Polycystic Liver Disease with a Large Intracystic Papillary Projection: Image-Pathologic Correlation

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Complicated cysts are not uncommon in patients with polycystic liver disease (PCLD) but prominent papillary projection within the cyst rarely occurs which may mimic malignant transformation, especially when there is progressively elevated serum tumor markers. We described such an unusual presentation in a patient with PCLD who had fever, diarrhea, and progressive elevated serum CA 19-9 and CEA. The imaging findings reflected the pathologic features of the papillary projection in a complicated cyst, featuring predominant fibrous stroma and some small bile ducts which appeared as sonographically anechoic tiny cystic spaces. These small bile ducts were consistent with hyperintense dots on T2-weighted magnetic resonance (MR) images. The findings suggested bile duct origin of the papillary projection. The clinical symptoms disappeared after operation. Familiarity with these imaging findings may be helpful in making differential diagnosis preoperatively.

Key words: Liver, cyst; Liver, Neoplasm; Magnetic resonance; Ultrasonography

Polycystic liver disease (PCLD) is an autosomal dominant inherent disease, which is characterized by the presence of multiple bile duct-derived epithelial cysts scattered in the liver parenchyma [1]. PCLD can be associated with polycystic kidney disease [1-3]. The imaging findings usually reveal an enlarged and diffusely cystic liver with the cysts varying from less than 1 mm to 10 cm or more in diameter. Infection or hemorrhage of the complicated cysts is not uncommon but intracystic papillary projection is an unusual presentation in PCLD [3, 4]. We describe the imaging findings of a patient with PCLD presenting as a large papillary projection in one of the large hemorrhagic infected cyst, which was suspected to be malignant transformation. There were characteristic features comparable with histopathologic findings on both ultrasound and magnetic resonance (MR) imaging.

CASE REPORT

A 47 year-old woman complained of watery diarrhea for several months and persistent high fever (up to 39°C) in recent one year. She had a family history of autosomal dominant polycystic kidney disease (ADPKD). Gradual elevation of serum tumor markers CA (cancer antigen) 19-9 from 42.3 to 240 u/ml and tumor marker CEA (carcinoembryonic antigen) from normal to over 500 ng/ml within nine months. She had body weight loss for 8 kilograms and progressively abdominal distension within one year. Laboratory findings revealed leukocytosis (21.77 k/ul) with neutrophil predominance at admission. Under the impression of infected cysts, she received intravenous antibiotic treatment (ciprofloxacin 200 mg every 12 hours). The white count declined to 12.43 k/ul but low grade fever persisted.

Abdominal ultrasound examination showed multiple variable-sized cysts in the liver, compatible with PCLD. Bilateral smaller renal cysts were also
noted. A large papillary projection (12 cm in diameter) was found in one large hepatic cyst at the inferior portion of the right lobe liver (Fig. 1). She also received computed tomography (CT) (Fig. 2) and magnetic resonance (MR) study (Fig. 3). The intracystic mass was not visible on CT. On T2-weighted MR images, a large intracystic papillary projection was seen with some small bright dots in the stroma (Fig. 3). The dot-like structures were invisible on T1-weighted MR images (not shown). The signal of the cystic content was bright in T1-weighted images which were suggestively of a hemorrhagic cyst. Due to progressive elevated serum level of tumor markers and unusual imaging findings for PCLD on ultrasound and MRI, she underwent surgical intervention under the impression of PCLD with suspected malignant transformation of the cystic wall. During operation, turbid yellowish brown fluid within the cyst was found in the large hepatic cyst which was located at the inferior portion of the right lobe liver. Trisegmentectomy for segment 5, 6, 7 and fenestration of hepatic cysts were performed smoothly. It revealed no evidence of malignancy and confirmed the diagnosis of an infected and hemorrhagic hepatic cyst in PCLD in histopathological study. The large papillary projection was composed of abundant fibrous stroma and some scattered bile ducts (Fig. 4). There was severe inflammation with infiltration of neutrophils and mononuclear cells coated with fibrinoid degenerated materials. There was cuboidal epithelium and abundant fibrous stroma with scattered small bile ducts (Fig. 5). Finally, PCLD with papillary projection due to fibroproliferation in a complicated

Figure 1. Abdominal sonography, right subcostal plane, reveals a large papillary lesion in the cyst at right lobe liver. Note the small anechoic spaces (arrows) within the echogenic stroma of the papillary mass, comparable with small bile ducts.

