggressive B-cell histology and include DLBCL and Burkitt's lymphoma, and they are frequently associated with EBV infection and are often advanced stage (III or IV) at diagnosis, with extranodal sites of involvement.

Patients with HIV-associated NHL are potentially curable with the multidrug chemotherapy regimens used for treating the NHL subtypes found in the general population. Treatment of the underlying HIV infection with highly active antiretroviral therapy (HAART) has improved the outcome and prognosis of patients with HIV-associated NHL.

Patients who have undergone allogeneic organ transplantation require potent immunosuppressive drugs (e.g., cyclosporine, tacrolimus, mycophenolate, corticosteroids, methotrexate) to prevent graft-versus-host disease in the case of bone marrow transplantation or allograft rejection in the case of solid organ transplantation. These medications can cause profound defects in T-cell function with an associated acquired immunodeficiency state, and transplant recipients are susceptible to a host of viral and protozoal infections.

Patients who receive potent immunosuppressive drugs are at risk for a lymphoproliferative disorder (i.e., post-transplantation lymphoproliferative disorder [PTLD]) that can behave clinically as an aggressive lymphoma. PTLD is an EBV-associated lymphoproliferative disorder characterized by a polymorphous or monomorphous population of B cells that can be monoclonal or polyclonal. Patients who develop PTLD are treated by reducing the doses of immunosuppressive drugs whenever possible. Patients with polymorphous disease early after organ transplantation may respond well to this approach. Patients who are not candidates for withdrawal of immunosuppression because of allograft rejection or who develop late monophorphic disease may respond better to treatment with rituximab alone or in combination with chemotherapy.

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