SPLENIC MARGINAL ZONE NHL.

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SMZL is a distinctive form of indolent lymphoma originating in the spleen, characterized by prominent splenomegaly and variable involvement of lymph nodes, bone marrow, peripheral blood, and other organs. The WHO definition is a B-cell neoplasm .It is an uncommon form of NHL, accounting for less than 1% of new cases. The immunophenotype of SMZL is similar to NMZL and MALT lymphoma Further evidence supporting SMZL as a distinct entity is a unique gene expression profile when compared to other indolent B-cell lymphomas. Association with hepatitis C infection has been reported.Patients often have modest cytopenias that are primarily due to splenic sequestration with a smaller contribution from marrow infiltration. Usually a bone marrow biopsy is the best initial diagnostic test and will often establish the diagnosis. Occasionally the distinction with other lymphoproliferative disorders, such as hairy cell leukemia (HCL), can be challenging. Flow cytometry of circulating lymphoma cells or the marrow can be helpful as SMZL is typically CD25 negative and CD103 negative, unlike HCL. If no blood or marrow involvement is present, the diagnosis is best established by splenectomy. The prognosis is usually good after a diagnosis of SMZL. Splenectomy is usually the treatment of choice for SMZL. In the absence of comparative trials, it is difficult to know if any particular regimens should be preferred, but it appears as though alkylating agents may be less active than purine analogues, and amongst the purine analogues, 2-chlorodeoxyadenosine may be less active than fludarabine and pentostatin. Interestingly, single-agent rituximab has been reported to be highly active in SMZL with an ORR of 100% and a CR rate of 71%.