

Abstract: Splenic marginal zone lymphoma (SMZL) is a subtype of non-Hodgkin lymphoma (NHL) which constitutes <1% of all NHL and up to 25% of low grade B cell neoplasms in histology of splenectomy specimens. Most patients are asymptomatic at diagnosis while others may present with splenomegaly and cytopenia (anemia and thrombocytopenia) with associated lymphocytosis. Cytopenias may be secondary to hypersplenism, bone marrow involvement or immune mediated. Autoimmune manifestations like autoimmune hemolytic anemia can occur over the course of disease, but it is rare on presentation.

Here we present a case of an elderly female who presented with fever, constitutional symptoms, isolated splenomegaly and autoimmune hemolytic anemia with normal bone marrow biopsy was found to have splenic marginal zone lymphoma on histology of splenectomy sample. Even though clinical symptoms are typical these patients constitute a diagnostic challenge as the investigations are inconclusive until last option of splenectomy.