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CASE REPORT | LIVER

Intracystic Hemorrhage Complicating Polycystic Liver Disease in a 90-Year-Old Woman

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ABSTRACT

Nonparasitic hepatic cysts are common benign tumors that are often asymptomatic and incidentally discovered on imaging. Intracystic hemorrhage is a rare complication of hepatic cysts. We review the literature and discuss a case of intracystic hemorrhage in a 90-year-old woman with polycystic liver disease. The patient underwent cyst aspiration and percutaneous drain placement with subsequent resolution of symptoms. To our knowledge, we report the oldest patient to present with hemorrhage into a hepatic cyst. This case presents unique challenges in management, both because of the patient's age and because of her polycystic liver disease.

INTRODUCTION

Simple hepatic cysts are benign lesions that are often asymptomatic and discovered incidentally on imaging, with a prevalence of 5%–15%.[1,2] Mass effect and compression of adjacent structures are the most frequent complications, and most patients present with abdominal pain.[3] In rare cases, they can be complicated by intracystic hemorrhage. After an extensive literature search, we identified 23 publications describing 27 cases of nonruptured hemorrhagic hepatic cysts and only 3 cases in patients with polycystic liver disease (PCLD). The mean age at presentation was 60 years. We report a case of intracystic hemorrhage in a 90-year-old woman with isolated PCLD. This is the oldest patient reported to present with a hemorrhagic hepatic cyst.

CASE REPORT

A 90-year-old woman with chronic thrombocytopenia, heart failure, and atrial fibrillation on warfarin presented with acute-on-chronic abdominal pain. She had multiple hepatic cysts, which were incidentally discovered 9 months previously. The largest cyst was 11 cm. She presented with mild abdominal discomfort that had acutely progressed to sharp right upper quadrant abdominal pain. Two months previously, she underwent transcatheter aortic valve replacement for aortic stenosis and was started on clopidogrel.

The patient was hemodynamically stable and had a large, palpable mass in the right upper quadrant with severe localized tenderness to palpation. Liver function tests, hemoglobin, and leukocytes were within normal limits, platelets were 42,000/mL, and international normalized ratio was 1.5. Computed tomography angiography revealed a polycystic liver with a dominant 11-cm exophytic cyst containing dependently layering hyperdense material consistent with intracystic hemorrhage (Figure 1). There was no active extravasation.

The patient's warfarin was held, and she was given 1 unit of fresh-frozen plasma and platelets. Given her age and multiple medical comorbidities, she was deemed a poor surgical candidate. Because the patient continued to have severe abdominal pain refractory to

