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RESEARCH ARTICLE

SPLENIC MARGINAL ZONE LYMPHOMA: A RARE CAUSE OF SPLENOMEGALY

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ABSTRACT

Splenic Marginal Zone Lymohoma(SMZL) is a very rare variety of lymphomas, accounting for only to 2% of NHL's (1,3) which has an incidence of 0.015% in India(2) .It originates from the memory B lymphocyte in marginal zone of secondary lymphoid follicle. Present as an indolent or disseminated disease at the time of diagnosis, with mass per abdomen, early satiety, incidentally detected altered blood counts. With a limited treatment options available like splenectomy, rituximab with or without chemotherapy, SMZL poses great challenge in diagnosis also. 65 year old gentleman, diagnosed as SMZL with symptomatic splenomegaly, underwent a splenectomy. IHC reports suggesting CD5(-), which is present only in 15 to 25% of the cases(4) of SMZL. In a developing country with restrained economic conditions, splenectomy can be a good diagnostic as well as treatment modality, as there is no consensus regarding the best treatment available at present.

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INTRODUCTION

Originating from the memory B lymphocytes in a microantomical compartment called marginal zone, SMZL accounts only for about 1 to 2 % of NHL's. Being a very rare entity, with a very few case reports from India, this case has CD 5 (-) making it even more rare. SMZL is more commonly associated with HCV in fections which is also negative in this scenario (Arcaini *et al*., 2006; Saadoun *et al*., 2005; Hermine *et al*., 2002; Thieblemont *et al*., 2003; Morse *et al*., 2001). Various studies quote that SMZL, characterised by splenomegaly with circulating CD 5 (-) villous B lymphocytes, described in malaria endemic areas, suggesting a possible role of infections in the pathogenesis (Bates *et al*., 2001). Since exact pathogenesis being unknown, even with advancement in classification of the disease, SMZL continues to be a disease, difficult to diagnose and categorise.

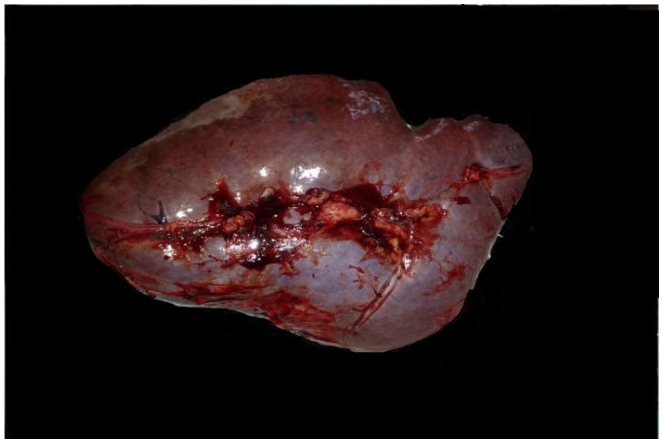
Revised Criteria For Diagnosis Of SMZL (Catherine Thieblemont, 2012)

- Based on the study of peripheral blood smear
- Bone marrow examination or,
- From the specimen of splenectomy.

65 year old man presented with huge mass per abdomen, decreased appetite, early satiety for a duration of six months with no significant family history or previous hospital admissions or treatment. Patient had huge, non tender, splenomegaly with spleen palpable per abdomen extending upto right ileac fossa along its axis. There was no other palpable organomegaly or lymphadenopathy.

Peripheral blood smear revealed mildanisopoikilocytosis, ormocytesmicrocytes and polymorpheic cells. TLC=5000 cells/cmm, DLC=N60, L34, E04, M02, B 00, adequate platelets and no parasites. Bone marrow aspiration suggestive of hypercellular bone marrow. Patient was negative for HIV, HbsAg, HCV serology and also negative for LD bodies. Serum LDH was raised, 434 U/ml, which is found in about 25% of the patients with SMZL (Mendes *et al*., 2014; Dreyling *et al*., 2013). OGD scopy revealed normal study. CT abdomen and pelvis suggestive of splenomegaly measuring 24 cm along its axis, liver being normal in size. Vaccination was given 2 weeks prior to the procedure. Patient underwent splenectomy under general anaesthesia. Histopathology of spleen suggestive of low grade B cell NHL-SMZL. IHC revealed – CD 19, 20 and BCl 2 positive, CD 3 reactive CD 5, 10, BCl 6, LDH3, cyclin D negative which was consistent with SMZL.

