Case Presentation: A 57 year old man with fever, anemia, and splenomegaly

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Background

Splenic marginal zone lymphoma is a rare B cell lymphoma that is often asymptomatic until spleen enlarges to cause left abdominal pain. We report here a case of 57 year old Caucasian male who presented with subjective fever. A contrast enhanced computerized tomography suggested a 30 cm spleen. Bloodwork showed lymphocytosis, and flow cytometry revealed monoclonal B cell lymphoma. Although splenomegaly with systemic symptoms is a very rare presentation of splenic marginal zone lymphoma, it should be kept in mind when investigating splenomegaly of unknown origin.

Case Report

- A 57 year old Caucasian male with past medical history of hypertension, hyperlipidemia and left retinal detachment presented to emergency room with three days of left-sided abdominal pain.
- He complained of left-sided pain and firmness in the abdomen. Pain was not relieved with bowel movements. His history was notable for 10 lb weight loss in the past 3 months, left shoulder and scapular pain, shortness of breath, subjective fever and intermittent night sweat. No nausea, vomiting, diarrhea, constipation or recent travel history.
- Vital signs:

  T=38.2 oral, BP=113/70, RR=16, Pulse=90, O2 sat 97% on room air.
- Physical exam was notable for decreased breath sounds and rales of left lower lung base, distended abdomen, and spleen palpable up to below umbilicus.

Hospital Course

- In the ER, CT of abdomen showed massive splenomegaly and CXR showed left pleural effusion.
- Differential diagnoses for splenomegaly include cirrhosis 33%, lymphoma 27%, infection (AIDS, endocarditis) 23%, inflammation 8%, and primary splenic disease (splenic vein thrombosis) 4%.
- Since CBC showed lymphocytosis with elevated atypical lymphs, there was a concern for lymphoma versus leukemia.
- Peripheral blood immunophenotyping revealed monoclonal B cell lymphatic disease. Bone marrow aspiration and biopsy showed low grade B cell lymphoma.
- He was started on Rituximab treatment, but within 10 minutes, he vomited and treatment was discontinued.
- Splenectomy was indicated. Splenic artery embolization was performed prior to splenectomy.
- The final pathology results: Splenic marginal zone lymphoma with predominant red pulp involvement.

Laboratory and Imaging

CBC: 29.7 | 10.6 / 32.3 | 92

BMP: 138 | 102 | 22 / 4.3 | 27 | 1.21 | 96

Laboratory	Result	Reference Range
RDW	17.1	11.5-14.5 %
MCV	77.8	81-100 fl
lymphocytes	63	19-48 %
seg Neutrophils	21	40-74%
atypical Lymphs	15	0-5%
lipase	20	8-57 u/l
lactic acid	14	5-18 mg/dl
amylase	35	28-100 IU/L
LDH	358	0-242 IU/L
AST	20	0 – 37 IU/L
ALT	9	0 – 35 IU/L
Albumin	3.7	3.5 – 5.0 g/dl
INR	1.0	0.9 – 1.1
Markers	Present CD19, CD20	Absent CD5, CD10, CD25
	JAK2	neg
	BCR-ABL	Neg

Fig 1. Laboratory result

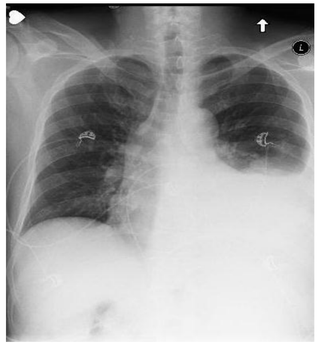

Fig 2. Chest x-ray shows left pleural effusion

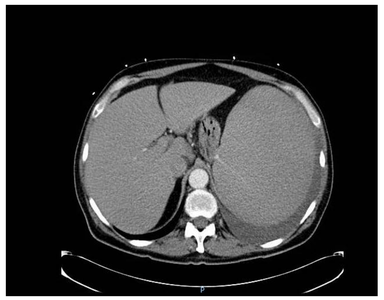

Fig 3a. Abdominal CT showing massive spleen **30 x 24 x 12 cm** , 3750 grams.

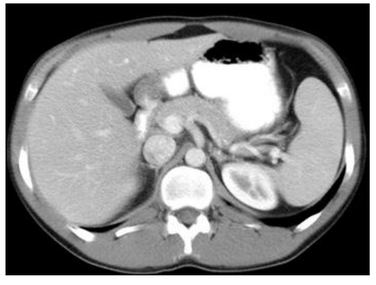

Fig 3b. Abdominal CT showing normal size spleen.

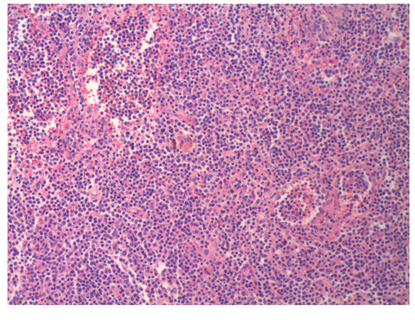

Fig 4. Pathology slide shows atypical proliferation of lymphoid cells with marginal zone pattern.

Discussion

Initial workup for splenomegaly:

- CBC can assess for cytopenia.
- CMP detects elevated direct and indirect bilirubin, which can be further investigated using hemoglobin electrophoresis, Coombs test, red cell enzyme testing, or osmotic fragility testing.
- Lymphocytosis on peripheral smear can lead to flow cytometry to diagnose CLL, hairy cell leukemia, or splenic marginal zone lymphoma.
- LDH indicates acute or chronic injury to tissue in NHL or AML.
- EPO level is low in polycythemia vera .
- Iron studies is abnormal in hemochromatosis.
- Amylase and lipase can be elevated in pancreatitis-like presentation in splenic vein thrombosis.
- Coagulation tests evaluate liver synthetic function.
- Hepatitis panel can evaluate for liver disease.

Splenic marginal zone B cell lymphoma

- Accounts for less than 1 % of all non-Hodgkin's lymphoma.
- Median age is 65 to 70 yo, it is uncommon to present before 50 yo.
- Patients present with splenomegaly, lymphocytosis and cytopenias.
- Lymphadenopathy and involvement of extralymphatic organs are uncommon.
- Treatment includes splenectomy, antibody treatment, chemotherapy, splenic irradiation and antiviral treatment.
- Prognosis score is based on hemoglobin <12 g/dL, LDH greater than normal, serum albumin <3.5g/dL.
- Median overall survival more than 10 years. It can transform into high grade lymphoma. Patients with aggressive tumors have median life expectancy of 18 months.

Conclusion

- Splenic marginal zone B cell lymphoma is rare in that it accounts for less than 1% of all non-Hodgkin's lymphomas.
- Splenic marginal zone B cell lymphoma has an indolent course and can be easily missed if patient remains asymptomatic.

References

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