Case Report

Intravascular Large B-Cell Lymphoma Presenting with Diffuse Gallbladder Wall Thickening: A Case Report and Literature Review

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Intravascular large B-cell lymphoma is a rare subtype of extranodal diffuse B-cell lymphoma characterized by proliferation of neoplastic cells within the lumen of small and medium sized vessels. Clinical and imaging findings are nebulous as the intravascular subtype of lymphoma can involve a multitude of organs. Involvement of the gallbladder is extremely uncommon, and imaging findings can be easily confused for more prevalent pathologies. We report a case of intravascular large B-cell lymphoma in an 83-year-old male and review clinical presentation and imaging findings on CT, ultrasound, hepatobiliary iminodiacetic acid (HIDA) scan, and MRI. It is important for the radiologist to know about this disease as the imaging findings are atypical of other types of lymphoma, and this may lead to a delay in diagnosis and treatment.

1. Introduction

Intravascular large B-cell lymphoma is a rare subtype of extranodal diffuse B-cell lymphoma characterized by proliferation of neoplastic cells within the lumen of small and medium sized vessels. Clinical and imaging findings are nebulous as the intravascular subtype of lymphoma can involve a multitude of organs. Involvement of the gallbladder is extremely uncommon, and imaging findings can be easily confused for more prevalent pathologies. This case is a reminder of a rare but important diagnostic consideration in the setting of a diffusely thickened gallbladder wall.

2. Case Presentation

An 83-year-old Caucasian male with past medical history of hypertension, diabetes, and coronary artery disease presented to the emergency department with extreme fatigue, weakness, and disorientation. He reported a history of 10–15-pound weight loss over the previous month. Physical exam did not yield any focal signs. Laboratory evaluation revealed pancytopenia (Hgb 9.6 g/dL, WBC 3.0 bil/L, and platelets 169 bil/L) and increased liver function enzymes (AST 311 U/L, ALT 62 U/L, and ALP 237 U/L). The patient was admitted for generalized weakness and fatigue.

An unenhanced CT scan of the abdomen and pelvis obtained in the emergency department showed a nondis tended gallbladder with diffuse wall thickening (Figure 1). A small calcified gallstone was identified in the neck of the gallbladder. Neither a soft tissue mass nor lymphadenopathy was present. Although acute cholecystitis was not a leading differential diagnosis based on the clinical presentation, HIDA scan was recommended based on the unexpected CT findings.

The patient developed worsening alteration of mental status, metabolic acidosis, further increase in liver function tests, and pancytopenia. D-dimer was elevated (1830 ng/mL). There was initially clinical concern for pulmonary embolism versus disseminated intravascular coagulation. Nuclear medicine testing with ventilation-perfusion (VQ) scan was performed and low probability for pulmonary embolism was found. Coagulation testing remained stable.

The HIDA scan revealed cystic duct patency (Figure 2). Subsequently, a targeted sonographic examination of the
Figure 1: Axial unenhanced CT of the abdomen obtained at presentation shows a diffusely thickened and low attenuation gallbladder wall.

Figure 2: Image obtained during HIDA scan shows early filling of the gallbladder by 30 minutes which was confirmed on delayed 40- and 60-minute images.

Figure 3: Grayscale longitudinal sonographic image of the right upper quadrant shows a decompressed gallbladder wall with diffuse wall thickening. The gallbladder wall has a striated appearance without increased vascularity.

The mucosa was uninvolved. These features explained all the imaging findings well in this particular case.

Flow cytometry of the peripheral blood revealed a small population of aberrant B-cells which correlate to the pathologic gallbladder wall findings. Subsequent bone marrow core biopsy and aspiration were performed. Pathologic evaluation demonstrated slight hypercellularity of the marrow and 10–15% involvement by diffuse large B-cell lymphoma. Immunohistochemical stain of bone marrow core biopsy sample was positive for CD79a, confirming B-cell origin.

Postoperatively, the patient’s condition continued to deteriorate requiring respiratory support in the intensive care unit. He developed severe metabolic acidosis and hemodynamic instability. Given the patient’s poor functional status and concomitant diagnosis of IVLBCL, the decision to withdraw supportive care was made by his family. The patient expired 7 days after initial presentation.