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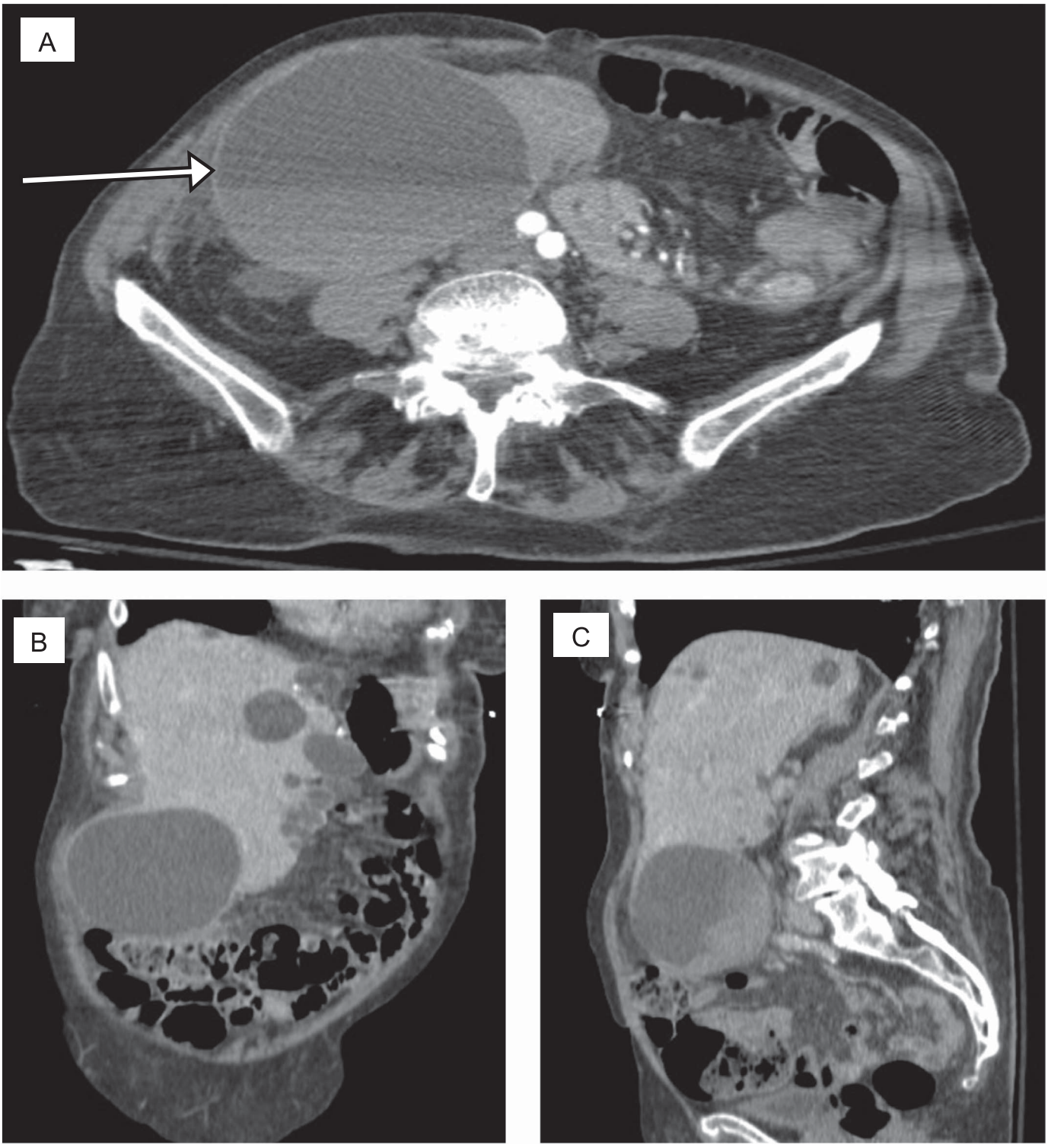

Figure 1. (A) Abdominal computed tomography angiography revealed an 11 × 9 cm exophytic cyst arising from the inferior right hepatic lobe containing dependently layering hyperdense material (arrow), consistent with intracystic hemorrhage. (B) Coronal and (C) sagittal views provide visualization of the patient's polycystic liver.

analgesics, the decision was made to proceed with drainage. Interventional radiology placed a percutaneous drain under ultrasound and fluoroscopic guidance and aspirated 500 mL of blood from the cyst (Figure 2). The patient had subsequent resolution in symptoms and remained hemodynamically stable with stable hemoglobin. She then returned to interventional radiology for cyst sclerotherapy 2 days later. Contrast injection into the cyst under fluoroscopy demonstrated extravasation,

