IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 9 Ver. XI (September). 2016), PP 38-40
www.iosrjournals.org

Splenic Marginal Zone Lymphoma: A Case Report with Review of Literature
Running Title: A Rare Case Report On Splenic Marginal Zone Lymphoma

Dr. Iffat Jamal[1], MD Pathology, Dr. Suryakant Nirala[1], MD Pathology Tutor,
All India Institute of Medical Sciences, Patna

Abstract:
Splenic marginal zone lymphoma(SMZL) is a rare B-cell neoplasm comprising less than 2% of lymphoid neoplasms ,composed of small lymphocytes which surround and replace the splenic white pulp ,germinal centres,follicle mantle and merge with marginal zone.We report a case of SMZL in a 40 year old male patient presenting with complaints of low grade fever and left sided abdominal discomfort for 6 months and was noted to have massive splenomegaly on physical examination. Laboratory evaluation revealed leucocytosis and presence of atypical lymphocytes which were larger than mature lymphocytes having moderately basophilic cytoplasm with cytoplasmic processes at the poles of the cell with round to oval nucleus with clumped chromatin and indistinct nucleoli.Immunophenotypic analysis revealed a B-cell population with CD -20 positivity.

Keywords: *Neoplasm,splenic,white pulp,germinal centre,splenomegaly*.

I. Introduction

Splenic marginal zone lymphoma(SMZL) is a rare primary splenic lymphoproliferative disorder that affects mainly older individuals belonging to age group of 40-60 years with male preponderance.It comprises less than 2% of lymphoid neoplasms.Patients present with moderate to massive splenomegaly .Bone marrow is frequently involved but peripheral lymphadenopathy is extremely uncommon.Patients have moderate leucocytosis with peripheral smear demonstrating the characteristic atypical lymphocyte with short cytoplasmic villous processes at their poles.[1,2]

II. Case report

We report a case of SMZL in a 40 year old male patient presenting with complaints of low grade fever and left sided abdominal discomfort for 6 months and was noted to have massive splenomegaly on physical examination.Complete blood count revealed moderate leucocytosis with Haemoglobin level of 10.0 g/dl,hematocrit ,29.3%,White blood cell count 20,000 /µl and platelet count was slightly reduced,1lakh/µl .The white blood cell differential was 49% neutrophils,44% lymphocytes ,5% monocytes,1% eosinophils and 1% basophils.Peripheral smear revealed presence of atypical lymphocytes which were larger than mature lymphocytes having moderately basophilic cytoplasm with cytoplasmic processes at the poles of the cell with round to oval nucleus with clumped chromatin and indistinct nucleoli. These cells with polar processes display fine granular positivity with Periodic Acid Schiff stain (PAS)(Figure 1 & 2). Immunophenotypic analysis revealed a B-cell population with CD -20 positivity. Bone marrow aspiration and biopsy was performed which showed nodular deposits of lymphoma cells .(Figure 3).Computed tomography of the abdomen was significant for massive splenomegaly.

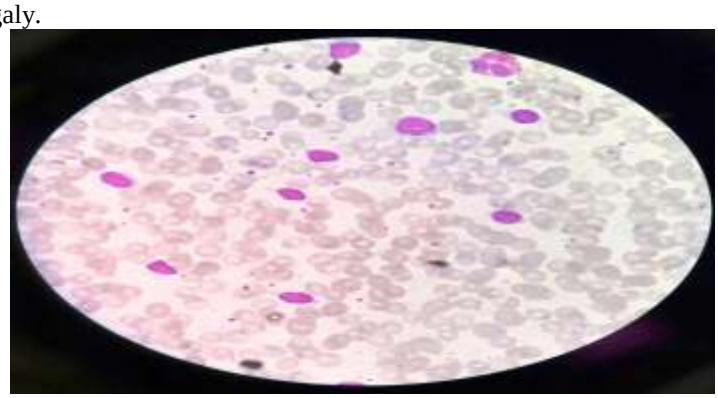

Figure:1 – Peripheral blood smear showing atypical lymphocytes with polar cytoplasmic processes, a characteristic feature of SMZL.(MGG,1000X)

