Management of pancreatic lithiasis in a 5-year-old girl

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Pancreatic lithiasis causing chronic pancreatitis is a very rare entity in childhood. A five-year-old girl presenting with pancreatic lithiasis was treated successfully with a well-organized diagnostic and therapeutic algorithm. The authors emphasize that early diagnosis followed by appropriate therapeutic pancreatographic procedures instead of primary invasive surgical approach can certainly ameliorate progressive and irreversible pancreatic damage. The clinicopathologic features of this rare entity are discussed, with emphasis on diagnosis and treatment.

Key words: child, pancreatic lithiasis, cholangiopancreatography.

Pancreatic lithiasis (PL) is a very rare entity in childhood and most of the PL series in the English-language literature contain many segmented or partial descriptions that are sometimes difficult to correlate. Herein, a five-year-old child with PL is presented with a brief literature review focused on clinicopathologic features, diagnosis and management scheme.

Case Report

A five-year-old girl was referred to our department with the complaint of recurrent abdominal pain lasting for two months. No abnormality was detected in physical examination or on plain abdominal X-rays. Laboratory research for the etiology of chronic abdominal pain including tests for fibrinogen, salmonella and brucella agglutination, and parasitosis were all normal, as were the values of pancreatic amylase (63 U/L) (normal range: 17-115 U/L), amylase-creatinine clearance (4%) (normal range: ≤5%) and steatocrit (negative) (normal value).

Abdominal ultrasonography (US) revealed dilatation of Wirsung canal up to 4-5 millimeters (mm), which was also confirmed by magnetic resonance cholangiopancreatography (MRCP). Multiple millimetric stones were lodged in the pancreatic head and the pancreatic canal was attenuated in this region (Figs. 1a and 1b).

Endoscopic retrograde cholangiopancreatography (ERCP) was performed under general anesthesia. Papilla of Vater was cannulated and the choledochus and intrahepatic bile ducts were visualized as normal after flush of contrast medium even though the pancreatic canal seemed to be dilated with the appearance of filling defects suggestive of multiple millimetric stones. Papillotomy was performed and stones in the form of sludge and debris were extracted. The patient had an uneventful postoperative course and was discharged on the third day of intervention.

The follow-up abdominal ultrasonography performed three months later revealed dilatation of Wirsung canal up to 4 mm with multiple stones 5 mm in size at the pancreatic head, and the parenchyma showed signs of chronic pancreatitis with multiple millimetric calcifications. Similar findings were also confirmed by MRCP together with the additional sign of pseudocyst of 2 cm located in the uncinate process.

Endoscopic retrograde cholangiopancreatography was performed for therapeutic purpose, and the minor duodenal papilla was cannulated, revealing stenosis 2 cm in length and dilatation proximally. After sphincterotomy, pancreatic plastic stent (5 F) 5 cm in length was inserted into the minor canal (Fig. 2). After an uneventful
postoperative course, the patient was discharged on the sixth day of intervention after performance of follow-up US demonstrating disappearance of pancreatic stones with multiple calcifications in the pancreatic head region (Fig. 3).

Discussion

Although pancreatitis is uncommon during childhood, it must be considered in every child with unexplained acute or recurrent abdominal pain. The etiology of acute pancreatitis in children is mostly idiopathic; while chronic, relapsing pancreatitis is frequently hereditary or due to congenital anomalies of the pancreatic or biliary ductal system. The hereditary form with autosomal dominant transmission has an onset with mild symptoms beginning in the first decade. It is diagnosed with the presence of the disease in successive generations of a family. Although spontaneous recovery from each attack occurs in a few days, episodes may become progressively more severe and evaluation during symptom-free intervals may be unrewarding until calcifications, pseudocysts or pancreatic insufficiency develops.

Pancreatic lithiasis, one of the most frequent causes of chronic pancreatitis, has two forms as calcic lithiasis, generally due to nutritional disorders, and proteic lithiasis, the hereditary cases of which show autosomal dominant transmission. Radiolucent PL, distinct from calcified PL, is a heterogeneous pancreatic disease with juvenile and senile presentation, representing 15% of PL, and evolution towards calcific stages could be related to a genetic factor or increased alcohol consumption.

In our case, research for the etiology of unexplained recurrent abdominal pain consisting of clinical, laboratory and radiologic evaluation was performed in a stepwise manner. After the demonstration of pancreatic duct dilatation
on US and PL additionally on MRCP, the etiology of recurrent abdominal pain was clarified. During the hospitalization period, the patient had no symptoms of fever, abdominal pain or vomiting. The laboratory findings were also not in accordance with an attack of pancreatitis. Hence, instead of a management with nasogastric suction and parenteral nutrition, as performed in acute pancreatitis, the patient was fed orally with close follow-up of clinical and laboratory findings.

The presenting findings of our case on the first admission aside from a pancreatitis attack are mostly in accordance with a symptom-free interval and probably suggest a hereditary form of PL, in which no progressive or irreversible pancreatic pathology has yet been clarified.

Endoscopic retrograde cholangiopancreatography is being used with increasing frequency in the pediatric population for both diagnostic and therapeutic purposes. With the introduction of pediatric duodenoscopes and expansion of indications in children, operative endoscopic procedures including endoscopic sphincterotomy, biliary drainage, common bile duct and pancreatic duct stone extraction, implantation of endoprosthesis, and pancreatic pseudocyst drainage are also being applied in childhood at an increased frequency and with an overall success rate similar to that seen in adult patients. Since the complications of ERCP, the most frequent being pancreatitis, cholangitis and hemorrhage, are seen at a higher rate in childhood than in adults (33.3%) even in experienced hands, MRCP should have priority over ERCP as a diagnostic modality, especially for cases in which the need for therapeutic intervention is uncertain, and strict selection criteria must be met before subjecting a pediatric patient to ERCP. During decision making for the management of our case, there was conflicted opinion as to whether or not to perform ERCP, since definitive diagnosis of PL has already been obtained by MRCP and there was no finding of acute pancreatitis necessitating urgent intervention. On the other hand, therapeutic intervention would be more logical before the onset of pancreatic parenchymal changes observed in chronic pancreatitis to achieve the most marked clinical improvement, as mentioned in a previous study, so ERCP with papillotomy was performed. On the second admission, the diagnostic studies revealed signs of emerging chronic pancreatitis, which forced us to perform urgent intervention with insertion of pancreatic stent.

A review of the literature reveals that invasive surgical approach for PL and chronic pancreatitis, such as longitudinal pancreaticojunostomy and pancreatic resection, has generally been replaced by endoscopic procedures supported by early diagnosis and early intervention. Some recently mentioned options for management are extracorporeal shock-wave lithotripsy combined with endoscopic pancreatic sphincterotomy for the management of giant stone in Wirsung canal in the case of severe chronic pancreatitis, intraoperative electrohydraulic lithotripsy of pancreatic duct stones used in combination with lateral pancreatojunostomy to avoid pancreatic resection in the case of chronic fibrocalcific pancreatitis, pulsed dye laser for pancreatic stones in the smaller pancreatic duct applied during ERCP, direct examination of pancreatic duct system with intraoperative pancreatoscopy using a pediatric cystoscope, and removal of pancreatic duct stone debris together with cyst excision in the case of choledochal cyst associated with PL, to prevent relapsing pancreatitis. In contrast, some other reports suggest only choledochal cyst excision without any intervention for pancreatic calculi, and mention that no evidence of pancreatic insufficiency or recurrence of preoperative symptoms occurred during the follow-up of the cases.

REFERENCES