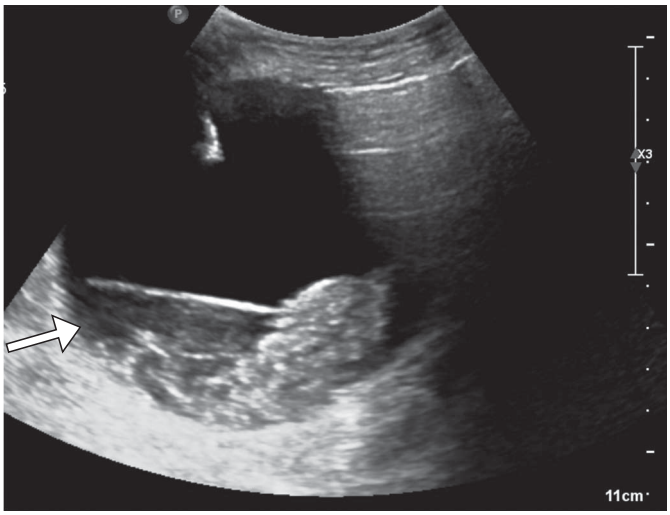

Figure 2. Ultrasound imaging of the cyst during percutaneous drain placement showing dependently layering hyperechoic material consistent with intracystic hemorrhage (arrow).

precluding sclerosis due to the concern of sclerosant extravasation (Figure 3). The drain was left in place for planned reattempt at sclerotherapy in 1 month. In the interim, the patient's drain became dislodged. One month later, ultrasound showed that the cyst had decreased to 7.8 × 7.0 cm and had minimal residual hyperechoic material, consistent with drainage and resorption of the previously seen hemorrhage (Figure 4). She remains asymptomatic 11 months later.

DISCUSSION

Nonparasitic benign liver cysts are usually asymptomatic and can be congenital or acquired, possibly arising from biliary

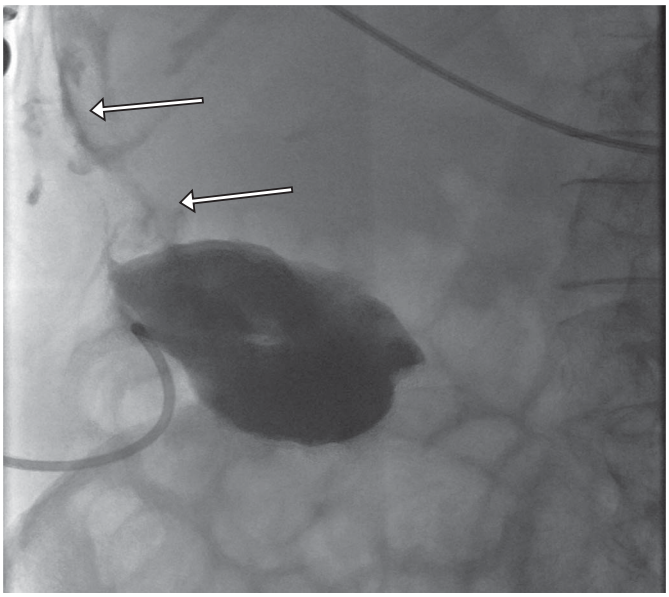

Figure 3. Fluoroscopic imaging during contrast injection of the drain showing contrast extravasation (arrows) from the cyst coursing along the lateral aspect of the liver.

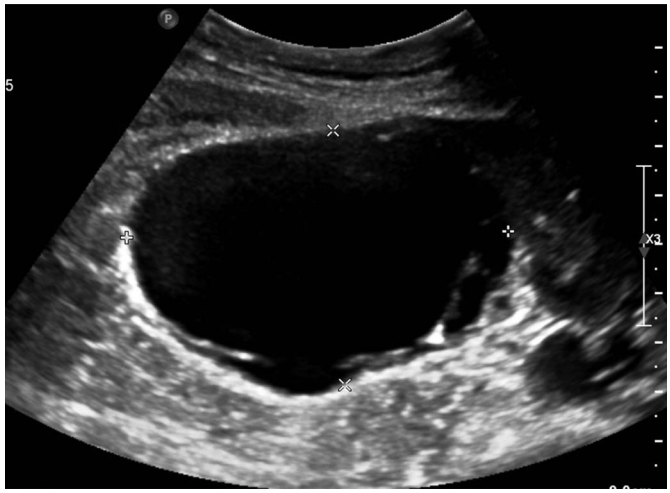

Figure 4. Ultrasound imaging showing persistent but decreased fluid in the cyst with minimal residual hyperechoic material, consistent with near-complete resolution of the previously seen blood within the cyst.

microhamartomas that lose their connection to the biliary tree.[4] They may enlarge over time as the epithelial lining secretes fluid into the cyst. Intracystic hemorrhage is a rare complication of hepatic cysts that may be due to increased pressure leading to cyst wall necrosis and sloughing of small blood vessels.[5] There are also reports of iatrogenic causes secondary to the initiation of anticoagulant or antiplatelet medication.[6–8] This likely played a role in our patient because she was on warfarin and recently started on clopidogrel.

