Intraductal papillary mucinous neoplasm of the pancreas and IgG4-related disease: A coincidental association.

Coexistence of autoimmune pancreatitis (AIP) and pancreatic cancer, elevation of serum IgG4 levels in pancreatic cancer patients, and infiltration of IgG4-positive plasma cells in peritumorous pancreatitis have been described in a few reports. This study examined the relationship between intraductal papillary mucinous neoplasm (IPMN) of the pancreas and peritumorous IgG4-positive lymphoplasmacytic infiltrates. Serum IgG4 levels were measured in 54 patients with IPMN (median 70 years, 26 males and 28 females; 13 main duct type and 41 branch duct type). Histological findings focusing on dense lymphoplasmacytic infiltration, storiform fibrosis, and obliterative phlebitis were reviewed, and immunostaining with IgG4 and IgG was performed in 23 surgically resected IPMN cases (18 main duct type and 5 branch duct type). The presence of IgG4-positive plasma cells>10/hpf and an IgG4-positive/IgG-positive plasma cell ratio>40% were considered significant. Serum IgG4 levels were elevated in 2 (4%) IPMN patients. Significant infiltration of IgG4-positive plasma cells was detected in 4 IPMN cases (17%). The IgG4-positive/IgG-positive plasma cell ratio was>40% in all 4 cases. In one
case with a markedly elevated serum IgG4 level (624 mg/dL), typical lymphoplasmacytic sclerosing pancreatitis (AIP type 1) lesions surrounded the whole IPMN. In the 3 other cases, infiltration of IgG4-positive plasma cells with fibrosis was focally detected mainly in the periductal area around the IPMN. In a few patients with IPMNs, IgG4-positive plasma cell infiltration can occur in the peritumorous area. The association of an IPMN with AIP type 1-like changes seems to be exceptional and coincidental.