Hereditary Pancreatitis Showing Numerous Cysts with Pancreatic Cancer

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A 49-year-old man presented with multiple pancreatic cysts without a history of acute pancreatitis. His mother had died of pancreatic cancer with multiple pancreatic cysts and diabetes mellitus. A blood examination showed elevated HbA1C (9.0%) and CA19-9 (304 U/ml). A gene analysis revealed a mutation in PRSS1 (N29I). Magnetic resonance cholangiopancreatography showed large cysts in the pancreas head and small cysts replacing the pancreas body and tail (Picture 1). Contrast-enhanced computed tomography showed a delayed-enhanced solid part at the center of cysts in the pancreas head (Picture 2). Endoscopic retrograde cholangiopancreatography revealed dilated branches without main pancreatic duct stricture (Picture 3). Total pancreatectomy was selected due to recurrence in the remnant pancreas. A histopathological examination showed pancreatic cancer surrounded by retained cysts in the pancreas head (Picture 4A), which originated from the epithelium of the cyst (Picture 4B). Highly atrophic pancreas parenchyma with dilated branches in the pancreas body and tail was also recognized (Picture 4C).

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