Case Report

A case of pancreas divisum in childhood diagnosed and treated by endoscopic retrograde cholangiopancreatography

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ABSTRACT

Pancreas divisum (PD) is the most common congenital anomaly of the pancreas. Its diagnosis is currently based on the magnetic resonance imaging (MRI). However, endoscopic retrograde cholangiopancreatography (ERCP) remains the reference examination, especially in difficult cases; it has a diagnostic and therapeutic role. The treatment of PD is recommended only for symptomatic patients; it is endoscopic and/or surgical. The response to treatment depends on the clinical present. We report a new case of PD in 13 years old child revealed by recurrent acute pancreatitis diagnosed by ERCP: the pancreatography via minor papilla revealed a very dilated duct of Santorini which is following main pancreatic duct. Sphincterotomy of the minor papilla was performed with good clinical and biological response. Through this case of PD and review of the literature, the authors insist on diagnostic challenges and the role of ERCP in the diagnosis and treatment of PD.

Key words: Pancreas divisum, endoscopic retrograde cholangiopancreatography, magnetic resonance imaging

INTRODUCTION

Pancreas divisum is the most common congenital anomaly of the pancreas resulting from the failure of fusion of the dorsal and ventral pancreatic ducts. The dorsal pancreatic duct or duct of Santorini is dominant and drains most of the pancreatic secretions via a non-adapted minor papilla. The high prevalence of PD in patients presenting recurrent acute pancreatitis, chronic obstructive pancreatitis and the results of targeted treatments on the minor papilla are the arguments for the pathogenicity of PD. The diagnosis of PD is currently based on MRI. Treatment is for symptomatic patients only; it is endoscopic and/or surgical. We report a new case of PD in childhood that illustrated the difficulties of diagnosis and the role of ERCP in a PD diagnosis and treatment.

CASE REPORT

A 13 years-old girl was admitted to the children’s hospital with an epigastric pain radiating into her back, accompanied by intermittent nausea and vomiting. His history was marked by the recurrent acute pancreatitis since the age of 12 years. Physical examination at admission has noted a moderate epigastric tenderness without significant abdominal distention. Vital signs were within normal limits. Serum amylase and lipase levels were markedly increased, while other serum biochemical levels were within normal ranges. Abdominal computed tomography (CT) scan showed Pancreatitis score C of Balthazar. MRI of the pancreas and magnetic resonance cholangiopancreatography showed that the common bile duct and the dorsal pancreatic duct had different duodenal openings and that the pancreatic duct was dilated (Fig. 1). The patient’s abdominal pain was improved by intravenous fluid hydration and restriction of food.

An ERCP was performed about one month later in our institution, the opacification via the major papilla showed a normal caliber cephalic duct of wirsung, no opacification of the dorsal duct and remainder of the principal pancreatic duct. The primary bile duct was no-dilated. The opacification via the minor papilla founded a very dilated duct of Santorini following the principal pancreatic duct, which is also very dilated (Fig. 2). The diagnosis of acute pancreatitis secondary to PD was retained on ERCP after having eliminated the usual causes of pancreatitis (biliary stones, drug-induced pancreatitis, abdominal trauma, disorders metabolic particularly those related to hyperlipidemia and hypercalcemia, tumors). An endoscopic
minor papilla sphincterotomy was performed with a good clinical and biological evolution.

**DISCUSSION**

Pancreas divisum is a common embryologic anomaly of the pancreatic ductal system.

There is a failure of fusion of the two pancreatic ducts. The dorsal duct or duct of Santorini provides drainage for the secretion of the majority of the gland. The smaller ventral pancreas is drained by the foreshortened duct of Wirsung. Only recently has an association between pancreas divisum and pancreatitis been postulated.

Opie first described this anatomical variant in 1903 and later reported a 10% frequency in postmortem examinations. The significance of this anomaly in pancreatic disease remained obscure until the introduction of ERCP in the 1970s as and the incidence was estimated at 7.5%. The role of PD in acute and chronic recurring abdominal pain is controversial. The disproportion between the pancreatic flow drained by the dorsal pancreatic duct and the small size of the minor papilla has long been incriminated in genesis of acute pancreatitis. However, the occurrence of these clinical manifestations at an adult age suggests the association with an acquired organic stenosis of the minor papilla probably related to the aging.

