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Case Report

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Hepatic Cysts with Cholesterol Crystal Formation: An Unusual Mimicker of Cyst Infection in A Patient with Autosomal Dominant Polycystic Kidney Disease

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1. Abstract

Hyperechoic echogenicity in a hepatic cyst is a sign of cyst infection. We report a case of autosomal dominant polycystic kidney disease with polycystic liver disease where the hepatic cyst showed glittering spots. Cyst fluid analysis revealed them as cholesterol crystals.

2. Introduction

The overall prevalence of hepatic cysts in patients with Autosomal Dominant Polycystic Kidney Disease (ADPKD) has been reported as 83% [1]. The complications of hepatic cysts include infection, hemorrhage, rupture, and torsion of the cysts [2]. Computed Tomography (CT) characteristics of infected hepatic cysts include pericystic hyperemia and higher intracystic attenuation than normal cysts. The ultrasound features of infected hepatic cysts include intracystic hypoechoic or hyperechoic or heterogeneous echogenicity [3]. Hepatic cysts with cholesterol crystals is a rare condition. The ultrasound features of these cysts are hyperechoic sludge with a glittering appearance that mimics cyst infection. Clinicians should be aware of this rare association.

3. Case Report

We report a 50-year-old woman with ADPKD with Polycystic Liver Disease (PLD). Written informed consent was obtained from the patient in all invasive procedures. She received alcohol sclerosing therapy for liver cysts in 2016 and left lobe hepatic cyst aspiration due to cyst infection in 2018. Upon admission, she had been suffering from abdominal pain and distension for 2 weeks. A liver ultrasound showed sludge in one of the cysts of the left lobe (Figure 1). The sludge showed a glittering appearance when we tilted the probe (Video 1). Because the ultrasound finding mimicked that of an infected cyst, we performed aspiration. The fluid was cloudy brown (Figure 2) and was sent for Gram stain, culture, total bilirubin, crystal analysis, and total cholesterol. The Gram stain did not reveal the presence of any organisms and no aerobes or anaerobes were isolated upon culture of the cyst fluid. The total bilirubin of the cyst was 0.80mg/dL and the total cholesterol was 168 mg/dL. Crystal analysis revealed multiple cholesterol crystals showing typical rectangular-shaped crystals with notched corners under light microscopy (Figure 3). The crystals were birefringent under polarized light microscopy (Figure 4). The patient underwent right lobe hepatic cyst Transcatheter Arterial Embolization (TAE) in 2019. Thereafter, her abdominal pain improved and the abdominal distension subsided. The whole liver cysts volume reduced by 10%.

4. Discussion

Hepatic involvement is the most common extrarenal manifestation in ADPKD. The overall prevalence of hepatic cysts in patients with ADPKD was reported as 83% with a prevalence of 85% in women and 79% in men. The older group had the highest prevalence (94%) and the younger group had the lowest prevalence (58%) [1]. A very large polycystic liver may cause abdominal distension and abdominal pain. Symptoms related to adjacent organ compression include easy satiety due to gastrointestinal compression, dyspnea due to diaphragm compression, and abdominal hernia or uterine prolapse due

to increased intra-abdominal pressure. The end stage of the disease is characterized by malnutrition, disability, and, finally, physical exhaustion [2]. The hepatic complications of ADPKD include infection, hemorrhage, rupture, and torsion of the cysts. Multiple liver cysts cause a mass effect, and complications include hepatic venous outflow obstruction, Budd–Chiari syndrome, inferior vena cava compression, portal hypertension with variceal bleeding, and bile duct compression with jaundice [2].