Specimen of spleen



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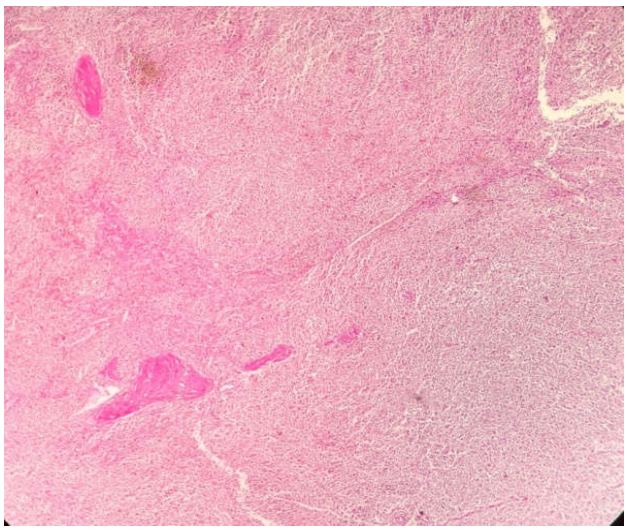
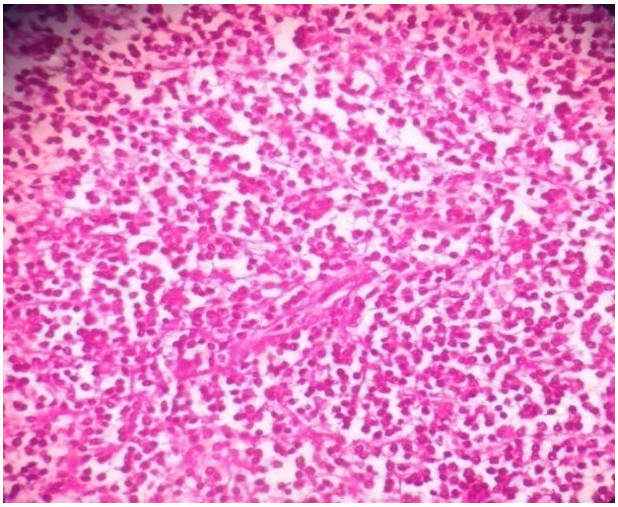
Tumor cells in low power field



Tumor cells in high power field



DISCUSSION

SMZL is a rare NHL. Presents at a mean age of 60 years with male to female ratio 1:1.8. Originating from spleen with variably involving lymph node, bone marrow, peripheral blood and other organs. Most common presenting complaints are abdominal discomfort due to splenomegaly. However may also present with abnormal blood count due to splenic sequestration than marrow infiltration. Patient may be asymptomatic with just a large spleen. B symptoms are uncommon.

WHO classified MZL into 3 types (Jaffe *et al*., 2001)

1. Extranodal Marginal Lymphoma Of MALT.
2. Splenic Marginal Zone Lymphoma.
3. Nodal Marginal Zone Lymphoma.

Even though it is well categorised, it is very difficult to diagnose and categorise. Disease can also be associated with HCV infection, malaria and auto immune conditions like primary biliary cirrhosis, rheumatoid arthritis, ITP, AIHA. Despite of low aggressive course there are reports with increased number of blastic crisis. In sporadic reports, Camacho *et al* had described blastic transformation in 13% of cases (Mollejo *et al*., 2005). Treatment options available at present are splenic irradiation (Troussard *et al*., 1996; El Weshi *et al*., 1998), chemotherapy, rituximab with (Cervetti *et al*., 2013) or without (Bennett *et al*., 2010; Kalpadakis *et al*., 2007) chemotherapy and splenectomy (Catherine Thieblemont, 2012; Evens and Blum 2015; Parry-Jones *et al*., 2003). Our patient had huge symptomatic splenomegaly, so splenectomy was justified in this case (Dreyling, *et al*., 2013; Silva dos Santos *et al*., 2017). Splenectomised patients show significant overall survival comparable with those treated with chemotherapy. The median overall survival in most series is about ten years, and 70% of the patients can remain treatment free for five years (Catherine Thieblemont, 2012; Evens and Blum 2015; Parry-Jones *et al*., 2003; Tayse Silva dos Santos *et al*., 2017). However, with extensive disease and debilitated patients, who cannot withstand surgery, should be considered for other modalities of treatment. This case report is to convey that, splenectomy is a good option as therapeutic as well as diagnostic modality, in patients with financial constrains and in patients with symptomatic splenomegaly, who cannot afford for chemotherapy or monoclonal antibody based treatment (Tayse Silva dos Santos *et al*., 2017).

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