

CASE REPORT



Left lower quadrant pain: an unlikely diagnosis in a case of acute abdomen

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ABSTRACT

Splenic infarct is an incredibly rare diagnosis for abdominal pain. One study involving two hospitals over ten years describes only 0.0037% of all hospital admissions presenting with splenic infarction. Our report describes a case of massive splenomegaly causing pain in a different location compared to the normal anatomical location of the spleen with an unexpected cause, lymphoma. High clinical suspicion for lymphoma as a cause of splenic infarction is critical because this disease can otherwise be clinically silent. This case emphasizes the importance of a thorough clinical workup and persistence in diagnostic pursuit.

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Lymphoma; splenomegaly; spleen; infarction; oncology

1. Introduction

Splenic infarct is not a typical differential diagnosis for left lower quadrant (LLQ) abdominal pain. In this case report we present a patient with massive splenomegaly causing the pain in a different location compared to the normal anatomical location of the spleen. Splenic infarct is a rare cause of abdominal pain with a broad range of etiologies, each of which necessitates immediate workup and treatment. It is understood that the possible causes of splenic infarct include thromboembolism, hematologic malignancy, hypercoagulable states, and blunt abdominal trauma [1]. Although the potential causes are known, the actual incidence of splenic infarct and the incidence of each cause is debated between only a handful of studies. One of these is a study involving two hospitals that admitted approximately 1,300,000 patients over ten years, of which only 48 (.0037%) presented with splenic infarction, showing how incredibly rare it is as a diagnosis [2].

2. Case presentation

A 77-year-old male with a past medical history significant for hypertension and coronary artery disease status post stent placement 3 years ago presented with acute LLQ pain. His medications included Aspirin, Famotidine, Metoprolol, Niacin and Pravastatin. He presented with a 2–3 month history of progressive abdominal distension, a palpable mass on his mid to lower left abdomen, and a sensation of a pulled abdominal muscle. He described associated indigestion with burping, constipation, nausea, anorexia with an unintentional 16lb weight loss, and night sweats. Two days prior to admission the patient felt a sharp, localized pain in his left lower quadrant that

did not radiate and was progressively more painful especially when palpated, which he rated as a 7/10 in severity. He described tea colored urine intermittently over the last few months. He denied any fever, chills, sick contacts, recent travel, vomiting, diarrhea, swollen glands or lumps and had a negative history for atrial fibrillation, clotting incidents, or bleeding.

On physical exam he had a palpable mass spanning across his abdomen and into his left lower quadrant with extreme tenderness to palpation. He had no hepatomegaly or other significant findings on exam. The initial laboratory work up showed a white blood cell count of 6.1×10^3 cells/mcL, hemoglobin of 11.6 g/dL, hematocrit of 36.6%, a platelet count of 87×10^3 cells/mcL, sodium of 135 mEq/L, potassium of 4.2 mEq/L, and creatinine of .98 μ mol/L.

CT angiogram (CTA) showed massive splenomegaly with a large infarct of the anterior inferior pole of the spleen (Figures 1, 2). It also showed a 2.2 x 4.5 cm subcarinal mass thought to reflect lymphadenopathy as well as left and right hilar adenopathy with an enlarged pretracheal lymph node, ranging from 1 to 2.1 cm in diameter. With evidence of a splenic infarct, the patient was started on IV heparin in the emergency department for suspected embolism as a cause of infarct.

Hypercoagulable workup (Table 1) was negative for antinuclear antibody (ANA), Factor V Leiden and thrombin mutations. B Microglobulin was elevated at 6.04 mcg/mL (Normal: <2.51 mcg/mL), homocysteine was elevated at 19.4 μ mol/L (Normal: <11.4 μ mol/L), protein C levels were normal at 79% (Normal: 70–180%), protein S levels were low at 29% (Normal: 70–150%), cardiolipin IgM antibody was elevated at 54 MPL (Normal: <12 MPL) haptoglobin low at <30 mg/dL (Normal: 30–300 mg/dL), dRVVT



Figure 1. CTA of the chest, abdomen, and pelvis. Coronal section showing splenomegaly and splenic infarct (arrows) of the anterior inferior pole of the spleen.

