

# Monosegmental Hepatobiliary Fibropolycystic Disease Mimicking a Mass: Report of Three Cases

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Hepatobiliary fibropolycystic diseases are a unique group of entities involving the liver and biliary tract, which are caused by abnormal embryologic development of the ductal plates at various stages. We experienced strange hepatobiliary fibropolycystic diseases with a complex mass composed of malformed ducts and biliary cysts, which did not belong to, and were different from, previously known malformations. They were unique in imaging and histologic features. We herein report three cases of monosegmental hepatobiliary fibropolycystic disease mimicking a mass.

**Index terms:** Ductal plate malformations; Fibropolycystic disease; Monosegmental disease; Caroli disease

## INTRODUCTION

The ductal plate is a double-layered cylindrical bile duct structure present in early fetal life. Biliary ducts are normally formed from remodeling and partial involution of these cylindrical ductal plates (1). The arrest or derangement of this process causes the persistence of excessive embryonic bile duct structures, which may be apparent microscopically or macroscopically. The diseases that develop in ductal plate malformation during different stages of remodeling—a continuum ranging from congenital hepatic fibrosis to Caroli's disease—can exist alone or in combination. These diseases are characterized by varying

degrees of persistent embryonic bile duct structures, fibrosis, and ductal dilatation (2). Congenital biliary malformations, including adult and infantile polycystic disease of the liver and kidneys, congenital hepatic fibrosis, Caroli's disease, von Meyenburg complexes, and choledochal cyst have been collectively termed ductal plate malformations or hepatobiliary fibropolycystic diseases (3-6). However, the present cases did not belong to, and were different from, the above described malformations. They were unique in imaging and histologic features. All of the biliary abnormalities in the present study resemble the human fetal ductal plate, suggesting that the present case is caused by the persistence of the fetal ductal plate. We herein report three cases with a complex mass composed of malformed ducts and biliary cysts.

