

negative for CD10 and largely negative for CD5. Some cases can have plasmacytoid differentiation and can be associated with a monoclonal expression of kappa or lambda light chains and with small monoclonal immunoglobulin spikes. Treatment is often similar to that of FL, with the exception that the BTK inhibitor ibrutinib is highly active in this disease, while largely disappointing in FL, and is a good treatment option for relapsed nodal MZL as well as other MZL subtypes.

Splenic MZL is largely a disease of older Caucasian patients; infection with hepatitis C is a risk factor for this disease, and treatment of hepatitis C can result in regression of the lymphoma. Patients present with a lymphocytosis with or without cytopenias and splenomegaly. Bone marrow involvement is common. Diagnosis can be made by flow cytometry of the peripheral blood; malignant lymphocytes will be positive for surface immunoglobulin, CD19, and CD20 and will generally lack CD5 and CD10. On peripheral smear, they have small nuclei and abundant cytoplasm with “shaggy” or villous projections. It can be differentiated from HCL by the absence of CD25, CD103, and annexin A1. Recurrent cytogenetic abnormalities include trisomy 3 and abnormalities of chromosome 7q. Therapy is indicated for symptomatic disease or significant cytopenias. Splenectomy is reasonable for selected patients with excellent relief of symptoms and cytopenias. Splenectomy is associated with an overall response rate of 85% and estimated progression-free survival and OS rates at 5 years of 58 and 77%, respectively. Single-agent rituximab can improve splenomegaly and cytopenias in >90% of patients. In a study of induction with weekly rituximab followed by maintenance, the response rate was 95%, with overall and progression-free survival rates at 5 years of 92 and 73%, respectively. Other options for therapy at relapse are similar to those used for FL and include retreatment with rituximab, alkylating agents, and purine analogues in combination with rituximab. The survival rate of patients is in excess of 70% at 10 years.

MALT lymphoma is an MZL lymphoma of extranodal tissue, most commonly the stomach, but other common sites include the skin, salivary glands, lung, small bowel, ocular adnexa, breasts, bladder, thyroid, dura, and synovium. It is associated with states of chronic