ABSTRACT

**Background:** Out of the various malignant tumours originating from the lymphatic hematopoietic system, lymphoma is one such important entity. It is divided into Non-Hodgkin’s Lymphoma (NHL) and Hodgkin Lymphoma (HL) depending on its cell source. A very rare type of malignant variant of lymphoma is the primary splenic lymphoma, involving exclusively the spleen and splenic hilar lymph nodes. Moreover, splenic marginal zone lymphoma (SMZL) is even more infrequent. SMZL is an uncommon chronic B lymphocyte proliferative disease, which only accounts for about 1–2% of all non-Hodgkin’s lymphoma. The mean age of SMZL incidence is about 65 years. There is no known significant gender predominance. A quarter of patients with early diagnosed SMZL have known to have vague symptoms like abdominal pain and distention; and other patients may be accompanied by loss of weight, malaise, cachexia, splenomegaly, or other manifestations.

**Conclusion:** Although, a good prognostic outcome is what is usually expected from most patients of Splenic Marginal Zone Lymphoma who undergo splenectomy, an aggressive transformation leading to a worse direction cannot be ruled out. SMZL is very challenging to be diagnosed pre-operatively due to the lack of specificity in clinical presentation.
Keywords: Lymphoma; primary splenic lymphoma and loss of weight.

1. INTRODUCTION

A very rare type of malignant variant of lymphoma is the primary splenic lymphoma, involving exclusively the spleen and splenic hilar lymph nodes. Moreover, splenic marginal zone lymphoma (SMZL) is even more infrequent [1]. SMZL is an uncommon chronic B lymphocyte proliferative disease, which only accounts for about 1–2% of all non-Hodgkin’s lymphoma. The mean age of SMZL incidence is about 65 years [2]. There is no known significant gender predominance. A quarter of patients with early diagnosed SMZL have known to have vague symptoms like abdominal pain and distention; and other patients may be accompanied by loss of weight, malaise, cachexia, splenomegaly, or other manifestations [3]. The origin of the cells of SMZL is believed to originate from memory B lymphocytes in the marginal zone of the secondary lymphoid follicles, yet more research is undertaken to prove its certainty [4].

Researchers are still exploring the pathogenesis and progression of the disease but there has been some evidence that it is related to changes in the short arm of chromosome 7. Even though SMZL is a slow-growing lymphoma, one-tenth of the affected individuals can progress to diffuse large B cell lymphoma, deteriorating the condition of the patient and developing symptoms [5]. SMZL progresses with a relatively better prognosis than other types of lymphomas. The median survival time of the disease is known to be 10 years provided the disease doesn’t progress or there is no obvious hypersplenism [6]. Considering how less the incidence of this tumor is, very little is known about the diagnostic and therapeutic approach of the patient and thus the treatment modalities are constrained to long-term follow-up, keeping in mind the absence of hypersplenism or disease progression. Generally, surgical excision for both diagnostic and therapeutic purposes, along with postoperative adjuvant chemotherapy and/or radiotherapy is recommended to improve the prognosis [7]. To date, SMZL clinically falls under Non-Hodgkin’s Lymphoma and therefore, it is classified as Stage III Non-Hodgkin’s Lymphoma as the spleen is involved. Though NHL is known to have an aggressive nature, SMZL is relatively indolent with a favorable prognosis [8]. Given this background, we herein reported a case of SMZL of a 49-year-old male with gross hepatosplenomegaly.

2. CASE REPORT

A 49-year-old male was admitted with complaints of generalized weakness, cough, and abdominal pain off and on for 1-2 months. On examination general condition was moderate, afebrile, pulse was 80 beats/min, blood pressure - 130/90 mmHg, respiratory rate - 22/min, pallor was present, while the cardiovascular, nervous and respiratory system examination revealed no abnormal findings. The abdomen was soft and non-tender. CT abdomen was advised which shows gross hepatosplenomegaly and a few mildly enlarged pedophiliac and paraaortic lymph nodes. To make a definite diagnosis, a splenectomy was planned after 7 days.

Splenectomy with lymph node biopsy with liver wedge biopsy was performed and the specimen was sent for histopathology. The Department of Pathology received a specimen of spleen measuring 26 x 17 x 7 cm in size. On the cut section, it was colored reddish-brown. On microscopy, the spleen shows congestion of sinuses and effacement of architecture by centrocytes-like cells. These cells are having cleaved nuclei and eosinophilic cytoplasm. Follicle formation was seen, so reported as suggestive of primary splenic lymphoma with infiltration in liver and lymph nodes. Immunohistochemistry was advised for confirmation which was reported as Splenic Marginal Zone Lymphoma (SMZL) with lymph node involvement and liver infiltration. It was positive for ki-67, CD20, and BCL-2 and negative for CD3, CD5, cyclin D1, and CD23. The oncologist decided to go with 6 cycles of chemotherapy. After 2 years of follow-up, the patient showed no signs of recurrence and spread.

3. DISCUSSION

SMZL forms less than 2% of all cases of Non-Hodgkin’s Lymphoma and thus is known to be a rare form of torpid B-cell neoplasm, affecting the bone marrow, spleen, and also the peripheral blood [7]. The definitive diagnosis of this condition pre-operatively has been demanding in the early stages of the disease due to the lack of specific clinical features, imaging, or laboratory findings and by the time patients present with symptomatic splenomegaly and cytopenia, the disease has already advanced in course. This fact makes histopathology the only modality to
give a definitive diagnosis. Splenectomy alone has been enough for patients with SMZL to have a maintained remission for several years [8].

Splenomegaly, sometimes very massive that the spleen weighs more than 2 kilograms is also seen. On microscopy, there is a proliferation of the lymphoma cells thereby replacing the white pulp and also infiltrating into the marginal zone. At times, there is infiltration in the splenic sinuses of red pulp which can be in a patchy or diffuse fashion [9]. The peripheral blood smear shows characteristic villous lymphocytes having basophilic cytoplasm with short polar villi, round nuclei with condensed chromatin. This feature is needed to differentiate SMZL from hairy cell leukemia. SMZL is immunophenotypically positive for CD79a, CD20, BCL-2 and IgM. It is negative for BCL-6, CD5, CD10, CD43, and CD103. Immunohistochemistry and flow cytometry is considered the gold standard to rule out other lymphomas [10].

SMZL is very challenging to be diagnosed pre-operatively due to the lack of specificity in clinical presentation. With this case report, we want to encourage further study on this lesion to understand the disease progression and thus channelize the treatment accordingly.

ETHICAL APPROVAL

Ethical clearance Taken from institutional ethics committee.

CONSENT

As per international standard or university standard, patient’s written consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES


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