Common presenting symptoms of hemorrhagic hepatic cysts include abdominal pain, palpable abdominal mass, and obstructive jaundice.[9] Diffuse peritoneal signs, hemodynamic instability, and progressive anemia are signs of hemorrhagic cyst rupture, which is a rare, life-threatening complication that occurs preferentially in patients with PCLD.[10] Thirteen cases of hemorrhagic rupture have been reported in the literature, resulting in 4 deaths, whereas no deaths secondary to intracystic hemorrhage without rupture have been reported.[6,7,10] Preventing rupture is therefore the top priority in the acute management of contained intracystic hemorrhage. Computed tomography angiography should be performed to assess for extravasation, which increases the risk of rupture and should be treated with urgent angioembolization.

Minimizing recurrence has proven to be one of the major challenges in the management of hepatic cysts. Loehe et al observed a 67% and 41% symptomatic recurrence rate in patients with PCLD and simple liver cysts after surgical unroofing, resection, or transplant.[11] Surgical unroofing is the definitive choice for single symptomatic hepatic cysts because it effectively relieves symptoms, has a low recurrence rate, and allows for analysis of the cyst wall.[9,12,13] However, percutaneous drainage followed by injection of sclerosing agents is the preferred option, particularly for poor surgical candidates or those who prefer a less invasive approach. It has been reported that this is as effective as

Table 1. Gigot classification of patients with adult polycystic liver disease[15]

Type	Definition
Type I	Limited number (<10) of large cysts (>10 cm)
Type II	Diffuse involvement of the liver parenchyma by multiple medium-sized cysts with remaining large areas of the noncystic liver parenchyma
Type III	Massive, diffuse involvement of the liver parenchyma by small- and medium-sized liver cysts, with only a few areas of the remaining normal liver parenchyma between cysts

laparoscopic unroofing but with a lower incidence of complications.[12,14] Sclerotherapy works by damaging the cyst epithelium and preventing additional fluid secretion into the cyst. Benzimra et al observed complete resolution of symptoms after percutaneous drainage and sclerotherapy in 83%, 80%, and 67% of patients with simple cysts, hemorrhagic cysts, and underlying PCLD, respectively.[12] The groups were not significantly different, and the mean follow-up period was 27 months.

Surgery is indicated when malignancy is suspected, in the presence of a communication between the cyst and biliary tree, and in cases of failed nonoperative management. When approaching these patients, it is helpful to use the Gigot classification as a guide (Table 1).[15] In a subset of patients with Type I PCLD who are symptomatic from 1 dominant cyst, laparoscopic unroofing is the procedure of choice. In patients with Type II and III PCLD presenting with symptomatic hepatomegaly, hepatic resection and cyst fenestration in the remnant liver is a more appropriate option.

Our patient had resolution of symptoms after percutaneous drainage and remains asymptomatic 11 months later. This supports previous reports suggesting that percutaneous drainage and sclerotherapy is safe and effective and should be performed as the first-line treatment of symptomatic hepatic cysts, even in the presence of PCLD and/or acute intracystic hemorrhage.[12,14] Patients with suspected malignancy, communication between the cyst and biliary tree, or recurrence of symptoms after percutaneous drainage and sclerotherapy should undergo surgery.

DISCLOSURES

Author contributions: AC Purdy performed the literature review and wrote the manuscript. A. Grigorian, D. Fernando, J. Nahmias, and AN Demirjian edited the manuscript. AN Demirjian is the article guarantor.

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Informed consent was obtained for this case report.

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