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A Rare Case of Splenic Marginal Zone Lymphoma

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Abstract: ***Splenic marginal zone lymphoma is a rare non - Hodgkin lymphoma. Although it is an indolent lymphoma, about 10% of patients can be converted to diffuse large B cell lymphoma. Clinical presentation varies from asymptomatic splenomegaly to abdominal pain, early satiety, fullness and weight loss. Splenectomy is instrumental in diagnosis and treatment of a rare subtype of non - Hodgkin lymphoma. A 55 years old female presented with left upper abdominal pain for 1.5 years and generalized weakness for 6 months and on examination grade - 3 splenomegaly was found. Complete blood count suggestive of pancytopenia and USG shows 22cm spleen and bone marrow biopsy was negative for malignant cells. The patient underwent splenectomy and had an uneventful recovery.***

Keywords: Splenic Marginal Zone Lymphoma (SMZL), Splenectomy

1. Introduction

Lymphoma is a malignant tumour originating from the lymphatic hematopoietic system. According to the different cell sources, it is divided into non - Hodgkin lymphoma (NHL) and Hodgkin lymphoma (HL).

Marginal zone lymphomas (MZLs) represent a heterogeneous group of indolent lymphoproliferative disorders originating from memory B - lymphocytes, which are normally present in the marginal zone - that is, the outer part of the mantle zone - of the secondary lymphoid follicles.

Primary splenic lymphoma is a rare type of malignant lymphoma, which involves only the spleen and splenic hilar lymph nodes. However, splenic marginal zone lymphoma (SMZL) is even rarer.

SMZL represents a rare chronic B lymphocyte proliferative disease, which only accounts for about 1–2% of non - Hodgkin's lymphoma.

Clinical presentation varies from asymptomatic splenomegaly to abdominal pain, early satiety, fullness and weight loss. Although it is an indolent lymphoma, about 10% of patients can be converted to diffuse large B cell lymphoma. [3]

2. Case Report

A 55 years old female presented with dull - aching, non - radiating and intermittent left upper abdominal pain for 1.5 years and generalized weakness for 6 months.

Patient's vital parameters were within normal limits. On per abdominal examination grade - 3 splenomegaly was present.

3. Investigations

Complete blood count suggestive of anaemia (7.4g/dl), leukopenia ($1.97*10^9$/L) and thrombocytopenia ($109*10^3$/L). Ultrasonography (USG) revealed 22cm spleen with smooth contour, uniform, and normal echotexture.

Contrast enhanced CT (A+P+T) shows gross splenomegaly displacing left kidney inferomedially and no lymph node enlargement in abdomen (Figure 1).

Bone marrow trephine biopsy shows normal maturation and neither evidence of granuloma nor malignancy in the tissue planes examined.

FDG PET - no evident FDG avid disease is seen in spleen, liver and lung.

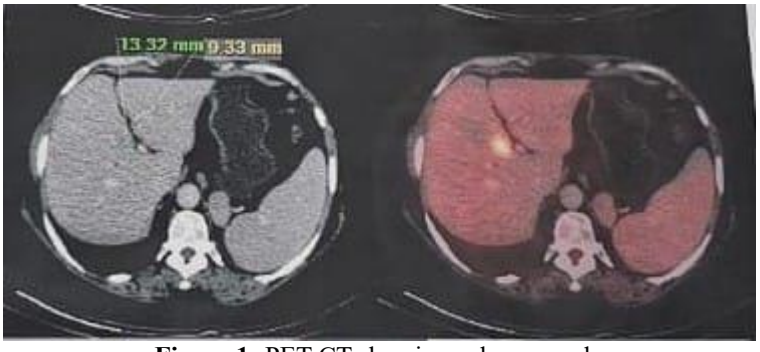

Figure 1: PET CT showing splenomegaly

Operative Intervention

Planned laparotomy done. Approximately 25*22*4 cm spleen with two splenunculi removed (Figure 2). Liver surface was normal. No enlarged splenic hilar lymph nodes. Postoperative period was uneventful. Patient was discharged on postoperative day 3 and followed up on POD 7, 15, 30 days and 6th month with no late post - op complication. No further radio - chemotherapy needed.

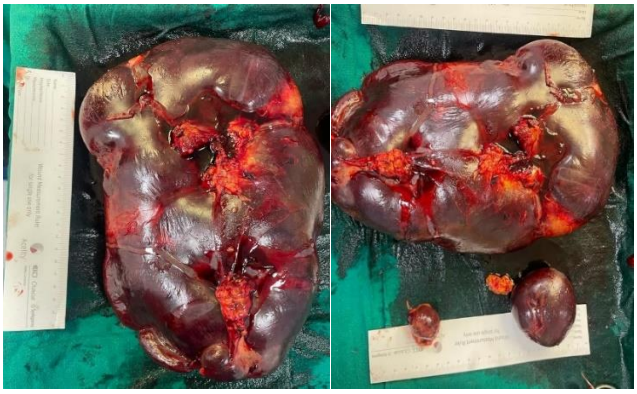

Figure 2: Removed spleen with two splenunculi

Biopsy Report

HPE confirmed diagnosis of low grade B cell non - hodgkins lymphoma - Splenic marginal zone lymphoma. No evidence of large cell transformation (Figure 3).

IHC markers CD 20 (Figure 4), BCL 2, Ki - 67 20% were positive and BCL6 & PAX5 negative.

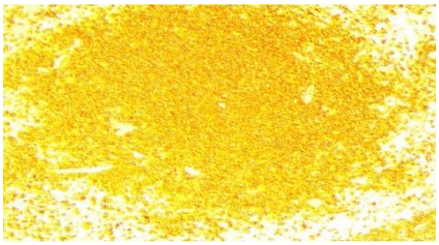

Figure 3: Enlarge white pulp of spleen

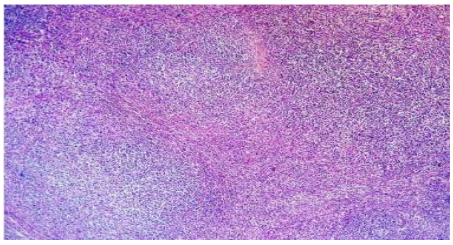

Figure 4: IHC CD20 positive

4. Discussion

SMZL is the second most common subtype of marginal zone lymphoma, comprising about 20% of the cases. It represents about 0.9% of all NHL.

In addition, patients with SMZL were frequently diagnosed as co - infected with hepatitis C virus (HCV). [2]

Splenomegaly is the most typical symptom of SMZL, and some patients present with decreased appetite, fever, night sweats, and sudden weight loss. Diagnosis of SMZL is based on clinical feature of unexplained lymphocytosis and splenomegaly. [1]

Staging relies mostly on integrated PET/CT, this also helps with targeting fluorodeoxyglucose - avid lymph nodes with biopsy.

Ideally, biopsy is obtained before initiation of steroid therapy as steroids would lyse lymphoid tissue and might obscure the diagnosis. [1]

Splenectomy is indicated for NHL patients with massive splenomegaly leading to abdominal pain, early satiety, and fullness.

It may also be indicated for patients who develop anaemia, neutropenia, and thrombocytopenia associated with hypersplenism.

For HCV - positive SMZL patients with no obvious surgical indications (such as hypersplenism), interferon alone or combined with ribavirin for anti - HCV therapy may be considered as an effective option. [2]

In patients with spleen - predominant features, survival is significantly improved after splenectomy and could be considered as a mainstay therapy for those that are surgical candidates. [1]

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