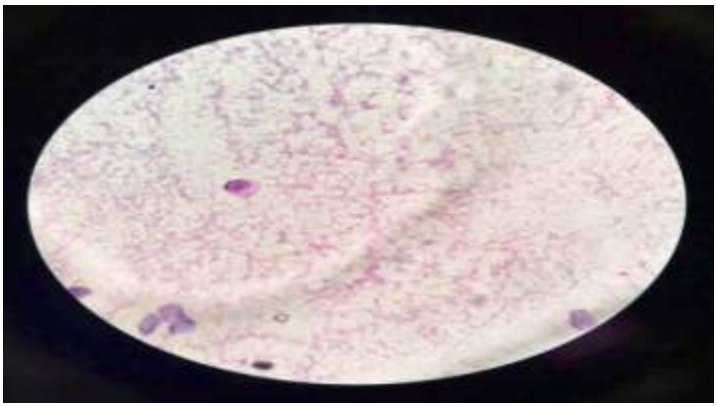

Figure 2: Photomicrograph showing granular PAS (Periodic Acid Schiff) positivity in villous lymphocytes in SMZL(PAS,1000X)

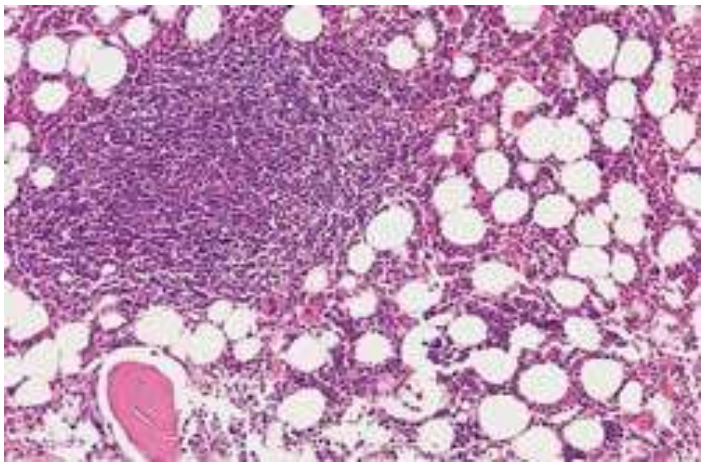

Figure 3: Photomicrograph showing Bone marrow infiltration by atypical lymphoid cells in SMZL(MGG,400X)

III. Discussion

SMZL is an uncommon B- cell neoplasm accounting for less than 2% of all Non-Hodgkin's lymphomas and only 8.3% of all Non-Hodgkin's Lymphoproliferative disorders involving spleen.[3] Patients with this disease typically are older men with widespread disease at the time of diagnosis.It has an indolent course with an average long term survival rate (10-15 years) without any treatment.[4]

The tumour involves spleen and splenic hilar lymph nodes .Bone marrow and peripheral blood are often involved .Peripheral lymph nodes are not frequently involved and extra nodal infiltration is quite uncommon.[5]

In the splenic white pulp ,a central zone of round lymphocytes surrounds or more commonly replaces reactive germinal centres with effacement of normal follicle mantle.This zone merges with the peripheral zone of small to medium sized cells with more dispersed chromatin and abundant pale cytoplasm .The red pulp is always infiltrated with both small nodules of larger cells and sheets of small lymphocytes which often invade sinuses.[5,6]

In Peripheral blood the characteristic atypical lymphocytes which are larger than mature lymphocytes are seen having moderately basophilic cytoplasm with cytoplasmic processes at the poles of the cell in the direction in which smears are made.These cells have round to oval nucleus with clumped chromatin and indistinct nucleoli .Some may appear plasmacytoid.The differential diagnosis includes other small B-cell lymphomas/leukemias including Chronic lymphocytic leukemia,hairy cell leukemia,mantle cell lymphoma,follicular lymphoma and lymphoplasmacytic lymphoma.[7]

Immunophenotypic analysis reveals tumour cells to be positive for CD20,CD79a, and negative for CD5,CD10,CD103,Annexin A1,and CyclinD1.

