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Pancreatitis and Portal Vein Thrombosis in Children: The Chicken or the Egg Causality Dilemma

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ABSTRACT

Background: The association of pancreatitis and portal vein thrombosis (PVT) is extremely rare in children. We report on 3 cases which suggest that there may be a causal relation between the two.
Methods: Clinical characteristics and evolution of 3 children with this particular condition were analyzed retrospectively. In this group of patients, the strategy consisted in opting for early surgical decompression of the portal hypertension, which was followed by a favorable outcome, not only in terms of complications related to the portal hypertension but also of a contemporaneous spontaneous regression of the concurrent pancreatic disease, in absence of any other specific management of the latter problem.
Results and conclusions: Combined PVT and pancreatitis is exceptional in children. Although this series is small, it provides insight and some evidence that the pancreatic disease might be secondary to the cavernomatous transformation of the regional venous system. More interestingly, it suggests that the appropriate management strategy should be to rapidly relieve portal hypertension after resolution of the acute phase of pancreatitis.

In recent years, an increasing number of reports have addressed the issue of acute and chronic pancreatitis in the pediatric population [1,2]. This has highlighted the need for a proper diagnostic and therapeutic algorithm for children, since most authors agree that the typical adult pattern and strategy is not applicable in the pediatric field.

In children, pancreatitis may result from congenital anatomic alterations such as pancreaticobiliary anomalies (pancreas divisum, annular pancreas, pancreaticobiliary maljunction, choledochal cysts), medication, or trauma [1]. Thrombosis of the portal vein trunk (PV), of the splenic vein (SV) or even of the superior mesenteric vein (SMV) [3] is not described in the literature as a cause of pancreatitis in children, and there is no previous description of pediatric cases with combined pancreatitis and thrombosis of one or more regional veins.

Because the association of PV thrombosis and pancreatitis is an exceptional condition in children, we present a short series of 3 cases managed according to an original strategy that was associated with excellent outcome. The objective of this study was thus to analyze the results of this treatment strategy in 3 children presenting with both thrombosis of PV ± SV and acute/chronic pancreatitis [2]. Particular attention was paid to the clinical evolution of this particular condition, and to determining whether or not the treatment of portal hypertension [4,5] could have an impact on the concurrent pancreatic disease.

1. Patients and results

1.1. Case 1

A 4-year-old female presented with a prolonged history of recurrent pleural effusions. She was a heterozygous carrier of a mutation of the CFTR ΔF508 gene (mutation 7T/9T on IVS8) and had repeatedly normal results for fecal elastase and sweat tests. Chemical analysis of the pleural liquid confirmed a high amylase and lipase content and abdominal imaging was performed; surprisingly, ultrasonography not only confirmed chronic pancreatitis with pancreatic pseudocyst but also showed signs of portal hypertension (splenomegaly, retroperitoneal venous collaterals), and a cavernomatous transformation of the portal vein (Fig. 1). On CT (computerized tomography) angioscan, the splenomesenteric confluence and splenic vein were replaced by a network of varices and venous collaterals running around and through the head and body of the pancreas (splenic cavernoma). The pancreas looked congestive and edematous, with numerous pseudocysts, and a retro-pancreatic fluid collection extending upward along the paravertebral space was observed, communicating with the pleura and causing the pleural effusion (Fig. 1). On CT angioscan the upper portion and bifurcation of the portal vein as well as the Rex recessus were patent. Because the patient had already been managed conservatively for 14 months with no resolution of the pancreatitis and only a

Abbreviations: PV, Portal vein; SV, Splenic vein; SMV, Superior mesenteric vein; INSPPIRE, International study group of pediatric pancreatitis: in search for a cure; CFTR, Cystic fibrosis transmembrane conductance regulator; CT, Computerized tomography; USD, Doppler ultrasonography; MRCP, Magnetic resonance cholangiopancreatography.

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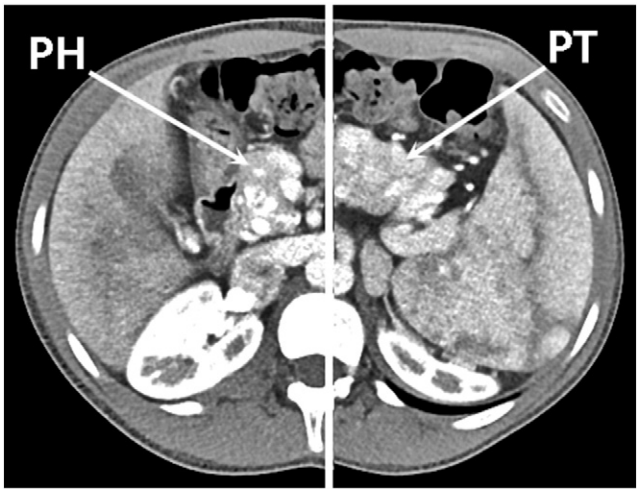

Fig. 1. CT imaging of a cavernoma of the splenic vein and subsequent transformation of the pancreas (PH, pancreas head; PT, pancreas tail).

moderate effect on the pleural effusion, it was proposed that the portal hypertension be treated before proceeding to endoscopic or surgical pancreatic interventions. One month later, a meso-Rex bypass was performed [4–6]. Although the pseudocysts were still present, the procedure was performed in a standard fashion without any specific access problems, and a bypass was interposed between the Rex recessus and the superior mesenteric vein immediately below the pancreas neck [6].

