Case	(086) Choledochal cyst with acute pancreatitis associated
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CASE PRESENTATION

A 2-year-old-girl was admitted with vomiting and a decreased level of consciousness. Past medical history includes hypertransaminasemia and cyclical vomiting. On examination, she looked healthy and had no fever. She also presented with metabolic alkalosis and a high ketone level in the blood. Treatment for vomiting and hydration was started.

Within 48 hours the patient deteriorated with fever, intense abdominal pain, and an elevation of serum lipase and amylase. An abdominopelvic ultrasound was performed which showed a 2.5 cm dilation of the extrahepatic bile duct with ascites.

After this, an abdominopelvic contrast CT revealed a fusiform dilatation of extrahepatic bile duct (3,3 x 5,5 cm) without evidence of gallstones, a proximal intrahepatic duct dilatation, a diffuse pancreatic parenchymal enlargement with multifocal parenchymal necrosis and a large volume of ascites. These findings were compatible with a choledochal cyst (type IC Todani´s classification) with acute pancreatitis associated.

DISCUSSION

- Choledochal cyst is a congenital bile duct anomaly that mainly affects the extrahepatic bile duct.

- Choledochal cysts are more common in female infants.

- Classic clinical presentation includes abdominal pain, obstructive jaundice and palpable mass in the right upper abdominal quadrant (hepatomegaly).

- Differential diagnoses includes primary sclerosing cholangitis, biliary lithiasis, cholangiocarcinoma, biliary hamartoma, biliary papillomatosis and pancreatic cystic lesions.

- The two most frequent complications of choledochal cysts are gallstones and malignancy as a cholangiocarcinoma. They also include bile peritonitis and pancreatitis.

- The diagnosis can be achieved with an ultrasound, CT or MRI, including MRCP.

- Choledochal cyst diagnosis is further supported by the presence of a direct communications between the biliary tree and cystic duct.

- The only possible treatment is surgical excision, with reconstruction.

- The Todani classification of bile duct cyst divides choledochal cysts into five groups.

- Type I: most common, characterized by dilatation of extrahepatic bile duct only.

- IA: Cystic dilatation

- IB: Distal choledocho dilatation

- IC: Fusiform dilation 2. Type II: Bile duct diverticulum 3. Type III: Choledochocele involving intraduodenal portion of the distal common bile duct 4. Type IV: Multiple communicating intra and extrahepatic duct cysts:

- IVA: Intra and extrahepatic cysts

- IVB: Multiple extrahepatic cysts 5. Type V: Multiple dilatations of intrahepatic ducts only (Caroli disease).

CONCLUSION

Choledochal cysts are a rare disease entity. It´s important to know the specific findings in the imaging tests and correlate it with the clinic in order to make a good differential diagnosis and avoid secondary complications (like malignancy, pancreatitis, biliary lithiasis, etc.).

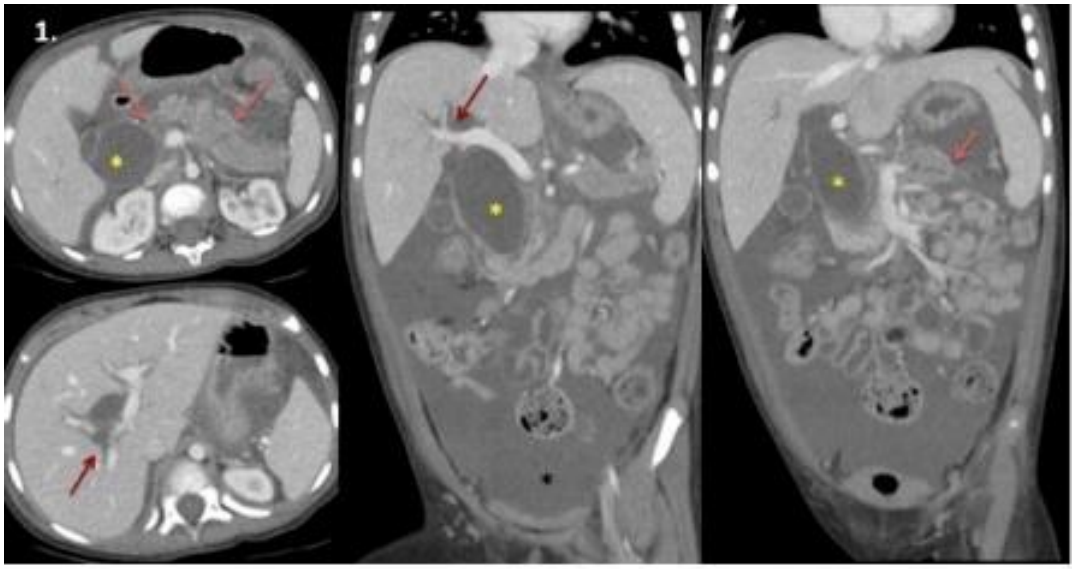

Figure 1: An abdominpelvic contrast CT revealed a fusiform dilation of extrahepatic bile duct, a proximal intrahepatic duct dilation, a diffuse pancreatic parenchymal enlargement with multifocal parenchymal necrosis and a large volume of ascites.

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