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## CASE REPORTS

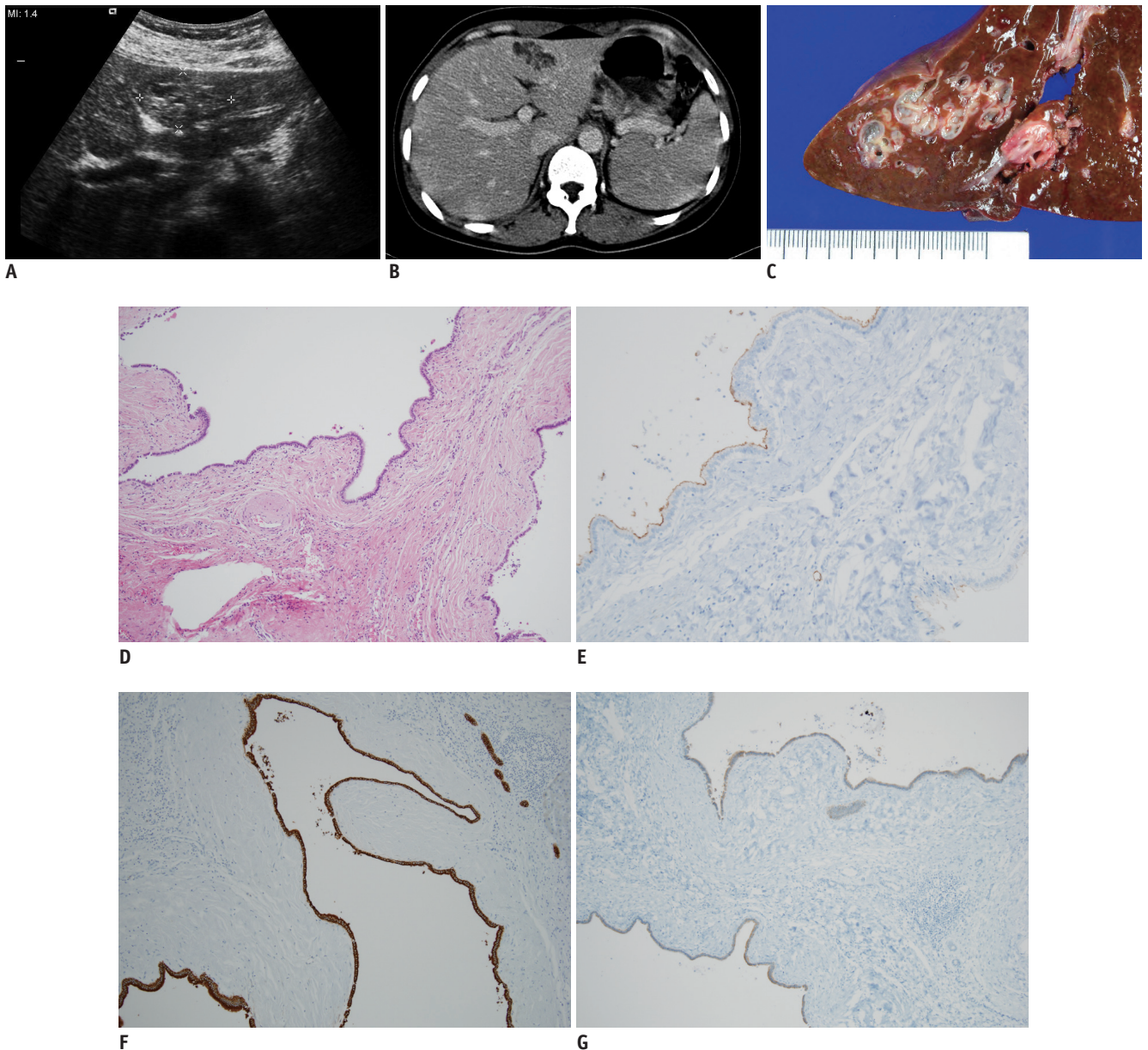
### Case 1

A 44-year-old woman presented with mild upper abdominal pain and a lobulated mass in the left lobe of the liver. This patient had undergone abdominal computed tomography (CT) scan for similar pain at a local hospital 4

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years previously, and a hepatic mass had been detected. The laboratory tests showed no abnormal findings and physical examination was normal. The evidence of polycystic kidney or liver disease was absent. There was no family history of polycystic kidney or liver disease. On the present admission, ultrasound showed an ill-defined, isoechoic mass, with internal cystic components in the lateral segment of the liver (Fig. 1A). A three-phase abdominal CT scan showed

an irregularly-marginated multicystic mass with tubular components, and with no contrast enhancement (Fig. 1B). The remaining liver was normal. This mass did not show a change in the size or contour as compared with that observed 4 years previously. Laparoscopic left lateral segmentectomy was performed. The cut surface of the hepatic left lobe showed aggregates of numerous small and large biliary cysts (Fig. 1C). Microscopically, the left



**Fig. 1. Case 1. Fibropolycystic liver disease mimicking hepatic mass in 44-year-old woman.**

**A.** Transverse abdominal ultrasound shows ill-defined isoechoic solid mass with multiple internal cystic components in lateral segment of liver. **B.** Axial contrast-enhanced abdominal CT scan shows ill-defined cystic mass with marginal tubular components. **C.** Gross findings of hepatic left lobe shows aggregates of numerous small and large biliary cysts. **D.** Microscopic findings of mass-like monosegmental fibropolycystic disease shows numerous dilated intrahepatic bile ducts, with irregular contours, infoldings, and protrusions, surrounded by thick fibrous tissue, which were features of ductal plate malformation (hematoxylin-eosin stain, x 200). **E-G.** Immunohistochemical stains for CD10 (**E**), CK7 (**F**), and MUC1 (**G**) show positive in lining (epithelial cells) of dilated cysts, as well as those of proliferative bile ductules (x 200).

hepatic mass showed numerous dilated intrahepatic bile ducts with irregular contours, infoldings, and protrusions, surrounded by thick fibrous tissue, which are known features of ductal plate malformation. There were ductal plate malformations of numerous dilated biliary cysts (Fig. 1D). Immunohistochemical stains for CD10 (Fig. 1E), CK7 (Fig. 1F), and MUC1 (Fig. 1G) showed positive reactions in the lining (epithelial cells) of dilated cysts, as well as proliferative bile ductules.

