



Marginal zone lymphoma – A rare finding on peripheral blood smear

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ABSTRACT

Splenic marginal zone lymphoma is a rare low grade B cell lymphoma arising from post-germinal center marginal zone B cells. Comprising less than 2% of the lymphoid neoplasm; affects mainly middle-aged and elderly patients. We report a case of 58 years old female with the history of fever, malaise, and abdominal discomfort. Laboratory evaluation revealed lymphocytosis with the presence of atypical lymphocytes having villous cytoplasmic projections and basophilic cytoplasm. Immunophenotyping analysis revealed IgM positive, CD 5 negative, CD10 negative. CD23 negative and cyclin D negative.

Keywords: Marginal zone lymphoma, Peripheral blood smear.

1. INTRODUCTION

The term splenic marginal zone lymphoma (SMZL) was first coined by Schmid and colleagues in 1992, as a rare indolent non-Hodgkin lymphoma (NHL) subtype originating from B memory lymphocytes present in the marginal zone of secondary lymphoid follicles.^{1,2,3} It is a rare disorder comprising of 0.9% of all Non Hodgkin's lymphoma(NHL).^{2,4,5}

SMZL is a B cell neoplasm of small lymphocytes that replace splenic white pulp germinal center resulting in effacement of follicle mantle zone and merge with the peripheral marginal zone. It involves splenic hilar lymph nodes and bone marrow. The peripheral blood involvement is seen in the form of villous lymphocytes. (Image -1)

It affects mainly elderly people over 50 years of age with equal sex incidence.⁶ although it is a rare indolent lymphoma it accounts for most of the cases of unclassified chronic lymphoid leukemias that are CD-5 negative. The tumor most commonly involves spleen, lymph nodes, bone marrow and often peripheral blood. Peripheral lymph nodes are not typically involved. The patients present with splenomegaly and occasionally with autoimmune thrombocytopenia or anemia. Cytopenias are uncommon as it is due to hypersplenism, and less frequently to auto-antibodies or bone marrow infiltration.^{3,7}

2. CASE REPORT

We report a case of 58 years old female with the history of fever, malaise and abdominal discomfort since last 15 days. On physical examination splenomegaly was noted which was confirmed by computerized tomography. No lymphadenopathy was observed. Complete blood count reveals total leucocytes count 12.29 x 10⁹ cells/microlitre, absolute neutrophils -3.42 cells/microlitre, absolute lymphocytes -8.64 cells/microlitre, eosinophils -0.50 cells/microlitre, monocytes- 0.17 cells/microlitre, basophils- 0.01 cells/microlitre, hemoglobin 12.4 gm%, platelets 1.53 lakh/ microlitre of blood. Reticulocyte count was 0.5%, vitamin B12-332, folate >20. Peripheral blood smear shows red blood cell normocytic normochromic, white blood cells shows total leucocytosis with absolute lymphocytosis with the presence of small to middle-sized basophilic lymphocytes along with few lymphocytes which were having villous cytoplasmic projection. [figure-1] platelets were adequate in number. With the view of this peripheral picture, we advised the patient to go for bone marrow examination and immunophenotyping to rule out the possibility of CLL/MCL/HAIRY CELL leukemia. The patient refused to go for bone marrow procedure. Peripheral blood was sent for immunophenotyping. The immunophenotyping shows CD3, CD5, CD7, CD10, CD25, and CD 23, CD 38 and bcl2 and IgM lambda negative. CD19, CD20, CD79b, CD 45, FMC-7, CD 11 C, IgM, kappa was positive.

