Successful Pancreatic Duct Stent Placement for Recurrent Pancreatitis in a Patient with Polysplenia with Agenesis of the Dorsal Pancreas and Peutz-Jeghers Syndrome

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Abstract

A 52-year-old woman presented with relapsing acute pancreatitis. A contrast CT scan revealed polysplenia, agenesis of the dorsal pancreas, perduodenal portal vein, inferior vena cava with persistent continuity of the azygos vein, abnormal lung lobation with bilateral left bronchial morphology, and intestinal malrotation (non-rotation type). To the best of our knowledge, this is the first report in which successful pancreatic duct stent placement for the treatment of recurrent pancreatitis was performed in a polysplenia patient with agenesis of the dorsal pancreas, separate bile and pancreatic ducts and Peutz-Jeghers syndrome.

Key words: Peutz-Jeghers syndrome, polysplenia, preduodenal portal vein, recurrent pancreatitis, short pancreas


Introduction

Polysplenia syndrome is often accompanied by a variable spectrum of visceral and vascular developmental anomalies. It is a rare disorder often involving multiple cardiac and gastrointestinal structural abnormalities, with potential effects on normal physiology (1). On the other hand, Peutz-Jeghers syndrome (PJS) is an inherited polyposis syndrome in which multiple characteristic polyps occur in the gastrointestinal tract with an increased risk of gastrointestinal and extraintestinal carcinoma associated with mucocutaneous pigmentation, particularly of the vermillion border of the lips. PJS is inherited in an autosomal dominant manner and caused by a germline mutation in the STK11 (LKB1) gene (2). We herein report a case of polysplenia complicated with agenesis of the dorsal pancreas, preduodenal portal vein, inferior vena cava with persistent continuity of the azygos vein, intestinal malrotation (non-rotation type). Both right and left bronchi demonstrated bilobed, left-sided conformation with the absence of an accessory fissure on the right lung (Fig. 1).

We made a definitive diagnosis of PJS with multiple polyposis according to the esophagogastroduodenoscopy and colonoscopy findings, the pigmentation of her side foot, and the family history of PJS of her younger sister, mother, and

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Received for publication July 7, 2015; Accepted for publication September 27, 2015
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Figure 1. Contrast CT from the chest to the abdomen. a: The portal vein (PV) was located in front of the duodenum. The pancreatic tail showed agenesis. GB: Gall Bladder, PV: Portal Vein, DU: Duodenum, Pa: Pancreas. b: Polysplenia was evident although she was not situs inversus. There is also a 35 mm hemangioma in S7. c: The intrahepatic inferior vena cava (IVC) was absent and the IVC was connected to the azygos vein above the heart. d: The right lung lacked a minor fissure and was bilobed.

Figure 2. We made a definitive diagnosis of Peutz-Jeghers syndrome on the basis of the esophagogastrroduodenoscopy and colonoscopy findings, and skin pigmentation. a: Esophagogastrroduodenoscopy revealed multiple 1-3 mm polyps around the gastric mucosa. b: It was difficult to detect the orifices of the bile and pancreatic ducts even by duodenoscopy. c: A polyp of about 3 cm in size in the duodenum was resected by endoscopic mucosal resection (EMR). d: Multiple polyps were also detected in the colon. e: There was a black pigmentation in her side foot.
endoscopic mucosal resection (EMR). They demonstrated a papilla due to the PJS polyps and intestinal malrotation.

The large polyps in the duodenum were initially removed by endoscopic mucosal resection (EMR). They demonstrated PJS pathological characteristics showing a frond-like elongated epithelial component and cystic gland dilatation extending into the submucosa or muscularis propria and arborizing the smooth muscle extending into the polyp fronds.

The removal of the large polyps made it possible to identify the papilla and pancreatic duct (Fig. 3a). Successful pancreatic duct cannulation was achieved. A pancreatogram showed a dilated pancreatic duct in the short pancreas, which branched off the patient’s left side and upward (Fig. 3b). As there was no obvious stricture in the main pancreatic duct orifice, endoscopic sphincterotomy to the pancreatic duct was performed to improve the flow of pancreatic juice. It was also difficult to identify the common bile duct orifice, which should be above the pancreatic duct, according to magnetic resonance cholangiopancreatography (MRCP) (Fig. 3c). Finally, a 5-Fr diameter and 10 cm long plastic stent was indwelled in the pancreatic duct which allowed the smooth flow of pancreatic juice for the first time (Fig. 3d). There were no procedure-related complications as regular stent exchange using a 10-Fr 10 cm plastic stent was performed every 3-6 months to prevent the recurrence of pancreatitis. As it may be difficult to find the pancreatic duct orifice after stent removal, we tattooed the duodenum near the pancreatic duct orifice for easier identification and planning on how to extract the pancreatic tube stent (Fig. 3e).

**Discussion**

The precise etiology of polysplenia remains unknown. Embryonic, genetic, and teratogenic components have all been implicated as causative factors. Polysplenia is associated with multiple congenital malformations that may involve solid organs and the digestive tube of the abdominal cavity, the heart, or the great vessels (3). A previous study has suggested that up to 2.2% of patients with congenital heart defects may have an underlying splenic abnormality (4). Although it is estimated that 2.5/100,000 infants are born with this anomaly, fewer than 5% are still alive at 5
years of age due to the associated severe cardiac anomalies (5). The most common vascular anomalies are agenesis of the suprarenal inferior vena cava with persistent continuity of the azygos vein and the preduodenal position of the portal vein. Biliary atresia is found in nearly 50% of cases, common mesentery in more than 75% of cases, and an abbreviated or annular pancreas in 85-90% of cases (6).

PJS is an uncommon disorder characterized by mucocutaneous pigmentation and gastrointestinal hamartomatous polyps. The incidence of PJS is estimated to be between 1 in 50,000 to 1 in 200,000 live births (7). Polyp-related symptoms typically arise in childhood and are observed by 10 years of age in 33% and by 20 years of age in 50% of all cases. Acute pancreatitis can be triggered by the temporary or permanent blockage of the pancreatic duct. Acute pancreatitis related to gastrointestinal polyposis syndrome has been reported in a few cases (8-10).

To the best our knowledge, there is no similar report on successful therapeutic ERCP in such patients. In terms of endoscopic retrograde cholangiopancreatography (ERCP), there are two interesting points in our report. as follows: 1) many polyps hindered the detection of the pancreatic duct orifice, and polypectomy was more effective in locating the orifice, thereby enabling pancreatic duct stent placement; 2) to readily detect the papilla during stent exchange, we tattooed the duodenum near the pancreatic duct orifice. In the present case, there was no clear answer regarding the timing of stent removal. As of this writing, we plan to schedule the complete removal of the stent 1 year after the initial stent placement.

In conclusion, we herein described successful pancreatic intervention in a patient who simultaneously presented with PJS and polysplenia with agenesis of the dorsal pancreas, preduodenal portal vein, inferior vena cava with persistent continuity of the azygos vein, intestinal malrotation, and separate orifices of the bile and pancreatic ducts.

All procedures were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5).

Informed consent was obtained from the patient to be included in the study.

The authors state that they have no Conflict of Interest (COI).

Acknowledgement

The authors thank the medical editors of the Department of International Medical Communications of Tokyo Medical University for editing the manuscript.

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