Cytogenetic study shows allelic loss of chromosome 7q31-32 in upto 40% cases of SMZL.Dysregulation of CDK6 gene located at 7q20 has been reported in several cases.[5,7]

The clinical course is indolent even with bone marrow involvement with an average survival rate of 10-15 years without any therapy. Surgical therapy with splenectomy appears to play an important role in the

treatment of some patients, with a resultant clinical response in most cases. Initial treatment with chemotherapy may not be beneficial and was associated with reduced overall survival in one study.[7,8]

However, in patients with clinical progression (ie, lymphocytosis), alkylating agents may produce a favorable response. Newer therapies using the purine analogs, specifically fludarabine phosphate, have shown some promise. Reported complete remissions have been seen in a number of patients treated with fludarabine phosphate who had a history of disease relapse or failed to respond to primary therapy.

Unfortunately, well-defined treatment strategies in SMZL and other indolent lymphomas have not been described. Coiffier et al reported that in older patients without adverse prognostic parameters, no therapy or splenectomy alone was associated with a long event-free survival.[9] Additionally, they stated that younger patients, patients with a high component of large cells, or those with adverse prognostic parameters probably have to be treated with a combination regimen, although they admitted that more prospective trials need to be undertaken to define the best options. If patients have signs of progression, high-dose chemotherapy may be used. Selected patients with SMZL have an aggressive course with death occurring within a year of diagnosis, despite surgical and chemotherapeutic intervention. Some authors have suggested that peripheral blood lymphocytosis and/ or extent of bone marrow involvement at presentation predict a more aggressive clinical course.

Additionally, a study by Lloret et al.[10] suggested that an increased number of large transformed cells in an otherwise typical SMZL predicts a more aggressive clinical behavior. This finding was not seen in our patient. Patient selection appears critical in SMZL, since some patients have long-term survival, while others have early progression. Unfortunately, for patients with recurrent disease, bone marrow transplantation may offer the only chance for cure. Newer agents, including the anti-CD20 monoclonal antibody (rituximab) may also offer hope in refractory disease. Future investigation is needed to better define clinical and morphologic characteristics that may predict disease outcome and thereby selecting those patients who need initial adjunct therapy to prevent recurrence and disease-related death.[9,10]

References

[1]. Harris NL, Jaffe ES, Stein H, et al. A revised European-American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group. Blood. 1994;84:1361–1392.
[2]. Troussand X, Valensi F, Duchayne E, et al. Splenic lymphoma with villous lymphocytes: clinical presentation, biology and prognostic factors in a series of 100 patients. Br J Haematol. 1996;93:731–736.
[3]. Brox A, Bishinsky JI, Berry G. Preliminary non-Hodgkin lymphoma of the spleen. Am J Hematol. 1991;38:95–100.
[4]. Falk S, Stutte HJ. Primary malignant lymphomas of the spleen: a morphologic and immunohistochemical analysis of 17 cases. Cancer. 1990;66:2612– 2619.
[5]. Pawade J, Wilkins BS, Wright DH. Low-grade B-cell lymphomas of the splenic marginal zone: a clinicopathological and immunohistochemical study of 14 cases. Histopathology. 1995;27:129–137.
[6]. Schmid C, Kirkham N, Diss T, Isaacson PG. Splenic marginal zone cell lymphoma. Am J Surg Pathol. 1992;16:455–466.
[7]. Mulligan SP, Matutes E, Dearden C, Catovsky P. Splenic lymphoma with villous lymphocytes: natural history and response to therapy in 50 cases. Br J Haematol. 1991;78:206–209.
[8]. Bolam S, Orchard J, Oscier D. Fludarabine is effective in the treatment of splenic lymphoma with villous lymphocytes. Br J Haematol. 1997;99:158–161.
[9]. Coiffier B, Thieblemont C, Felman P, Salles G, Berger F. Indolent non-follicular lymphomas: characteristics, treatment, and outcome. Semin Hematol. 1999; 36:198–208.
[10]. Lloret E, Mollejo M, Soc Mateo M, et al. Splenic marginal zone lymphoma with increased number of blasts: an aggressive variant? Hum Pathol. 1999;30: 1153-60