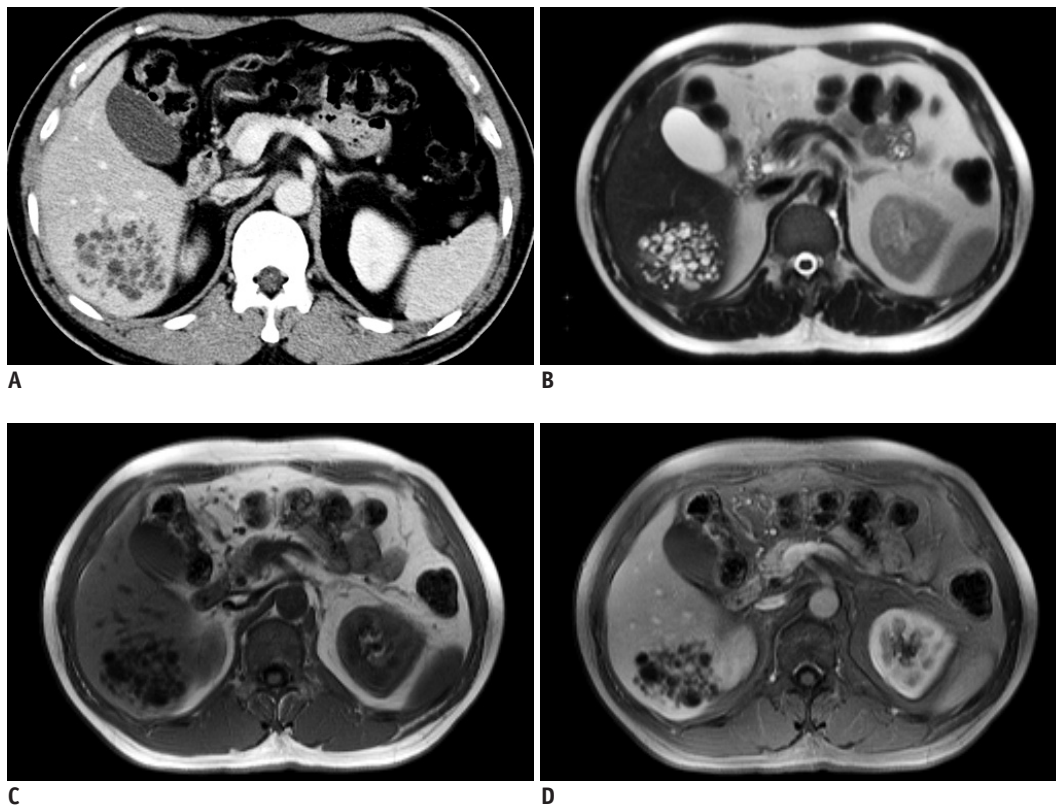
**Case 2**

A 46-year-old man presented with incidentally discovered hepatic mass in the right lobe of the liver. The laboratory tests showed no abnormal findings, and physical examination was normal. The evidence of polycystic kidney or liver disease was absent. No family history of polycystic kidney or liver disease was recognized. A three-phase abdominal CT scan showed an ill-defined mass composed of numerous cysts and tubular lesions in segment VI of the liver (Fig. 2A). Magnetic resonance (MR) images showed a cystic mass with very high T2 and low T1 signal intensities (Fig. 2B, C). Post-contrast MR images showed no contrast

enhancement of this mass (Fig. 2D). The remaining liver was normal. Percutaneous biopsy was performed. Microscopic findings of the needle biopsy specimen showed several, irregular-shaped, dilated intrahepatic bile ducts, lined by the cuboidal biliary epithelial cells (Fig. 2E). There were also ductal plate malformation and numerous inflammatory cells in a background of fibrous tissue. Immunohistochemical stains for CD10 (Fig. 2F), CK7 (Fig. 2G), and MUC1 (Fig. 2H) showed positive reactions in the lining (epithelial cells) of dilated cysts, as well as proliferative bile ductules. Follow-up imaging 18 months later showed no interval change of this cystic mass.

