OMICS Group Conferences
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2nd International Conference on
Gastroenterology & Urology
June 10-12, 2013 Hilton Chicago/Northbrook, USA

More than the size of a fist: A case of small cell lymphocytic lymphoma in a 47 year old male

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Introduction: We are presented with a clinical scenario of diagnostic dilemma in establishing the cause of massive splenomegaly which requires not only a pathologic staging but moreover pathologic diagnosis of which splenectomy was an option.

Description: A 47 year old male with incidental finding of isolated massive splenomegaly associated with hepatomegaly with liver span of 11 cm midsternal line, 15 cm midclavicular line and lymphadenopathies involving the cervical, axillary and inguinal lymph nodes pauses dilemma in establishing diagnosis since laboratory results showed inconsistent findings.

Discussion: A wide array of diseases associated with massive splenomegaly were considered from infectious process (Chronic Hepatitis Infection, Extrapulmonary Tuberculosis, Schistosomiasis), Neoplastic process (Non Hodgkins and Hodgkins Lymphoma) to Congestive Splenomegaly (liver cirrhosis with portal Hypertension). Though CT scan result with splenic index of 4337 and hematology findings of pancytopenia were noteworthy, other diagnostic results were not consistent. Biopsy of the inguinal mass revealed diffuse hyperplasia. Immunohistochemistry result of CD10, CD15, CD20, CD30 were all negative. Rectal biopsy was done which was negative for schistosoma ova. BSMP, rectal biopsy, serum AFP and Hepa B were also negative. Splenectomy was done with intraoperative findings of massive splenomegaly with spleen weighing 2.5 kilogram, an enlarged splenic hilum node and splenic artery, hepatomegaly and multiple hepatic masses at segments 4, 5, 6. Histopathologic finding of the spleen was confirmatory of small cell lymphocytic lymphoma with a pseudofollicular pattern composed of small lymphocytes, prolymphocytes and paraimmunoblast.

Small cell lymphocytic lymphoma accounts for about 4-5% of non-hodgkins lymphoma. There are 5 significant risk factors to the prognosis of overall survival which includes age, serum LDH levels, performance status, stage and extra-nodal site involvement. In this case two risk factors were identified and with the presence of extra-nodal site involvement the patient belongs to SCCL stage IV-B hence CHOP regimen was given with a note of lymph node regression. This indicates that the patient has less than 50% chance of relapse free and overall survival of five years.

Although the implication of splenectomy is controversial considering the risk of surgery and its post op complication, performing the procedure in this case is of imperative significance in establishing diagnosis and alleviating symptoms of our patient.

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