SPLENIC LYMPHOMA IN HCV POSITIVE PATIENT; A CASE REPORT

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ABSTRACT

Splenic Lymphoma is a marginal zone lymphoma (MZL`s) that is a type of Non-Hodgkin`s Lymphoma. It is characterized by a lymphocytic tumor of marginal zone of spleen that may involve splenic hilar lymph nodes and may spill the abnormal B-Lymphocytes in the blood. A 38 years old male, known HCV, presented with pain in left hypochondrium and an abdominal mass in left hypochondriac region. He was diagnosed clinically as a case of splenic hematoma which turned to be splenic lymphoma after investigations and was treated.

Key Words: Splenic lymphoma, HCV positive Patient, Splenectomy

INTRODUCTION

The most common splenic malignancy is lymphoma.[1] Splenic marginal zone lymphoma (SMZL) with or without villous lymphocytes is a disorder that was recently recognized as a distinct pathological entity in the World Health Organization classification. At diagnosis, SMZL presents as an indolent and disseminated disease that is originally recognized after histopathological examination of surgically removed spleens as SMZL itself, or by means of morphological and immunophenotypic characterization of circulating neoplastic lymphocytes as splenic lymphoma with villous lymphocytes.[2] Cytopenia and lymphocytosis are frequently observed.

According to WHO classification, lymphomas are divided into Hodgkin`s and Non-Hodgkin`s. Non-Hodgkin`s lymphomas are adult onset usually in late 40`s and present as lymphadenopathy of various regions notably in the groin, spleen, in the axilla and the neck. Marginal Zone lymphomas is a further classification of Non-Hodgkin`s lymphomas which present as a tumor like growth in secondary lymph node follicles and begin in the marginal zone of the node, later eroding adjacent zones of the node.[2] It presents as hard node and involves the whole node later on. Splenic Marginal Zone lymphomas, a malignant condition, are found in the white pulp of the spleen and involve invariably the red pulp later on. It may also involve splenic hilar lymph nodes, bone marrow and blood. A particular glycoprotein of the Hepatitis C virus may be able to activate a specific B-Lymphocyte in the spleen which may act as origin of the tumor. Characteristic basophilic villous shaped B-Lymphocytes may or may not be found in blood, but when present are pathognomonic of Splenic Lymphoma.

Lymphomas are 5.3 % of all cancers and 55.6 % of all blood cancers. Marginal Zone lymphomas represent 5-17 % of all Non-Hodgkin`s lymphomas and SMZLs represent 20 % of all MZLs.[3] According to WHO, every person has 1 in 50 chance of developing this malignant condition. It is found preferably in males after the age of 50. In a study from Surveillance, epidemiology and End result (SEER) program in united states, out of 116411 cases of Non-Hodgkin`s lymphoma, 763 cases were Splenic Marginal Zone lymphomas. Exact etiology is still unknown, but chemotherapy, Hepatitis C virus infection, decreased immunity states such as AIDS, various chemicals like PCB(polychlorinated biphenyl), Radiation therapy, alkylating agents may induce this malignant state of spleen.[4] The purpose of this case was to enlighten the surgeons as well as other healthcare professionals so that they may keep this condition in mind while treating a case of HCV positive having mass in abdomen.[4]

CASE REPORT

A 38 years old male of 5.9 feet height and 68 kg weight, labourer by occupation, resident of Faisalabad city presented with pain in the left hypochondrium and left lumbar region for three months, with feeling of lump in the left hypochondriac region as well. Pain was moderate in intensity and increased with cough. He had no history of vomiting, weight loss, night sweats. History revealed no significant past history, medical

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history of relevance. There was no family history of similar disease. On examination there was no pallor, cyanosis, anemia and lymph node enlargement throughout the body. Abdominal examination showed tenderness in left hypochondrium and spleen was moderately enlarged in size with no other visceromegaly. No clinical evidence of hypertension, diabetes mellitus or any other significant systemic disease. Patient had been positive for HCV on chromatographic method for 8 months and was confirmed positive on ELISA as well as on PCR at the time of admission. He was diagnosed previously as a case of splenic hematoma but on ultrasonography, its non-communicating nature was revealed and was considered lymphoma.(Figure I). Its size was 9.1x9.0 centimeters. CT scan abdomen and pelvis confirmed a localized mass within the spleen. All the other viscera of the abdomen and pelvis were in normal shape and texture both on ultrasonography and CT. His hemoglobin was 12.8 g/dl and Total leucocyte count was 11500/cmm. Prothombin Time, Activated Partial Thromboplastin Time and other Liver Function Tests revealed no significant hepatic abnormality. Splenectomy was done and specimen was sent for histopathology in the laboratory which confirmed it to be splenic marginal zone lymphoma.(Figure II). Patient was discharged after seven days uneventfully. Pneumococcal, meningococcal and Hib vaccines were given at the time of discharge. He was counseled for post-splenectomy consequences and was called for follow up after 3 weeks.

