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Malignant Intraductal Papillary Mucinous Neoplasms of the Pancreas in Two Identical Twins

Enrico Dalla Bona[1], Valentina Beltrame[1], Stella Blandamura[2], Marco Pizzi[2], Claudio Pasquali[1], Cosimo Sperti[1]

[1]Third Surgical Clinic, Department of Surgery, Oncology and Gastroenterology, and
[2]Department of Medicine (DIMED); University of Padua. Padua, Italy

Context Intraductal papillary mucinous neoplasm (IPMN) is a rare pancreatic tumor defined as intraductal mucin-producing neoplasm with tall, columnar, mucin-producing epithelium. IPMNs have already been described in association with inherited genetic disorder including familial adenomatous polyposis and Peutz-Jeghers syndrome. However, description of familial history of IPMN is very rarely reported. We present two cases of malignant IPMN in identical twins. **Case report** A 54-year-old woman was admitted in August 2010 with epigastric pain and high serum levels of amylase and lipase. Abdominal CT revealed dilation of main pancreatic duct (5 mm) with multiple cysts in the head of the pancreas. Serum CA 19-9 was in the normal range, and positron emission tomography with CT acquisition (PET/CT) was negative. A diagnosis of mixed type IPMN was made, and the patient underwent pancreatico-duodenectomy. Pathologic examination showed a tubular, poorly differentiated adenocarcinoma, tubular type, in combined type IPMN (pancreatico-biliary type) of the head of the pancreas with high grade dysplasia (pT3N0M0 G3). Postoperative course was uneventful, and the patient underwent adjuvant chemo-radiotherapy. Three years after surgery, there was no evidence of tumor relapse. Her sister, a 57-year-old woman, was admitted in February 2013 for obstructive jaundice and weight loss. Serum CA 19-9 was 86.2 U/mL. Magnetic resonance of the abdomen showed a 4.8 cm, pluricystic mass of the head of the pancreas, with marked dilation (10 mm) of the main pancreatic duct. FNAC under EUS examination showed mucinous epithelial cells with low-moderate dysplasia. PET/CT revealed a pathologic uptake of the radiotracer in the pancreatic head and in the right colon. Colonoscopy did not show colonic lesions. In March 2013 the patient underwent pylorus-preserving pancreaticoduodenectomy. Pathologic examination showed a colloid, poorly differentiated adenocarcinoma in main duct IPMN (intestinal type) of the head of the pancreas (pT3N0M0G3). Adjuvant therapy with gemcitabine was started, and the patient is alive 3 months after operation, without tumor relapse. **Conclusion** Although rare, the coexistence of IPMN reported here in two identical twins, suggests that it is due to a genetic inherited factor that remains to be elucidated. Physicians should carefully study the familial history of patients with IPMN.