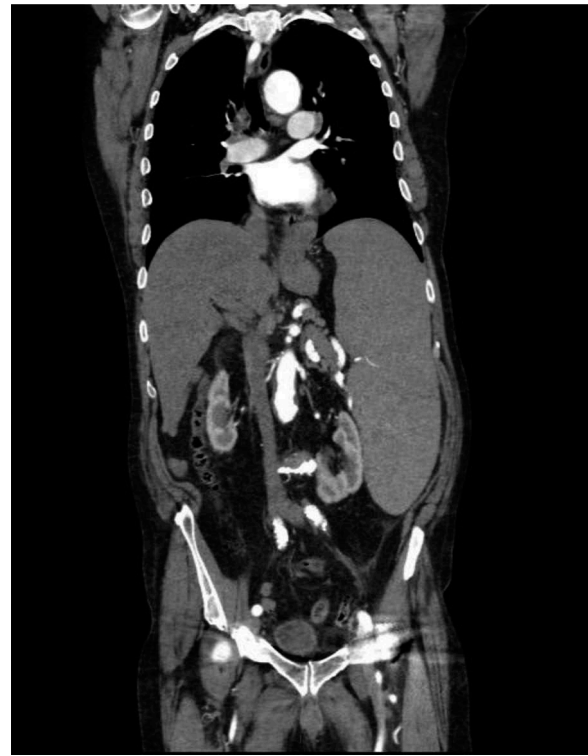


Figure 2. CTA of the chest, abdomen, and pelvis. Coronal section showing massive splenomegaly and compression of left kidney.

screen high at 46 seconds (Normal: <45 seconds) and LDH high at 477 IU/L (Normal: 98–225 IU/L).

Abdominal ultrasound of hepatic and splenic vessels was negative for emboli. CTA was negative for cardioembolic thrombi. Transthoracic and transesophageal echocardiograms were negative for a cardioembolic source of the infarct.

A peripheral blood smear showed only mild pancytopenia with atypical lymphocytes. Bone marrow biopsy was ordered and flow cytometry of the aspirate showed 110:1 kappa B cell population in 8.3% of the cells. The flow cytometry results showed the abnormal

B cells expressing CD45+, CD5-, CD10-, CD11b-, CD11c±, CD19+, CD20+, CD22+, CD23-, CD38-, CD103-, FMC7+, HLA-DR+, kappa+. A diagnosis of splenic marginal zone lymphoma was made. At this point in time, the patient is following up with oncology for treatment directed at the cell marker CD20.

3. Discussion

LLQ pain is an extremely uncommon presentation of splenic infarction. In the literature, the most common spleen-related cause of LLQ pain is wandering spleen,

Table 1. Diagnosis-focused work up for splenic infarct.

Rule Out Diagnosis	Test	Result
Cardioembolic Cause	CT Angiogram	No proximal or descending aortic thrombus
	Transthoracic Echocardiogram	No cardiac thrombus
	Transesophageal Echocardiogram	No cardiac thrombus
Hypercoagulable State	Factor V Leiden	Mutation not detected
	ANA Screen	Negative
	Cardiolipin IgM	54 MPL High (Normal: <12 MPL)
	dRVVT Screen	46 seconds High (Normal: <45 seconds)
	Protein Factor II Mutation	Negative
	Protein C	79% (Normal: 70–180%)
	Protein S	29% Low (Normal: 70–150%)
	Haptoglobin	<30 mg/dL Low (Normal: 30–300 mg/dL)
	Homocysteine	19.4 μmol/L High (Normal: <11.4 μmol/L)
	LDH	477 IU/L High (Normal: 98–225 IU/L)
	Abdominal Ultrasound	No venous thrombus
Autoinfarction Due to Malignancy	B Microglobulin	6.04 mcg/mL High (Normal: <2.51 mcg/mL)
	Peripheral Smear	Mild pancytopenia with atypical lymphocytes
	Bone Marrow Aspirate	Rare atypical lymphocytes
	Flow Cytometry	Monotypic Kappa B-cell population in 8.3% of the sample
	Cell Markers	CD45+, CD5-, CD10-, CD11b-, CD11c+/-, CD19+, CD20+, CD22+, CD23-, CD38-, CD103-, FMC7+, HLA-DR+, kappa+

which is a condition where the suspensory ligaments of the spleen are weakened or lost, allowing the spleen to move away from the left upper quadrant to anywhere within the abdominal cavity [3]. Not commonly described in the literature is a case of LLQ pain caused by massive splenomegaly. Neither splenic infarct nor lymphoma would have been on the differential diagnosis for LLQ pain if not for the initial CT scan in the emergency department, which showed the infarcted portion of the spleen. As is often the standard of care for severe acute abdominal pain, imaging will be obtained in these situations and the search for the cause of splenic infarct can begin. We hope this case will encourage keeping lymphoma high on the differential diagnosis of any splenic infarct.

