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Isolated Lymphoma of Spleen – A Rare Entity With Difficult Pre Operative Diagnosis

KEYWORDS

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ABSTRACT *Lymphocytes collects in lymph nodes,spleen and thymus.If lymphocytes start to divide and multiply uncontrollably ,they can built up and form a lymphoma.*

A 46 year old women came with complaints of pain abdomen for 5 months and swelling in left upper abdomen for 3 months, which is not progressive.USG of abdomen showed enlarged spleen of size 163 mm in long axis with suspicious SOL, subsequent CT abdomen revealed non enhancing hypodense lesion in mid polar region of spleen.With the suspicion of splenic tumour open splenectomy was performed, intraoperatively irregular growth was noted in spleen with splenic hilar lymphadenopathy.Splenectomy with hilar lymphadenectomy was performed and the specimen was sent for HPE and later it came out to be lymphoma of spleen.

Lymphoma of spleen is a rare phenomenon.Although it is rare it responds well to treatment.Clinical recognition of this is difficult but it is critical to institute appropriate treatment.

INTRODUCTION

As a secondary lymphoid organ, the spleen is a common site of lymphoma dissemination and can be involved with any lymphoid malignancy.However, splenomegaly as the predominant presenting symptom is relatively uncommon. Primary splenic lymphoma is rare with a reported incidence of less than 1 % of all lymphomas.(1)Several lymphoma subtypes may present with isolated splenomegaly, including diffuse large B-cell lymphoma, mantle cell lymphoma, hairy cell leukemia, splenic marginal zone lymphoma, prolymphocytic leukemia, chronic lymphocytic leukemia, and Waldenström macroglobulinemia.

CASE REPORT

46 year old female presented with chief complaits of pain in epigastric region for 5 months, swelling in left upper abdomen for 3 months.No other significant history.On palpation there is a swelling in left hypochondrium extending 7 cm below costal margin.

INVESTIGATIONS

Haemogram – Hb-9.9 g/dl,TLC – 8910 cells/cu mm,DLC N - 66.8%,L – 25.6%,M -2%,E – 5.2%,B – 0.4%.Peripheral smear – normocytic normochromic, no immature cells. Platelet – 3.12 lakhs /cu mm.HbsAg, HIV, HCV – negative. Sickling test – negative.Chest X ray – normal.ESR - 15mm. Mantoux – negative.

USG abdomen and pelvis – enlarged spleen measures 163 mm in long axis with inhomogenous echotexture ? SOL. Rest of the abdominal organs are normal.CECT of abdomen and pelvis – Splenomegaly – 11.5 x 9.3 cm.Non enhancing hypodense lesion in midpolar region of spleen? Infarct.

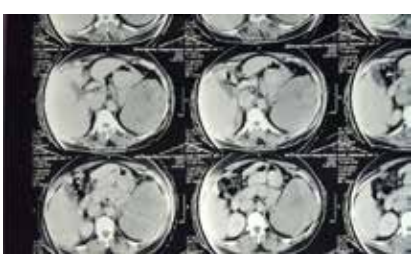

Fig. 1 CT showing non enhancing hypodense lesion in midpolar region of spleen.

INTRA OPERATIVE FINDINGS

Intra operative examination of abdomen and pelvis ruled out any other lymphadenopathy.Liver and other intraabdominal organs are normal.Resected specimen of spleen measured 18 x 12 x 8.5 cm with intact capsule weighed 980 grams.Surface – multinodular.Cross section – large nodular appearance.Hilum of spleen contained matted lymph nodes and 2 matted lymph nodes in hilar region resected seperately showed homogenous pattern.

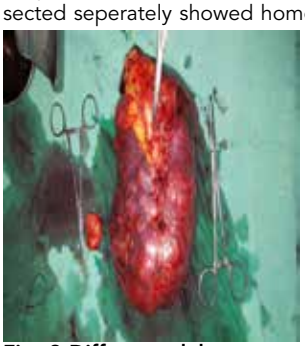

Fig. 2 Diffuse nodular appearance of spleen.

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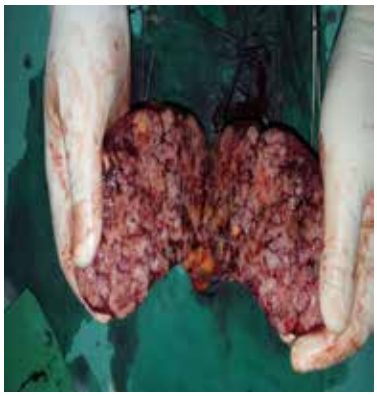

Fig 3 Specimen showing focal area of necrosis.

HISTOLOGY

Monotonous pattern of the large lymphoid cells in the diffuse pattern with increased mitotic figures with foci of necrosis.Lymph nodes also showed similar histological picture.Diagnosis of diffuse large B cell non hodgkin's lymphoma of spleen with stage 2 is made.Bone marrow study found to be normal.Patient referred for chemotheraphy R CHOP regime.

