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Case Report

Splenic Marginal Zone Lymphoma (SMZL): A Rare Case Report

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Received: September 15, 2021; **Published:** October 27, 2021

Abstract

The spleen may be the primary site of the lymphoma or it may be a part of a disseminated malignancy. There is no precise definition and it is not included in the recent World Health Organization classification of hematolymphoid neoplasms. The diagnosis of malignant lymphoma presenting as an initial splenic manifestation may go unrecognized. In this article, we are reporting a case of SMZL in a 40 year old female who presented with abdomen distension associated with abdominal pain diagnosed in the operation after that she given adjuvant chemotherapy and she is still a life without any relapse or metastasis till now.

Keywords: *Primary Splenic Lymphoma; Splenic Marginal Zone Lymphoma; Splenectomy; Adjuvant Chemotherapy*

Introduction

The number of primary splenic lympho mas being reported is increasing despite the rarity of this malignancy, but what really constitutes a lymphoma arising primarily in the spleen is still a matter of discussion [1]. The splenic presentation is the rule for some pathological entities, such as Splenic Marginal Zone Lymphoma (SMZL) [2].

Case Report

A 40 years old women presented with abdomen distension for eight months and a two months history of abdominal pain, not associated with fever or night sweats, no any lymph nodes enlargement or palpable, no weight loss, Comorbidity is AHT on regular treatment. Her HBV was positive. Ultrasonography of whole abdomen revealed only a large splenomegaly. She did abdomen and pelvic CT Scan appeared moderate splenomegaly with no focal lesion. At first, she treated medically as Gastritis case without any benefit and her complain continuous. Last months before operation her complain is a progressive and not response to any pain Killer. She underwent to splenectomy on July 2017. The histopathological examination is revealed Huge and an intact spleen follicular type non-Hodgkin's lymphoma. Immunohistochemistry (IHC) results was: strong positivity CD20, LCA, Bcl2 and negativity of CD3, CD5, TDT, Cyclin-D2.IHC was confirmed the splenic marginal zone lymphoma (SMZL) as the final diagnosis. Post-Operative CT-scans for re-evaluation are including: Neck and Chest CT scan: several small posterior cervical L.N (1.3 cm) and the other is Normal, Abdomen and Pelvic CT Scan was appeared Left partial nephrectomy, soft tissue at surgical bed (2.2 cm). Blood investigations were ESR = 69, B2- macroglobulin = 7.2, LDH = 700. She did receive adjuvant chemotherapy, six cycles of CHOP protocol with well tolerance and good response. Para clinical where reduced to the normal range LDH from 700 pre-chemotherapy to 370 after that, B2- macroglobulin from7.2 to 1.7 later on. The patient was evaluated in regular interval at first every 3 months for the first year then every 6 months after that annually. Follow- up evaluation consisted of interval history and physical examination. The CT scans and tumor marker including ESR, B2-microglobulin, LDH were used during the follow-up always staying in the

normal range. Six months after completion of therapy the follow up results is LDH 372, B2- macroglobulin 0.8. Neck, Chest, Abdomen and pelvic CT-Scans are appeared two nodules at site of surgical bed (2 cm) and the other are normal. Last evaluation results were Neck and Chest CT scans are normal and Abdomen and Pelvic CT scan is stationary course related to the two nodules at site surgical bed (2 cm) and the other are normal. ESR 36, LDH 107, B2micro 0.9, Calcium 7.4, CA15.3 (9.2). Breast ultrasound is normal. Our patient is complete remission without any complain or any relapse according to the last evaluation on September 2020.

Discussion

Primary splenic lymphoma is a relatively infrequent cause of splenomegaly, its reported incidence being less than 1% [3].

The clinical course of PSL is thought to be more favorable than NHL that originates elsewhere and eventually spreads to the spleen and necessitates splenectomy [4]. The spleen may be the primary site of the lymphoma or it may be a part of a disseminated malignancy [5]. Primary splenic lymphoma is a relatively infrequent cause of splenomegaly, its reported incidence being less than 1% [3]. Splenic marginal zone lymphoma (SMZL) is a distinctive and well-characterized B-cell neoplasm that involves the spleen and various organs. Although SMZL is accepted as an entity in the World Health Organization (WHO) classification [6], Splenic marginal zone lymphoma (SMZL) is a relatively rare neoplasm accounting for 1 - 3% of all NHLs but it represents the most frequent lymphoma presenting in the spleen. SMZL was first described in 1992-63 [7]. The peak of incidence of SMZL is in the seventh decade of life and, at variance with other mature B-cell lymphoproliferative disorders, it is more frequent in female subjects. Select chemotherapy regimen for PSL, according to the individual cases despite the lack of clinical trials on the subject [8]. SMZL is universally considered a low-grade lymphoma with an indolent clinical course. Many cases show a protracted uncomplicated evolution, a good response to splenectomy or chemotherapy [2].

Conclusion

PSL is a very rare condition, often presenting with non-specific features and a splenic mass. Most often, the diagnosis is made after the histopathological evaluation of the splenectomy specimen. Splenectomy offers both definitive histological diagnosis and therapeutic resection and represents the mainstay of treatment. When splenectomy is combined with combination chemotherapy, outcomes in terms of remission and survival are improved. Our case was treated by splenectomy then chemotherapy with good response without any relapse or metastasis until published this article.

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Volume 4 Issue 11 November 2021