**Case 3**

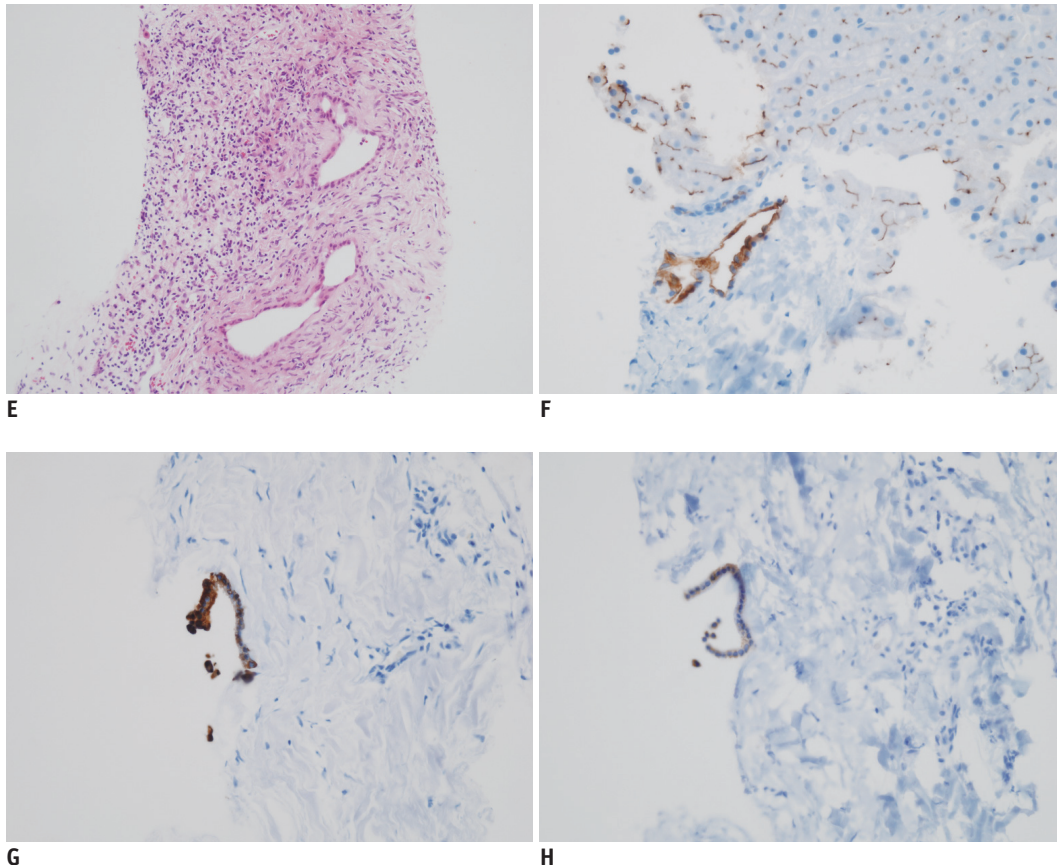
A 50-year-old man presented with incidentally discovered hepatic mass in the right lobe of the liver, during evaluation of sigmoid colon cancer. The laboratory tests showed no abnormal findings, and physical examination was normal. The evidence of polycystic kidney or liver disease was absent. No family history of polycystic kidney or liver disease was recognized. Ultrasound showed an ill-defined echogenic mass with multiple varying-sized cystic



**Fig. 2. Case 2. Fibropolycystic liver disease mimicking hepatic mass in 46-year-old man.**

**A.** Axial contrast-enhanced abdominal CT image shows ill-defined mass composed of numerous cysts and tubular lesions in segment VI of liver. **B, C.** Axial T2- and T1-weighted abdominal MR images show ill-defined mass composed of numerous cysts and tubular lesions. **D.** Axial contrast-enhanced abdominal MR image shows tubular and cystic lesions with no contrast enhancement.





**Fig. 2. Case 2. Fibropolycystic liver disease mimicking hepatic mass in 46-year-old man.**

**E.** Microscopic finding of needle biopsy specimen shows several, irregular-shaped, dilated intrahepatic bile ducts, lined by cuboidal biliary epithelial cells. There is also ductal plate malformation and numerous inflammatory cells in background of fibrous tissue (hematoxylin-eosin stain, x 200). **F-H.** Immunohistochemical stains for CD10 (**F**), CK7 (**G**), MUC1 (**H**) shows positive in lining (epithelial cells) of dilated cysts (x 200).