The pathophysiology of splenic infarct from lymphoma is tissue necrosis and parenchymal ischemia [4] due to autoinfarction. The two primary causes of autoinfarction include rapid growth of the spleen leading to a loss of blood supply from branches of the splenic artery and venous congestion due to abnormal cells [5].

The most common presenting signs of lymphoma are enlarged lymph nodes, fever, sweating, chills, weight loss, swollen abdomen, early satiety, chest pain and shortness of breath [6]. Many of these are non-specific or can be attributed to the splenomegaly itself, making it important to keep lymphoma on the list of differential diagnoses even when the symptoms can otherwise be explained. As in our case, the patient's weight loss, anorexia, and hematuria were likely due to the extreme mass effect of the spleen compressing the stomach and kidney.

Our patient was found to have splenic marginal-zone lymphoma (SMZL), a small B-cell lymphoma involving the spleen and bone marrow that represents 1–2% of all lymphomas. This prevalence may be underestimated because until recently diagnosis was made with splenectomy specimens. Definitive diagnosis can now be made with immunophenotyping, genetic features, and cytologic composition. Patients typically present at 65 years of age with splenomegaly being the most common sign, observed in 75% of patients. Less common presenting symptoms include anemia, thrombocytopenia, leukocytosis, and autoimmune hemolytic anemia. Differential diagnoses include chronic lymphocytic leukemia, mantle cell lymphoma, follicular lymphoma, lymphoplasmacytic lymphoma, and MALT marginal zone lymphoma. Patients are typically diagnosed with SMZL at a late stage, as it almost always involves the bone marrow at the time of diagnosis [7].

The tests ordered for this patient were triaged to rule out any immediate life threatening conditions while collecting as much information as possible. The prospect of a cardioembolic source of infarct is

well documented as a source of splenic infarction originating from damaged valves, mural thrombi, left sided chamber abnormalities or plaques in the aorta [8]. This diagnosis carries with it the potential sequela of stroke and other harmful results of emboli in the arterial vasculature mandating it be ruled out immediately as the cause of infarct. In our case, the cardiac work up was both extensive and negative for every result.

Due to the massive splenomegaly, it was more likely that the spleen autoinfarcted from either malignancy or hypercoagulability. Splenic ischemia and infarct can occur when malignant splenic parenchyma outgrows its splenic arterial bloody supply or when abnormal cells cause venous congestion [5].

Markers for hypercoagulable states were easily available through blood draw, which made the information accessible while simultaneously doing a work up for a cardioembolic source. There were a number of abnormal results from the hypercoagulable work up, but many labs may have altered results due to heparin's anticoagulative effects [9]. Because of this and the lack of a visualized venous thrombus in the splenic vasculature on abdominal ultrasound or CTA, hypercoagulability was ruled out. Also, the elevated levels of B microglobulin, an MHC class I molecule on all nucleated cells and a marker for multiple myeloma and lymphomas, led us to look for a diagnosis of malignancy [10].

The results of the bone marrow biopsy allowed us to come to a final diagnosis of splenic marginal zone lymphoma after ruling out an urgent cardioembolic cause and the easily obtainable hypercoagulable work up. Bone marrow biopsy is an invasive procedure that was crucial for diagnosis in this case and potentially many others, with some studies showing up to 29% of splenic infarcts having undiagnosed origin [2]. Lymphoma needs to be ruled out in a timely manner because a biopsy and immediate treatment can significantly affect outcome in aggressive disease.

4. Conclusion

We aim to contribute to the medical literature by increasing awareness of splenic infarct as the first and primary presenting symptom of lymphoma. High clinical suspicion for lymphoma as a cause of splenic infarction is important because this disease can otherwise be clinically silent. Without a clear diagnosis and with resolution of symptoms, attaining a biopsy to rule out lymphoma is critical in providing thorough patient care.

'Symptoms, then, are in reality nothing but the cry from suffering organs.'

Jean-Martin Charcot

Disclosure statement

No potential conflict of interest was reported by the authors.

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