Abstract:

BACKGROUND: Autoimmune pancreatocholangitis (AIPC) is an emerging, not completely characterized disease. Aim of this study was the comprehensive evaluation of a series of AIPC patients, who were diagnosed and treated in a European institution between January 2003 and July 2006. METHODOLOGY/PRINCIPAL FINDINGS: Thirty-three patients with histologically confirmed AIPC were analyzed and compared to 20 patients with non-autoimmune chronic pancreatitis (CP) and 14 patients with primary sclerosing cholangitis (PSC). Clinical features and conventional histopathology were taken into account. Immunohistochemistry and real-time quantitative PCR were used for the characterization of the inflammatory infiltrate and the stromal reaction. AIPC was localized in the pancreatic head in 94% of the patients. Intra- and/or extrapancreatic biliary tract involvement was present in 64% of the cases. The number of infiltrating T-lymphocytes, macrophages and total plasma cells was significantly higher in AIPC than in CP (3-, 4- and 8-fold increase, respectively). The absolute number of IgG4-positive plasma cells was higher in AIPC than in CP and PSC (7-fold and 35-fold increase, respectively), but significance was only reached in comparison with PSC. CXCR5- and...
CXCL13-positive cells were almost exclusively detected in AIPC. CONCLUSIONS/SIGNIFICANCE: AIPC is mainly a disease of the pancreatic head with possible extension into the periphery of the gland and/or into the biliary tract/gallbladder. The morphology of AIPC, as well as the immune- and stromal reaction is characteristic and comparable between cases with and without biliary tract involvement. Immunological markers (IgG4, CXCR5, CXCL13) can be of diagnostic relevance in specific settings.