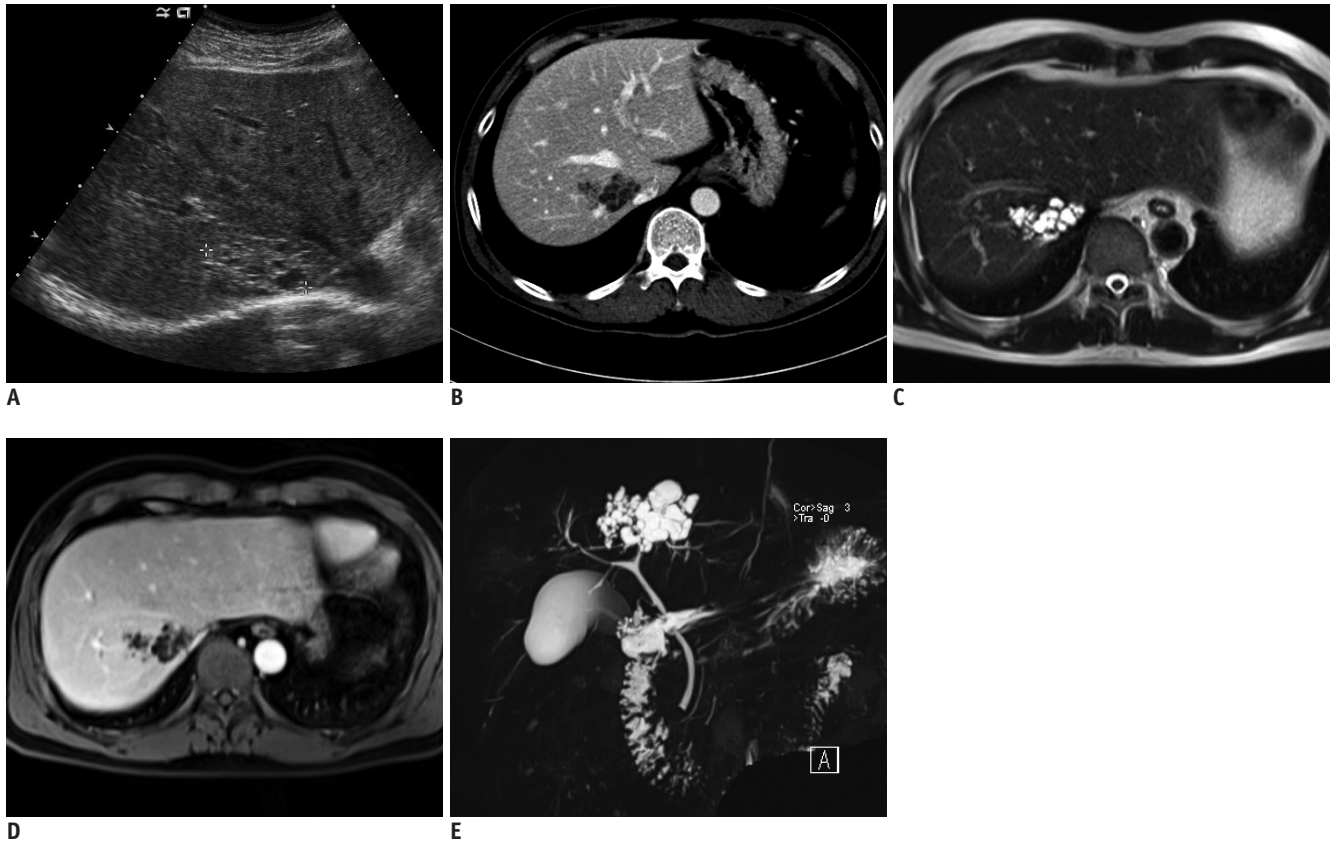
components in the right lobe of the liver (Fig. 3A). A three-phase abdominal CT scan showed an irregularly-marginated mass with tubular components but with no contrast enhancement (Fig. 3B). MR imaging showed a cystic mass with very high T2 signal intensities (Fig. 3C). Post-contrast MR image showed a cystic mass with no contrast enhancement (Fig. 3D). 3 dimensional magnetic resonance cholangiopancreatography (MRCP) showed a cystic mass with tubular and cystic components (Fig. 3E). The remaining liver was normal. Anterior resection of sigmoid colon cancer was performed, and TNM staging according to the American Joint Committee on Cancer was a stage II lesion (T2N0M0). A follow-up imaging study 18 months later showed no interval change of this cystic mass.