Fig I: Ultrasonographic findings of the splenic marginal zone lymphoma

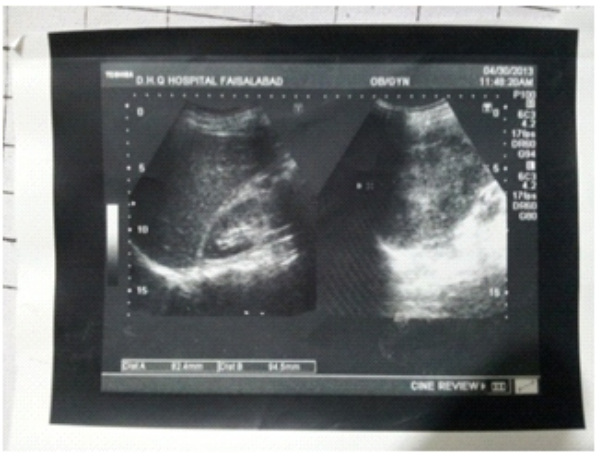
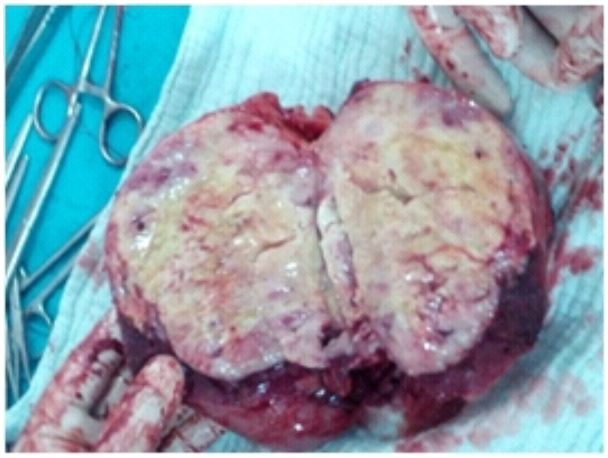


Fig II: Gross appearance of splenic marginal zone lymphona



DISCUSSION

The case of splenic lymphoma associated with Hepatitis C virus or caused by it, is quite a rare case in Faisalabad. It is considered to occur after the age of 50 years but in our case the patient was 38 years old which is rare. The risk of SZML to run in the family is almost negligible.

No abnormal basophilic villous shaped cells were found in blood. These cells may or may not be present in the peripheral blood. When present, these confirm diagnosis. The hilar splenic lymph nodes and bone marrow were not infiltrated. Patient was a known case of Hepatitis C for more than 3 months. In a study, the hepatitis C virus was found to have influence on the development of lymphoma in spleen and various other sites. A specific glycoprotein (E2) was found to interact with CD81 receptor in the precursor B-cell that lead to proliferation of B-cells. Bone marrow biopsy (BMB) is done to confirm the presence of cells in it. Bone marrow biopsy has led to spare the biopsy of spleen. In a study bone marrow biopsy was proved to as efficient as spleen biopsy itself in diagnosing and confirming the disease.[5] Splenectomy was considered frontline treatment plan for it but in various centers chemotherapy is done and splenectomy is spared for recurrent splenomegaly associated with this condition. Chemotherapy is recommended for patients who can`t undergo surgery, elderly or those who have recurrent disease. After the discovery of rituximab,[6] the treatment of splenic marginal zone lymphoma was revolutionized. Now the primordial treatment for splenic marginal zone lymphoma is rituximab therapy. Rituximab is given to all patients irrespective of their age, gender and co-morbidity.

Splenectomy is done only in aggressive disease and in cases where splenomegaly is massive. Various studies have proved rituximab to be more beneficial than splenectomy and overall prognosis of rituximab was much better than splenectomy. In cases, where splenectomy was inevitable due to its size, laparoscopic approach was done. Laparoscopic splenectomy is considered superior to open methods.[7] Median survival rate of patients is 5-10 years but in more aggressive disease it can be less than 4 years. Precise decision depends upon the growth size and grade of tumor. In the study done in USA on epidemiology, the median survival rate of patients after treatment was 5 years. Comparatively, the overall survival rate of splenic zone lymphomas was less than other variants of marginal zone lymphomas such as nodal marginal zone lymphomas(NMZL) and mucosa associated lymphoid tissue (MALT) which have better prognosis as well as longer survival rates.[8,9]

CONCLUSION

This case report suggested that while treating HCV positive case having mass in left hypochondrium, Splenic Marginal Zone lymphoma, may be kept in differentiate diagnosis.

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