DISCUSSION

Isolated presentation with splenomegaly is more common in indolent lymphoma than in aggressive lymphoma. Many patients seek medical attention because of progressive discomfort in the left upper quadrant. Initial laboratory assessment should include a complete blood cell count and differential; basic metabolic panel; liver function tests; calcium, uric acid, lactate dehydrogenase, and β2 microglobulin levels; serum protein electrophoresis with immunofixation; and hepatitis B and C serology.CT scan of abdomen and pelvis a should be performed.Fluorodeoxyglucose positron emission tomography (FDG-PET) imaging is adjunctive but optional in these cases. Many splenic lymphomas have a tropism for the bone marrow and a high likelihood of producing circulating malignant cells.Thus, evaluation of the peripheral blood with the assistance of a skilled hematopathologist may lead very rapidly to a diagnosis.A bone marrow biopsy may be required to establish the diagnosis.

Patients without detectable blood or bone marrow involvement but continued suspicion of splenic lymphoma represent a difficult diagnostic dilemma.Splenectomy should be discussed with the patient, as it can have the dual benefit of establishing the diagnosis while simultaneously providing therapeutic benefit. Patients with splenomegaly due to macroscopic splenic nodules can be considered for core needle biopsy.(2)

previous concerns about bleeding, a recent meta-analysis showed a major complication rate of only 1.3% when needles smaller than 18 gauge were utilized(2).In cases of diffuse splenic infiltration, splenectomy is preferred over core needle biopsies, as the former is more likely to establish the diagnosis while simultaneously providing initial therapy.

The clinical presentation of NHL varies tremendously depending upon the type of lymphoma and the areas of involvement. Aggressive lymphomas commonly present acutely or subacutely with a rapidly growing mass, systemic B symptoms (ie, fever, night sweats, weight loss), and/or elevated levels of serum lactate dehydrogenase and uric acid.Examples of lymphomas with this aggressive or highly aggressive presentation include diffuse large B cell lymphoma, Burkitt lymphoma, adult T cell leukemia-lymphoma, and precursor B and T lymphoblastic leukemia/lymphoma.

Indolent lymphomas are often insidious, presenting only with slow growing lymphadenopathy, hepatomegaly, splenomegaly, or cytopenias. Examples of lymphomas that typically have indolent presentations include follicular lymphoma, chronic lymphocyticleukemia/small lymphocytic lymphoma, and splenic marginal zone lymphoma.

Lymphomas involve the white pulp of the spleen, in contrast to the leukemias and histiocytomas which involve the red pulp.They involve all the malphigian bodies uniformly and cause an increase in the number of white pulp nodules per unit of the spleen, without associated germinal center formation.(3)

Ahmann et al staged primary splenic lymphomas as

Stage 1 : disease confined to spleen.
Stage 2 : disease involving the hilar nodes also.
Stage 3 : disease spreading to other intraabdominal lymph nodes.

Two thirds of patients present in stage 2 or more.(4)

Gorg et al studied pattern of splenic involvement in series of 680 cases and identified four types, diffuse(37%), focal small nodular(39%), focal large nodular(23%), and bulky disease(2%).(5)

High grade and large cell lymphomas showed large or small nodular lesions, while low grade lymphomas and hodgkins disease showed diffuse or small nodular pattern. The small lymphocytic type is the most difficult to diagnose, however run the most indolent course.Large cell type has a more fulminant course associated with tissue necrosis and degeneration.

Chemotherapy – R – CHOP,rituximab, cyclophosphamide, doxorubacin, vincristine and prednisolone.R – EPOCH(etoposide).ABVD, which contains adriamycin, bleomycin, vinblastine and doxorubicin.R-CVP, which contains rituximab, cyclophosphamide, vincristine and prednisolone.A stem cell transplant is the treatment of choice for DLBCL patients whose cancer has returned or relapsed.High-dose chemotherapy coupled with a stem cell transplant can be used to treat patients with DLBCL who have failed initial chemotherapy, but are responsive to a second chemotherapy regimen.

Newer agents like alisertib, bortezomib, everolimus, vorinostat are currently investigated for the treatment of lymphoma.Patients in remission should have regular visits with a physician who is familiar with their medical history as well as with the treatments they have received.Blood tests and computed tomography (CT) scans, may be required at various times during remission to evaluate the need for additional treatment.

CONCLUSION

When we see a patient, our initial focus is on determining urgency of diagnosis and treatment. The history, focusing on symptom duration and severity, along with a focused exam assessing for haemodynamic abnormalities, signs of marrow failure, and size of spleen/presence of symptoms

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of rupture, can help identify patients who may need admission for expedited evaluation.

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