3. Discussion

Intravascular B-cell lymphoma is a highly aggressive form of extranodal B-cell lymphoma characterized by neoplastic cell growth within the lumen of small sized vessels [1]. It presents as a disseminated aggressive disease without focal mass or lymphadenopathy [2]. Neoplastic vascular involvement can affect all types of organs, such as bone marrow, CNS, skin, lung, adrenal gland, liver, kidney, spleen, thyroid, pituitary gland, and gastrointestinal tract [3]. In our patient, no additional lesions were identified in the base of the lungs, abdomen, or pelvis. No cross-sectional imaging was performed on the chest or brain during this hospitalization, so involvement of the lungs or brain cannot be excluded. Overall, findings in this case are consistent with secondary IVLBCL of the gallbladder wall. IVLBCL presenting with gallbladder symptoms is very rare with only a handful of cases described in the literature [4]. Common clinical presentation includes nonspecific systemic symptoms, such as fever of unknown origin, fatigue, deterioration of performance status, and neurological alteration.

This case highlights the diagnostic conundrum of IVLBCL. In our case, the patient experienced nonspecific intermittent altered mental status and fatigue without focal
abdominal pain. Furthermore, imaging evaluation revealed only nonspecific findings of striated gallbladder wall thick-ening. The lack of lymphadenopathy in patients with IVLBCL also makes timely diagnosis challenging. Additionally, staging of this disease is difficult secondary to the lack of discrete mass.

To our knowledge, the multimodal imaging characteristics of IVLBCL affecting the gallbladder have not been described in the literature. Diffuse gallbladder wall thickening is nonspecific. It can be secondary to primary gallbladder pathology such as acute and chronic cholecystitis, adenomyomatosis, or gallbladder carcinoma [5]. It may also be a manifestation of a host of conditions extrinsic to the gallbladder such as hypoalbuminemia, congestive heart failure, hepatitis, renal failure, and physiologic contraction after a recent meal. A thickened gallbladder wall with a striated sonographic appearance (thick echogenic wall with a homogenous sonolucent central band) is more characteristic of benign noninflammatory conditions [6].

MRI evaluation is helpful in characterizing gallbladder wall thickening as those cases which show early and pro-longed enhancement tend to be malignant whereas benign etiologies generally have less rapid and prolonged enhance-ment [7]. However, our case did not follow this concept. With vessel occlusion by intravascular neoplastic cells, one would expect to find decreased postcontrast enhancement as seen in our case. The lack of a focal mass, lymphadenopathy, and postcontrast enhancement make for an unusual imaging constellation for lymphoma [8]. While findings of neurologic alteration, pancytopenia, and elevated liver function studies have been described in cases of IVLBCL, they are not specific [3]. Additional potentially useful imaging, not obtained in this case, includes diffusion weighted imaging and positron emission tomography (PET). Diffusion weight imaging (DWI) would likely show abnormally restricted dif-fusion due to the presence of high grade malignant cells. PET would likely show increased FDG activity as with large B-cell lymphoma elsewhere. Unfortunately, these findings overlap with acute cholecystitis and would not help in distinguishing lymphoma from acute inflammation.

IVLBCL is a highly aggressive neoplastic proliferation of B-cells within medium and small sized vessels. The clinical and laboratory findings are not specific, and the diagnosis is often delayed. Furthermore, the imaging findings
in our case highlight the diagnostic challenge of IVLCL. The lack of a focal mass, lymphadenopathy, and abnormal contrast enhancement misleadingly point to a nonneoplastic process. Furthermore, diffuse gallbladder wall thickening with a trilaminar appearance is most characteristic of benign noninflammatory conditions, which would not prompt tissue diagnosis. A benign condition such as diffuse adenomyomatosis with numerous Rokitansky-Aschoff sinuses could be considered, but this case had no cystic changes and the trilaminar appearance of the gallbladder would be an unusual imaging finding. Although no diagnostic imaging features of IVLCL exist, with increased awareness, the antemortem identification of this elusive diagnosis may be suggested.

Consent

Written informed consent from the patient for submission of this manuscript for publication was not obtained. No identifying information is included.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

Authors’ Contributions

Sayf Al-Katib is the primary author of this manuscript. Robert Colvin was a contributing author and assisted in write-up and editing. Farnoosh Sokhandon is a contributing author and provided guidance in the write-up.

References