## DISCUSSION

Ductal plate malformation is a persistent fetal ductal plate in the postnatal livers. Desmet (5) considered that these congenital biliary diseases may be derived from the

abnormal development at different levels of the biliary tree, and named these diseases “ductal plate malformation”. Summerfield et al. (6) proposed the term “hepatobiliary fibropolycystic disease” for this congenital biliary disease group, because they show cystic lesions of the liver and bile ducts and overlap each other relatively frequently. We experienced monosegmental hepatobiliary fibropolycystic disease mimicking a hepatic mass different from previously described patterns in imaging and histologic features. To the best of our knowledge, such cases have not been reported in the literature.

Remodeling of the ductal plate results in the formation of bile ducts and ductules via epithelial proliferation and cell death, with subsequent resorption of the remaining ductal plate elements. Either a complete or partial arrest or a derangement in ductal plate remodeling is the primary abnormality in all variants of fibropolycystic liver disease, producing various degrees of duct pathology, such as dilatation and cyst formation (2, 5, 7). Remodeling of the ductal plate begins *in utero* from the hilum to the periphery



**Fig. 3. Case 3. Fibropolycystic liver disease mimicking hepatic mass in 50-year-old man.**

**A.** Transverse abdominal ultrasound shows ill-defined hyperechoic solid mass with multiple internal cystic components in segment VIII of liver. **B.** Axial contrast-enhanced abdominal CT scan shows ill-defined cystic mass with marginal tubular components. **C, D.** Axial T2- and contrast-enhanced T1-weighted abdominal MR images show ill-defined mass composed of numerous cysts and tubular lesions. **E.** Three dimensional magnetic resonance cholangiopancreatography shows aggregates of cystic and tubular lesions.

of the liver, with successive development of the large ducts first, then the segmental ducts, the interlobular ducts, and finally, the smallest bile ductules. Biliary anomalies may develop at various stages of this involution-remodeling process, and the timing or stage of maldevelopment determines the resulting clinicopathologic disorder. An understanding of abnormal embryogenesis helps to explain the characteristic imaging features of these disorders (1).

Congenital hepatic fibrosis is characterized by ductal plate malformation of small interlobular bile ducts and fibrous bands of variable thickness that link adjacent portal tracts, and is a rare autosomal recessive disorder with a variable clinical course affecting both the hepatobiliary and renal systems. Biliary hamartomas are characterized by a focal disorderly collection of bile ducts and ductules, with ductal plate malformation surrounded by abundant fibrous stroma, caused by the maldevelopment of small intralobular bile ducts. Autosomal dominant polycystic liver disease is characterized by multiple biliary cysts of variable diameter that are disseminated throughout the liver, caused

by maldevelopment of medium-sized intrahepatic ducts. Caroli's disease is characterized by a congenital dilatation of the larger intrahepatic bile ducts, which is caused by the arrested remodeling of the ductal plates of the large intrahepatic ducts: pure type characterized by only ectasia of intrahepatic bile ducts (Caroli's disease) and a combined type caused by the arrest of remodeling, both in the early period of bile duct embryogenesis, and later, during the development of the more peripheral biliary ramifications, with subsequent simultaneous maldevelopment of large-sized intrahepatic duct and small-sized interlobular bile duct (Caroli's syndrome). Choledochal cysts are characterized by ductal plate malformation of large extrahepatic ducts. Fibropolycystic liver diseases usually do not exist as single entities, but occur in conjunction with a number of other symptoms and diseases, which can be found in various combinations (1, 7, 8).

Ductal plate malformations can be involved monosegmentally or monolobarly during the process of bile duct development, as well Caroli's disease (3, 4). In the

present cases, ductal plate malformations were involved monosegmentally, and the remaining liver was normal. Therefore, we could diagnose monosegmental hepatobiliary fibropolycystic disease in all cases.

