A Case Report of T-cell Lymphoma Presenting with Massive Splenomegaly and Pancytopenia

Bipin Saroha MD[1], Karolyna Rey MD[1], Chandler Griffin[2], Babitha Thatiparthi MD[1], Osmany Deangelo DO[1]

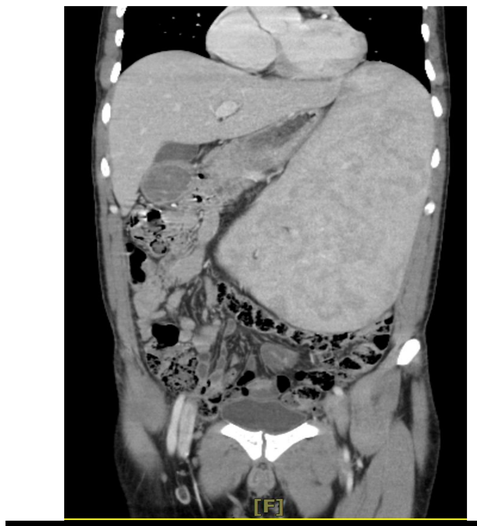
[1] Larkin Community Hospital, Department of Radiology, Miami, FL
[2] Lake Erie College of Osteopathic Medicine

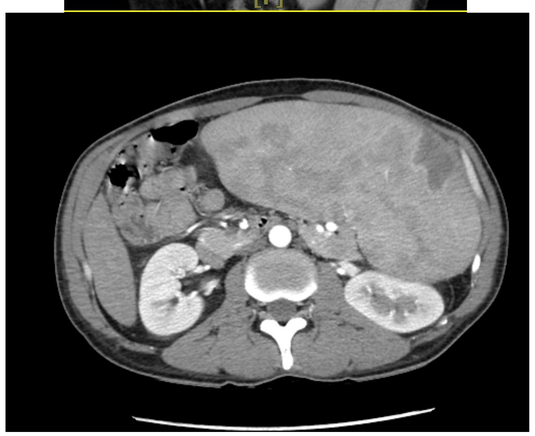
BACKGROUND

A 50-year-old male with no significant past medical history presented to our institution with severe weakness and fatigue. He noted right-sided chest pain since the past 3 months but because it was not limiting his daily activities, he decided not to see a doctor until he started to feel weak and fatigued. On physical exam the patient was awake, alert, and oriented to person, place and time. He appeared cachectic and his sclera were icteric. He did not appear to be in acute respiratory distress. His abdominal exam was significant for splenomegaly. His neurological exam showed no focal deficits. Admission CBC was significant for a hemoglobin of 4.3 g/dL, WBC 0.35 x 10^9/L, and platelet count of 88 x 10^9/L. Admission CMP was significant for total bilirubin of 3.81 mg/dL and direct bilirubin of 0.93 mg/dL. Due to the patient's significant anemia, he underwent a transfusion of 3 units packed RBCs and a pancytopenia workup was started. Bone marrow biopsy was completed and pathology showed hypercellular bone marrow with an atypical sinusoidal T-cell infiltrate. Multiple immunostains were performed with atypical cells positive for CD3 and CD7 with a loss of CD5 and CD2. The atypical cells also stained positive for TIA1, granzyme B, and TCR delta and highlighted a sinusoidal pattern of involvement. Negative stains included CD57, TCR Beta F1, CD10, BCL6, PD-1, and ICOS. Cellularity was approximately 95-100%. These factors combined led to a diagnosis of T-cell lymphoproliferative disorder with favoring of a hepatosplenic T-cell lymphoma.

IMAGING

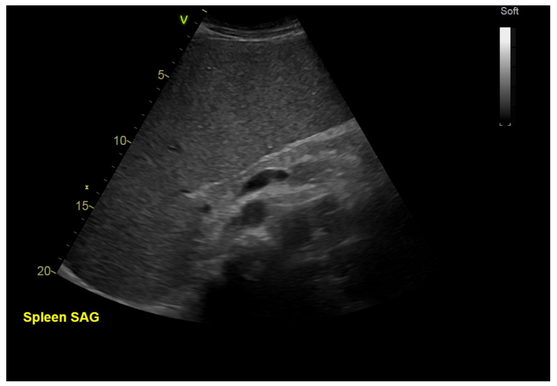
CT abdomen and pelvis with contrast showed massive splenomegaly measuring 28cm in length craniocaudal. Multiple peripherally located hypoattenuating wedge-shaped areas which may be secondary to regions of infarct. There was no evidence of occlusion of the splenic, superior mesenteric, or hepatic portal veins. There is no evidence of hemodynamically significant stenosis, occlusion, aneurysm formation, or vascular malformation in the visualized abdominal aorta, celiac or splenic arteries, SMA and IMA.





IMAGING (CONTINUED)

Ultrasound of the abdomen showed a liver of normal size measuring 16.3cm in length. The intrahepatic ducts were not dilated and the visualized portion of the portal vein demonstrates hepatopedal flow and measured 11mm in diameter. The common bile duct was not dilated and measured 3.4mm. The spleen was massive in size. Impression was splenomegaly without ultrasonographic evidence of space occupying lesions. There was no ultrasonographic evidence of hepatomegaly.



DISCUSSION

In 2016, there were approximately 72,000 new cases of Non-Hodgkin Lymphoma (NHL) diagnosed in the United States [1]. Of these, T-cell lymphoma are much rarer than B-cell lymphoma making up approximately 10-15% of all NHL [1]. Furthermore, hepatosplenic T-cell lymphoma is an exceptionally rare subtype of peripheral T-cell lymphomas representing less than 5% and only a few hundred cases are reported in literature [2]. Cases of hepatosplenic T-cell lymphomas typically present in young male patients and there is usually hepatosplenomegaly with thrombocytopenia.

DISCUSSION (CONTINUED)

This case further solidifies previous cases; the presentation and imaging of this rare disease presents with massive splenomegaly and pancytopenia. One of the most important differential diagnoses of hepatosplenic T-cell lymphoma is T-cell large granular lymphocytic leukemia. Certain criteria have been defined which help to differentiate between the two [3]. Some of the notable criteria for differentiating between the two include the presence of splenomegaly, higher bone marrow cellularity and the absence of CD57 which help to favor hepatosplenic T-cell lymphoma over T-cell large granular lymphocytic leukemia.

REERENCES

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CONTACT

Please send all questions to
bsaroha@larkinhospital.com
krey@larkinhospital.com