Autoimmune pancreatitis (AIP) has been established as a distinct form of chronic pancreatitis that is distinguishable from other types such as alcoholic, hereditary or obstructive chronic pancreatitis. AIP seems to be a global disease, since it has been reported in many different countries, especially from Japan, USA and Europe (Germany, Italy, United Kingdom). Typical histopathological findings in the pancreas in AIP include a periductal lymphoplasmacytic infiltration with fibrosis, causing narrowing of the involved ducts. The typical clinical features include presentation with obstructive jaundice/pancreatic mass and a dramatic response to steroids. However, while the reports from Japan describe uniform changes called lymphoplasmacytic sclerosing pancreatitis (LPSP) in the pancreas from AIP patients, the reports from Europe and USA distinguish two histopathologic patterns in AIP patients: one with the characteristics of LPSP and another with slightly different histological features, called idiopathic duct centric pancreatitis (IDCP) or AIP with granulocytic epithelial lesions (GELs). This article reviews the evidence that GEL-positive AIP or IDCP is a second type of AIP, distinct from LPSP, in regard to pancreatic pathology, immunology and epidemiology.