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**Type 1 Autoimmune Pancreatitis Associated with Mucinous Cystic Neoplasm***Christina DiCarlo MD, Jan Silverman MD, FASCP, Mark Bunker MD, Allegheny General Hospital, Pittsburgh, PA*

The relationship between autoimmune pancreatitis and pancreatic neoplasia is poorly understood. We report a case of a 26-year-old woman who presented with a 2-month history of upper abdominal pain. Computed tomography scan revealed a 5.8 cm cyst within the pancreatic tail. Fine needle aspiration revealed mildly atypical mucinous cells. Fluid analysis revealed elevated CEA (>4000 ng/mL) and amylase (>7000 U/L). A distal pancreatectomy was performed. On examination, a 6.5 × 6.5 × 5.5 cm unilocular cyst was identified. Histology revealed a low-grade mucinous cystic neoplasm with ovarian-like stroma that was positive for progesterone receptor. Surrounding the neoplasm and extending to peripancreatic fat but not to the pancreatic specimen edge was a band of dense fibrosis with lymphocyte-plasma cell aggregates in part involving venules. By immunohistochemistry, IgG4+ plasma cells were present in large numbers, forming up to 100% of IgG plasma cells (each up to 160/hpf). This process therefore met criteria for diagnosis of type 1 autoimmune pancreatitis/IgG4-related sclerosing disease (AIP) in association with mucinous cystic neoplasm (MCN). Previous literature includes small numbers of cases of concurrent type 1 AIP and pancreatic neoplasms, specifically ductal adenocarcinoma and intraductal papillary mucinous neoplasm. Opinions have been divided on whether the mutual association represents causation, result, or coincidence. In addition, pseudocyst formation has been noted in cases of type 1 AIP. However, only one previous report has linked a MCN with type 1 AIP. The authors attribute the association to inflammatory antitumor response via Th2 cytokines. Sampling of tissue surrounding a pancreatic neoplasm could lead to a misdiagnosis of AIP. Clinical, imaging, histologic, and immunohistochemical data, with relevant literature review, are presented.

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