CONGENITAL FIBROCYSTIC LIVER DISEASE.

Imaging features and differential diagnosis

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- * Ductular cells known as the ductal plate are a precursor unit for the set up of the definitive intrahepatic bile ducts.
- * Remodeling and partial involution of cylindrical ductal plates finally cause bile ducts to arise, beginning from the hilum to the periphery of the liver.

Malformations of:

- **Small-sized intra-hepatic duct:**
 - CONGENITAL HEPATIC.
 - FIBROSISBILIARY HAMARTOMAS.
- **Large and medium-sized intra-hepatic ducts:**
 - CAROLI'S DISEASE.
 - AUTOSOMAL DOMINANT POLYCYSTIC LIVER DISEASE (ADPLD).
- **Larger extra-hepatic biliary ducts:**
 - CHOLEDOCHAL CYSTS.

CONGENITAL HEPATIC FIBROSIS:

AUTOSOMAL RECESSIVE POLYCYSTIC DISEASE (ARPKD)

Some distinct CT features:

- Hypertrophic left lateral segment.
- Normal or hypertrophic left medial segment.
- Atrophic right lobe.

Varices, splenomegaly, associated ductal plate malformations, and renal abnormalities.

DIFERENTIAL DIAGNOSIS: CIRRHOSIS

Medial segment is normal in size or enlarged in congenital hepatic fibrosis, but is usually small in patients with cirrhosis due to other causes.

BILIARY HAMARTOMAS:

On ultrasound: Tiny hypoechoic or hyperechoic foci measuring less than 10 mm and distributed uniformly throughout the liver, and may demonstrate comet-tail artefacts.

On CT: low-attenuation, non-enhancing lesions.

On MRI: Lesions are hypointense on T1-weighted and hyperintense on T2-weighted images.

DIFERENTIAL DIAGNOSIS: LYMPHOMA

Multiple low-attenuation focus through the liver and spleen, and hepatosplenomegaly in a patient with lymphoma.

AUTOSOMAL DOMINANT POLYCYSTIC DISEASE:

AUTOSOMAL DOMINANT POLYCYSTIC LIVER DISEASE (ADPLD)

CT shows multiple cysts with low attenuation.

MRI: T1-weighted imaging demonstrates low signal intensity fluid-containing cysts, while T2-weighted imaging shows cysts with high signal intensity .

MRI: Contrast enhancement (multihance) delayed sequences confirms the absence of communication between the biliary tree and the cystic lesions.

AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE (ADPKD)

Both renal and kidney cystic involvement.

CAROLI DISEASE:

MR cholangiography demonstrates the communication between the cystic dilations and the biliary tree.

Contrast-enhanced CT demonstrates intra-hepatic saccular or fusiform cystic dilations which contains calculi and sludge.

CENTRAL DOT SIGN

Fibrovascular bundles with strong contrast enhancement within dilated cystic intrahepatic duct.

CHOLEDOCHAL CYSTS:

TYPE I: Cystic or fusiform dilatation of the extrahepatic common bile duct.

TYPE III: CT shows periampullary cystic dilatation of the extrahepatic common bile duct. The presence of an adjacent duodenum diverticulum is noted

TYPE IV: Combination of both intrahepatic and extrahepatic bile duct dilatation