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Intraductal Papillary Neoplasm of the Pancreas Oncocytic: Why Resection Isn't Enough

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Abstract

Intraductal Oncocytic Papillary Neoplasm of the pancreas [IOPN] is the least common subset of intraductal papillary mucinous neoplasms [IPMN], and is an exceedingly rare cystic pancreatic tumor. In fact, IPMN makes up less than 5% of all pancreatic neoplasms, and IOPN makes up less than 5% of IPMN. Studies on this particular tumor are sparse to date, with only a few case reports and small series. It is distinct from other forms of pancreatic tumors in its regard to develop into invasive carcinoma, even after resection. The reason for such an aggressive recurrence after resection is the multifocal nature of IOPN; this elusive nature of IOPN gives practitioners a false sense of treatment success when a focus of IOPN is removed with clear margins, because a different focus may be close-by without being detected. Current treatment guidelines for benign IPMN recommend yearly surveillance with CT or MRCP for patients who undergo surgical resection without invasive characteristics. Additionally, even with negative surgical margins additional investigation with modalities such as pancreatoscopy is recommended in order to exclude non-contiguous spread of disease. This case illustrates the need for extreme vigilance both at the time of surgery and after in order to detect occult or recurrent tumor. There is a relative paucity of data on these neoplasms and it is possible more aggressive surgical approaches and surveillance are warranted.

Keywords: Intraductal oncocytic papillary neoplasm; Pancreas; Cyst

Description

A 79-year-old African American male presented with bilateral lower extremity swelling, anorexia, 15-lb weight loss, and shortness of breath. Past medical history included resection of a pancreatic intraductal oncocytic papillary neoplasm [IOPN] 2 years ago and a presumed unprovoked DVT 1 year ago. Surgical margins of the mass were negative, and 2 regional lymph nodes were also negative for metastatic

disease. The body and tail of the pancreas were resected, leaving behind the head. There were no signs of invasive disease at the time of resection, or during 1 year follow-up where CT scans of the chest/abdomen/pelvis showed no recurrent or metastatic disease. He hadn't followed-up in the past year. Medications were coumadin and ferrous sulfate. Vital signs revealed hypotension and tachycardia. Examination revealed a frail septuagenarian with decreased breath sounds at lung bases, areflexia, and 4+ pitting edema to the thighs. Laboratory studies were remarkable for anemia, thrombocytopenia, and an elevated cancer antigen 19-9 level of 192 U/ml (reference range: 0-35 U/ml). Alpha-fetoprotein, parathyroid hormone-related protein, carcinoembryonic antigen, thyroid functions, renal and liver function tests, and calcium were normal. CT scan of the pancreatic head showed post-surgical changes with no acute findings or pancreatic mass. CT scans of the chest/abdomen/pelvis revealed multiple hepatic and bladder masses, peritoneal carcinomatosis, anasarca, and pleural effusions. Bone scan revealed diffuse uptake in the axial skeleton. In light of widely metastatic disease, the patient elected for hospice care.

IOPN, the least common subset of intraductal papillary mucinous neoplasms [IPMN], is an exceedingly rare cystic pancreatic tumor. In fact, IPMN makes up less than 5% of all pancreatic neoplasms, and IOPN makes up less than 5% of IPMN. Studies on this particular tumor are sparse to date, with only a few case reports and small series. It is distinct from other forms of pancreatic tumors in its regard to develop into invasive carcinoma, even after resection. The reason for such an aggressive recurrence after resection is the multifocal nature of IOPN; this elusive nature of IOPN gives practitioners a false sense of treatment success when a focus of IOPN is removed with clear margins, because a different focus may be close-by without being detected. Current treatment guidelines for benign IPMN recommend yearly surveillance with CT or MRCP for patients who undergo surgical resection without invasive characteristics. Additionally, even with negative surgical margins additional investigations with modalities such as pancreatoscopy are recommended in order to exclude non-contiguous spread of disease. This case illustrates the need for extreme vigilance both at the time of surgery and after in order to detect occult or recurrent tumor. There is a relative

paucity of data on these neoplasms and it is possible more aggressive surgical approaches and surveillance are warranted.