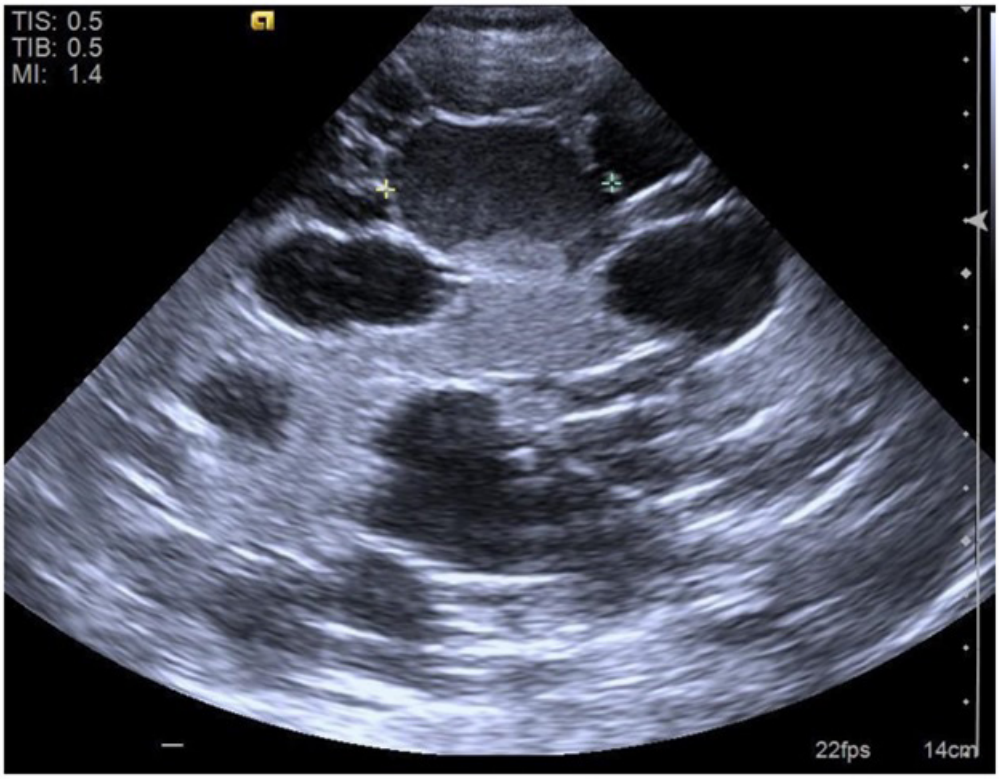

Figure 1: Liver ultrasound of an ADPKD patient with PLD. A cyst 4.2cm at left lobe showed hyperechoic sludge with glittering spots.

Figure 2: Cloudy brown cyst fluid was aspirated from the cyst with sludge and glittering spots.

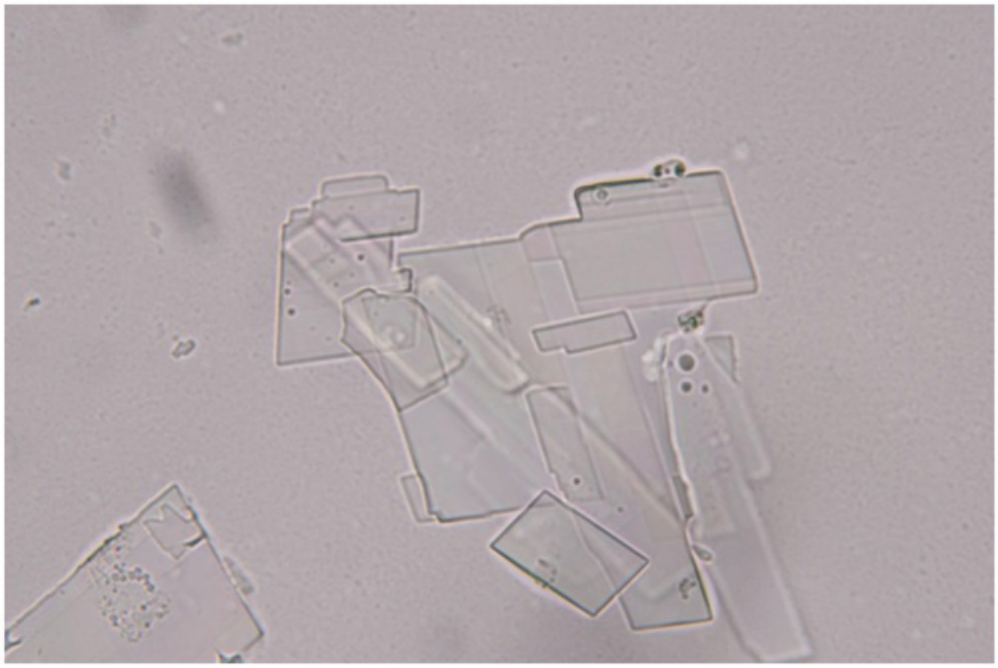

Figure 3: Crystal analysis of the cystic fluid revealed typical rectangular-shaped crystals with notched corners under light microscopy, indicating cholesterol crystals.