Only 5% of patients with PD abnormalities are symptomatic. The PD may be responsible for recurrent acute "idiopathic" pancreatitis (PAIR), whose exhaustive etiological investigation reveals no other abnormality associated with the ductal abnormality of the PD. Cotton reported a series of 37 cases of recurrent idiopathic acute pancreatitis (PAIR), of which 25% had PD. Chronic abdominal pain and chronic obstructive pancreatitis are also a form of PD disclosure.

Traditionally, ERCP has been the procedure of choice for diagnosis and it is commonly used as the conventional option for diagnosing pancreas divisum. However, ERCP is an invasive diagnostic method which is associated with possible serious consequences. Magnetic resonance cholangiopancreatography (MRCP) is non-invasive diagnostic technique that evaluates the pancreaticobiliary ductal system and has been shown to have a good sensitivity and specificity for the diagnosis of pancreas divisum. It makes the positive diagnosis of PD with a high interobserver concordance by finding "Cross duct sign": the dorsal pancreatic duct drained in the minor papilla, crossing the common bile duct that open in in the ventral pancreatic duct in the duodenum at the major papilla. Secretin could increase the volume of ductal fluid and the secretions by the exocrine pancreas, so MRCP after secretin stimulation (S-MRCP) could improve the visualization of pancreatic ducts and enable the assessment of exocrine function of pancreas. Its results are comparable to ERCP, which is the gold standard in PD. The pancreatogram via major papilla revealing in the case of complete PD, a short and fine ventral segment duct (2 to 3 cm) without opacification of the corporeocaudal portion. In one third of PD cases, the ventral duct may be absent and the pancreatogram obtained by canulation of the minor papilla finds the dorsal duct running across the length of pancreas, crossing the bile duct and ending independently in the duodenum.

In our observation, the opacification via the minor papilla showed a dilated dorsal duct opening independently in the duodenum at MRI which not founded 'Crossing sign' because the sinuous aspect of the terminal part duct of the dorsal. ERCP is the reference examination to diagnose difficult cases of PD, it showed the dominance of the dorsal duct I to drain the major part of the pancreatic secretions through an accessory papilla.

PD treatment is only for symptomatic patients. It removes the obstacle in the minor papilla and facilitates the drainage of the pancreatic secretion along the dorsal pancreatic duct. It is endoscopic and/or surgical. Endoscopic sphincterotomy, reported for the first time by Cotton in 1978, is the endoscopic treatment of choice. It consists at a catheterization of the minor papilla and implantation the pancreatic prosthesis in the dorsal duct. The gesture ends with an incision of the papilla with the bistoury, prosthesis in place. Papillary dilatation by balloon was abandoned due to a high rate of pancreatic trauma. The second alternative endoscopic method is the injection of botulinum toxin in the minor papilla used by Wehrmann et al in 5 patients with PD with PAIR in order to identify patient’s candidates for sphincterotomy. The disappearance of symptoms was noted in 3 patients with subsequent endoscopic sphincterotomy.

In our patient, the sinuous and hooked aspect of the terminal part of the dorsal duct made it difficult to put in place a pancreatic prosthesis, a simple sphincterotomy of the accessory papilla was performed with a good clinical evolution.

The results of endoscopic sphincterotomy depend on the clinical presentation. They are favorable in 75% of patients with recurrent acute pancreatitis compared to less than half of patients with chronic pancreatitis or chronic pancreatic pain.

The most commonly used surgical treatment is the transduodenal sphincteroplasty of the minor papilla. This technique must be associated with cholecystectomy in order to find a microlithiasis. The pancreatico-jejunal derivation is possible in the case of associated canal obstruction. Finally, left pancreatectomy and cephalic duodenopancreatectomy are performed after failure of previous surgical techniques.

Endoscopic treatment is generally recommended as a first-line treatment for a PD responsible for PAIR. Surgery is indicated in case of failure or complications after endoscopic treatment. In case of pain or chronic pancreatitis, surgical sphincterotomy gives better results (40 to 60% good results) than endoscopic sphincterotomy (25% good results).
CONCLUSION
Pancreas divisum is the commonest congenital anomaly of the pancreas that can be responsible for recurrent acute pancreatitis. Abdominal MRI is noninvasive examination to confirm the diagnosis of PD. Nevertheless, ERCP remains the reference examination especially in difficult cases, it has a diagnostic and therapeutic role. Treatment of pathological PD is endoscopic or surgical. The response to treatment depends on the clinical presentation.

REFERENCES