Fibropolycystic liver disease can be clinically silent or can cause numerous signs and symptoms, such as cholangitis, portal hypertension, gastrointestinal bleeding, infections, and space-occupying masses, according to each entity (1, 6). In the present cases one patient complained of mild upper abdominal pain, and the remaining two patients did not complain of pain or discomfort. At follow-up, the present cases did not change in size or shape.

The different types of fibropolycystic liver disease demonstrate characteristic findings at CT and MR imaging, according to each entity (1, 7, 9). In Caroli's disease, cystic or fusiform dilatation of the intrahepatic ducts is seen, as well as the central dot sign, which corresponds to a portal vein branch protruding into the lumen of a dilated bile duct. MRCP, percutaneous transhepatic cholangiography, and endoscopic retrograde cholangiopancreatography can show the dilated biliary trees. In biliary hamartoma, multiple small lesions (less than 15 mm) of uniform size throughout the liver are seen, which are cystic with or without peripheral rim enhancement. Congenital hepatic fibrosis frequently shows alternating hypertrophy and shrinkage of hepatic segments or lobes, periportal hepatic fibrosis, intrahepatic ductal dilatations, and renal cysts. This entity has nonspecific clinical and imaging findings, but ultrasound, CT, and MR imaging can help to evaluate this entity. Autosomal dominant polycystic disease shows multiple hepatic cysts, which are observed at the time of puberty, and increase slowly in size into adulthood (1, 7, 9, 10). Awareness of these CT and MR imaging features are essential in detecting and differentiating between various fibropolycystic liver diseases, and can assist in proper management (1).

We performed immunohistochemical analyses to characterize the biliary epithelium. All the cyst-lining epitheliums were positive for CK7, CD10, and MUC1, suggesting biliary epithelium with ductal plate malformation. MUC1 is expressed in the fetal ductal plate and fetal bile ducts, but not in postnatal biliary epithelial cells. The present findings, that MUC1 apomucin was expressed in the ductal plate malformation suggest that the ductal plate malformation in the present study was a persistent fetal ductal plate. CD10, a microvilli-associated antigen, is well-known to be expressed in hepatocytes

(canaliculi), but not in bile ducts. The reason for positive CD10 in biliary elements in the present study is unclear. Ductal transformation of hepatocytes is unlikely in the present case, but may be due to fetal biliary antigen. Examination of CD10 in the fetal ductal plate is mandatory (3, 4).

The present cases resemble monosegmental Caroli's disease, a rare disease. Caroli's disease is defined as cystic or fusiform, non-obstructive dilatation of intrahepatic bile ducts, as well as the central dot sign. Caroli's disease is characterized by retaining their communication with biliary trees (1). Monolobar or monosegmental Caroli's disease mainly affects the left hepatic lobe, and is only composed of non-obstructive segmental dilatation of the intrahepatic bile ducts (2). However, there have been no definite descriptions of the presence of proliferated intrahepatic peribiliary glands, peribiliary cysts, or ductal plate malformations in monolobar or monosegmental Caroli's disease. Caroli's syndrome is characterized by the simultaneous maldevelopment of large-sized intrahepatic ducts and small-sized interlobular bile ducts, as well as non-obstructive dilatation of the intrahepatic bile ducts, as well as other findings of ductal plate malformations. The present cases did not involve biliary trees, and showed grape-like aggregates of numerous biliary cysts, with ductal plate malformation different from the appearance of localized Caroli's disease.

In the present cases, ultrasound revealed an isoechoic or hyperechoic mass, studded with multiple varying-sized cystic components, whereas CT and MR showed mainly cystic nature, with no contrast enhancement. This discrepancy may be attributed to the echogenic solid nature resulting from interface echoes of numerous small cystic lesions at ultrasound like hepatic hemangioma. Numerous aggregated small and large biliary cysts and ductal plate malformations seemed to produce an echogenic solid nature studded with multiple larger cysts.

In summary, we report three cases of monosegmental fibropolycystic disease mimicking a mass. Awareness of this entity and image feature is of importance for differential diagnosis from incidentally discovered other hepatic masses.

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