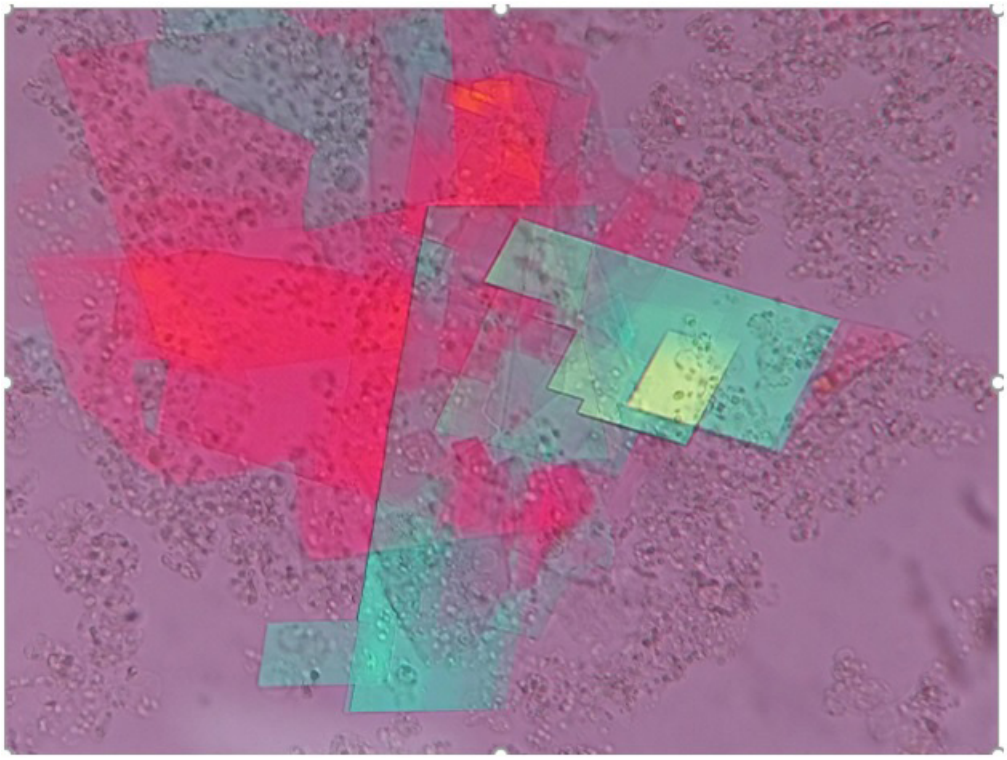

Figure 4: Under polarized light microscopy, the cholesterol crystals showed bifringence.

The initial treatment for PLD and ADPKD includes the termination of oral anticonceptives, conservative treatment for asymptomatic patients, and analgesic administration for patients with acute or chronic pain or tenderness. Other invasive treatments include percutaneous aspiration combined with sclerotherapy, installation of a stent for compression syndrome, laparoscopic fenestration, liver resection, and liver transplantation [2]. In several case series studies have reported TAE for treatment of the PLD as an acceptable treatment option for ADPKD patients [4]. Furthermore, 83% of patients experience an improvement in symptoms like abdominal distension and pain 2–6 months after TAE. The mean liver cystic volumes were reported as reduced by 33%, and liver parenchyma volumes increased by 35% at 36 months after TAE. TAE is a safe procedure without major complications. Post-treatment syndrome is self-limiting and includes

the development of abdominal pain, low-grade fever, and loss of appetite within 1 week of TAE.

A complication in ADPKD with PLD is hepatic cyst infection, which occurs in approximately 1% of all patients with hepatic cysts. The most common organisms isolated from blood culture or aspirated hepatic cysts fluid are *Escherichia coli* and *Klebsiella* sp., indicating that the bacteria have translocated across the intestinal barrier [5]. The symptoms of hepatic cyst infection include right upper-quadrant pain, fever, and malaise. Sepsis may occur without adequate treatment. CT shows infected hepatic cysts with pericystic hyperemia and higher intracystic attenuation than normal cysts (median, 19.0 Hounsfield unit (HU) vs. 8.5 HU). The ultrasound features of infected hepatic cyst lose their anechoic property and present intracystic hypoechoic or hyperechoic or heterogeneous echogenicity [3].

Cholesterol crystals in hepatic cyst is a rare condition, and has not been reported in the literature. The crystals showed hyperechoic echogenicity that mimicked cyst infection. The glittering appearance under ultrasound may be related to the flat, flaky crystal structure of cholesterol. Clinicians should be aware of this rare association.

5. Acknowledgements

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