3. DISCUSSION

Marginal zone lymphomas represent a group of lymphoma that arises from post-germinal center marginal zone B cells. It includes three specific entities according to the organ affected as extranodal marginal zone lymphoma (EMZL) or mucosa associated lymphoid tissue (MALT) lymphoma, Splenic marginal zone lymphoma (SMZL), nodal marginal zone lymphoma. (NMZL). EMZL is the most common entity, this neoplasm at virtually any extranodal site and commonly associated with chronic antigenic stimulation of either infection e.g. H.pylori in the stomach or autoimmune disorder as Sjogrens syndrome and salivary gland. Patients with NMZL have lymph nodes disease without the involvement of spleen or extranodal sites. SMZL is usually diagnosed incidentally either due to splenomegaly or cytopenias or peripheral blood lymphocytosis with compatible phenotyping.⁸

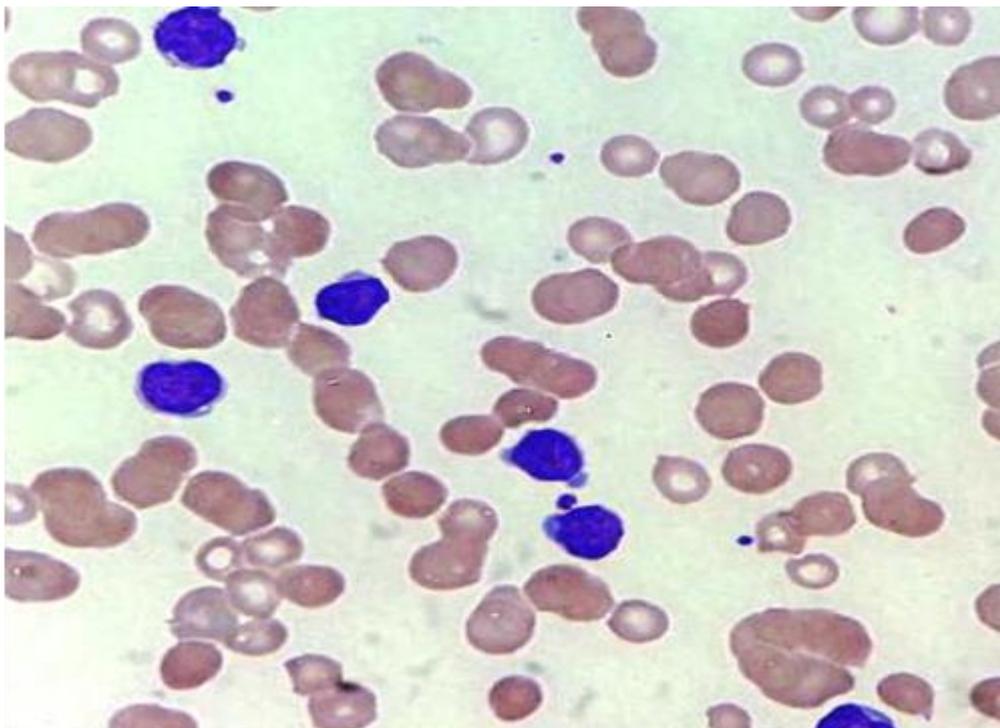
SMZL has splenic involvement with IgM, CD 5 and CD10 positivity. It mainly affects the elderly people over the age of 50. Both male and females are equally affected. Immunophenotyping in our case shows CD 5, CD 23 negative which excludes chronic lymphocytic leukemia, CD 5 and cyclin D1 negative which excludes the possibility of mantle cell lymphoma, CD10 negative helps us to exclude follicular lymphoma ; CD -103 negative rules out hairy cell leukemia.

Antigen	Status in MZL
CD20	Positive
CD79b	Positive
CD5	Negative
CD10	Negative
CD23	Negative
CD25	Negative
cyclin D1 CD 103	Negative

Thus the relevant markers that define the immunophenotype for SMZL are shown in the table shown above. The lack of CD5 expression is helpful in the discrimination between SMZL and chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL), and the lack of CD10 expression argues against follicular lymphoma (FL). Mantle cell lymphoma (MCL) is excluded due to the lack of CD5 and cyclin-D1 expression.

4. CONCLUSION

Splenic marginal zone lymphoma is a difficult entity to diagnose especially on peripheral blood smear as in this case due to the presence of villous lymphocytes we reach to various differential diagnosis otherwise only we can make out the diagnosis of CLL on peripheral film with lymphocytosis history of an elderly person. Keeping in view of this case it is very necessary to see the peripheral blood film with a proper clinical history of a patient to make an early right platform of final diagnosis.



Peripheral blood smear Leishman stain (100 x)

5. REFERENCES

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