The Natural History of a Branch Duct Intraductal Papillary Mucinous Neoplasm in a Patient with Lady Windermere Syndrome: A Case Report

Vien X Nguyen¹, G Anton Decker¹, Ananya Das¹, M Edwyn Harrison¹, Alvin C Silva², Idris T Ocal³, Joseph M Collins², Cuong C Nguyen¹

Departments of ¹Gastroenterology/Hepatology, ²Radiology, ³Laboratory Medicine/Pathology; Mayo Clinic. Scottsdale, AZ, USA

ABSTRACT

Context “Low-risk” branch duct intraductal papillary mucinous neoplasm (LRB-IPMN) is defined as pancreatic epithelial cellular proliferation of small branch ducts that lack malignant characteristics. At present, our understanding of the natural history of LRB-IPMN is still evolving. Lady Windermere Syndrome (LWS) is a disorder seen in nonsmoking women with no pre-existing pulmonary disease affecting the lingula and/or RML with Mycobacterium avium-intracellulare complex (MAC). We present a case with pancreatic adenocarcinoma after a six-year surveillance of LRB-IPMN in an asymptomatic elderly white woman with LWS. Case Report A 79-year-old woman was referred to our institution because of pancreatic cystic abnormalities and elevated carbohydrate antigen 19-9 (CA 19-9). While at our institution, she was also diagnosed with LWS. Multiple abdominal imaging studies, endoscopic retrograde cholangiopancreatography (ERCP), computer tomography (CT), and magnetic resonance cholangiopancreatography (MRCP) were performed in the ensuing 6 years, all consistent with LRB-IPMN. No progression was seen until year 6 when MRCP showed a 2 cm pancreatic cancer. Because of multiple comorbidities, the patient chose chemotherapy over a pancreaticoduodenectomy. She developed respiratory failure and died after one cycle of gemcitabine. Conclusions: LRB-IPMN may be a heterogeneous disease in which some cases can transform into malignant pancreatic neoplasms despite the absence of the so-called “high risk” features on imaging studies. Clinical management, therefore, requires individualized flexibility. In addition, when there is coexistence of LWS and pancreatic cancer, prompt diagnosis and treatment of LWS should be considered prior to chemoradiotherapy or surgery.