Figure 2. Contrast-enhanced CT scan of abdomen shows typical appearance of polycystic liver disease with some renal cysts. The papillary lesion cannot be visualized in the same cyst shown on Figure 1.

Figure 3. Axial fast spin-echo T2-weighted image (TR/TE 6300/105) shows a distinct papillary lesion in the largest cyst. There are some scattered small bright dots (arrows) in the papillary stroma, comparable with small bile ducts.
A cyst was considered. She recovered without symptoms after operation and was followed up regularly in outpatient clinic two years after operation.

**DISCUSSION**

PCLD occurs in association with ADPKD (71-93%) or as an isolated disease [1-3]. The most common form of autosomal dominant PCLD coexists with ADPKD and is linked to mutations in either PKD1 or PKD2 [1]. PCLD is typically a benign disease which is rarely associated with hepatic malignancy. However, there are some complications including infection, perforation, spontaneous hemorrhage, obstructive jaundice and neoplastic degeneration. Intrahepatic malignancy from hepatic cysts was rarely reported [5, 6]. Lin demonstrated adenocarcinoma and infection in a solitary hepatic cyst with significant elevated level of tumor marker CA 19-9 and CEA in the cystic fluid [6]. In their reported case, the tumor presented as papillary growth on the cystic surface [6]. In contrast to the reported case, intracystic papillary projection in PCLD can mimic malignant tumor even though clinical evidence of infection and marked elevation of tumor markers.

Typical histologic findings of the cysts in patients with PCLD are similar to simple hepatic cyst. These cysts consist of cuboidal epithelium originating from bile duct. Classic MR appearance of hepatic cysts is well known with a well-defined, discernible thin wall, occasionally thin internal septa, and very bright signal intensity comparable with cerebrospinal fluid (CSF) on T2-weighted images [3]. Cysts of PCLD have similar appearance as simple cysts on MRI and CT, but numerous cysts in variable sizes are usually characteristic for making the diagnosis [3]. As shown in this case, sonography and MRI are more sensitive for detecting internal septa and intracystic lesion than CT. Infection or hemorrhage of the cysts is not uncommon in PCLD [1, 3]. MRI is more useful than CT in demonstrating different signal in the hemorrhagic cyst [3]. The intracystic fluid of a hemorrhagic cyst shows bright T1 and T2 signal but can be hypodense on CT [3, 4].

In this report, sonographic and MR findings of the papillary mass in the large hemorrhagic and infected cysts reflected the features of the pathologic specimen. The histologic findings of the papillary projection included abundant fibrous stroma and some scattered bile ducts, which were suggestive of its bile duct origin. The small T2-bright dots in the papillary projection of hepatic cyst represented bile ducts which were presumed to be a sign of benign lesion in this particular condition. The etiology of the presence of the large intracystic mass was not clear because no prior imaging study was available for comparison. It was presumed to be inflammatory mass developed in the infected cyst. Sonography is not sensitive in detecting intracystic hemorrhage as compared with MR study. CT is less sensitive than ultrasound in detecting intracystic papillary lesion in PCLD.

Though there is no effective medical therapy, surgical intervention, including liver resection and fenestration, is indicated for infected or symptomatic cysts in patients with PCLD [7, 8]. As in our reported case, this patients recovered completely after resection.
of hepatic cysts and fenestration.

In summary, sonographic and MR studies of the large papillary projection in the cyst of PCLD reflected its histopathologic features and was suggestive of its bile duct origin. Familiarity with the imaging findings may be helpful in making differential diagnosis preoperatively.

REFERENCES

多囊性肝疾病含一巨大囊内乳突状突起：影像-病理之对照

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多囊性肝疾病有合併症水囊並非罕見，但罕以囊內明顯乳突起表現，尤其又有逐漸上升之血液腫瘤標記，此時可以惡性轉化。我們描述此種影像於囊性肝疾病病人，臨床上以發燒、腹瀉及逐漸升高之CA 19-9與CEA表現。此病例之影像表現表現了病理特徵，即主要纖維基質中含一些小膽管，其對照於超音波下是無回音的微小囊腫及磁振造影中是高訊號亮點，因此推測囊內乳突應源自膽道。病人在術後症狀消失。我們認為瞭解影像所見有助於術前之鑑別診斷。

關鍵詞：囊腫；感染；肝臟；磁振；超音波