Postoperative course was uneventful, and she was discharged on day 6 after surgery. As per our protocol for bypass surgery, anti-aggregant (dipyridamole 7 mg/kg/day and acetylsalicylic acid 3 mg/kg/day) and anti-acid therapy were administered for 3 months.

At last clinical visit (14 months after surgery), Doppler ultrasonography (USD) and CT scan have demonstrated the patency of the shunt and a regression of the pancreatic disease (Fig. 1), with spontaneous resolution of the pancreaticopleural fistula and pleural effusion and of the pancreatic pseudocysts. Also, the spleen span (USD) is back to normal, with no hypersplenism (platelet count, 235.000/μL; white cell count, 5460/μL); blood amylase and lipase levels, fecal elastase and repeated sweat test results are all normal.

1.2. Case 2

This male patient first consulted at 6 years of age for upper gastrointestinal bleeding. Esophagogastroduodenoscopy showed F2–F3 esophageal varices (according to the classification of the *Japanese Research Society for Portal Hypertension* [7]. USD and CT scan showed a cavernoma of the PV, with the thrombosis slightly extending into the cephalic portion of the superior mesenteric vein (patent SV), and splenomegaly. On transjugular retrograde portography [8], the Rex recessus and the intrahepatic portal venous system were thrombosed. The patient was enrolled in an endoscopic variceal band ligation program and oral beta-blocker treatment was started [9,10].

About 3 years after the first assessment, the patient was hospitalized for a sudden episode of acute pancreatitis with increased pancreatic enzymes (twice the upper normal laboratory values). USD, CT scan and MRCP (magnetic resonance cholangiopancreatography) imaging confirmed an acute edematous pancreatitis with a pseudocystic lesion of the pancreas head (Fig. 2). Esophagogastroduodenoscopy showed no recurrence of esophageal varices, but a severe hypertensive gastropathy was present.

Two months after this event, USD showed that the pancreatic pseudocyst had reduced in size by half; the patient was clinically well but pancreatic edema was unchanged. Because a meso-Rex bypass was not feasible, a distal splenorenal shunt (modified “Warren” type; no division of gastric veins) was performed [9–11]; at operation, no specific difficulties were encountered when approaching the splenic and renal veins; only a mild edema of the root of the mesentery was observed. Postoperative course was uneventful, and the patient was discharged on day 7 after surgery. As per our protocol for bypass surgery, anti-aggregant therapy (dipyridamole 7 mg/kg/day and acetylsalicylic acid 3 mg/kg/day) was administered for 3 months.

Six months after surgery, USD showed complete resolution of the pseudocyst and of signs of pancreatitis, and a well-functioning shunt (Fig. 2); esophagogastroduodenoscopy confirmed absence of esophageal varices and only a slight residual gastropathy.

1.3. Case 3

A 16-year-old female presented to our hospital for recurrent abdominal pain; she had a prolonged history of chronic pancreatitis and multiple pancreatic pseudocysts. The latter diagnosis had been made 5 years previously in another hospital where she had undergone surgical exploration for the pseudocysts (simple evacuation). USD and CT angioscan confirmed the chronic pancreatitis, with a slightly atrophic pancreas and multiple pseudocysts containing heterogeneous fluid (Fig. 3). Absence of factors favoring pancreatitis, but presence of a related moderate exocrine pancreatic insufficiency, was also confirmed; clinical symptoms improved after oral enzyme supplementation was started. Interestingly, USD and CT angioscan also showed a cavernomatous transformation of PV (SV and SMV and their confluence were patent). Although the patient presented with huge splenomegaly and an extensive network of venous collaterals around the pancreas and within the porta hepatis (Fig. 4), her platelet count was at the lower end of the normal range (169.000/μL) (a higher value than expected, possibly because of an inflammatory condition), and endoscopy of the upper gastrointestinal tract showed no esophageal varices. Transjugular retrograde portography [8] confirmed the thrombosis of the extrahepatic portion of the portal vein with a patent intrahepatic portal system and Rex recessus. A conservative initial treatment strategy was used to try to reduce the pancreatic inflammation and pseudocysts before portal decompressive surgery, but without success. Ten months after the initial admission, a meso-Rex bypass was performed. At that time, the pseudocystic lesions of the pancreas had not changed, but there were no intraoperative problems in creating or positioning the bypass as it followed a straight and easy route between the pseudocysts of the pancreas head and those of the pancreas body. Only minor adhesions were found between the stomach and the pancreas, and the meso-Rex bypass was performed in a standard manner [6].

Postoperative course was uneventful, and she was discharged on day 6 after surgery. As per our protocol for bypass surgery, anti-aggregant (dipyridamole 7 mg/kg/day and acetylsalicylic acid 3 mg/kg/day) and anti-acid therapy were administered for 3 months. Pancreatic enzyme replacement therapy was continued as used preoperatively.

During the first 6 months after surgery, she presented with 3 episodes of abdominal pain and vomiting that required short observation: at that time, pancreatic enzyme levels were normal and there were no signs of pancreatitis on imaging. Subsequently, she improved progressively; pancreatic enzyme substitution was reduced (fecal elastase results were normal) and then stopped during the second year after surgery. At 22 months after surgery, she was asymptomatic with USD imaging confirming the patency of the meso-Rex bypass, a complete disappearance of the pancreatic pseudocysts (Fig. 3), and a normal spleen span (measured at 10 cm on USD).

2. Discussion

In children, extrahepatic portal vein thrombosis is a relatively frequent cause of portal hypertension, while pancreatitis associated with thrombosis of the portal vein is exceptional. Because the Bambino