Autoimmune pancreatitis (AIP) has been extensively reported from Japan, Europe, and the United States. Whereas the descriptions of AIP from Japan have predominantly been based on the presence of a distinct clinical phenotype, reports from Europe and the United States describe at least 2 histopathologic patterns in patients' condition currently diagnosed as AIP, viz, lymphoplasmacytic sclerosing pancreatitis (LPSP) and idiopathic duct centric pancreatitis (IDCP) or granulocyte epithelial lesion (GEL)-positive pancreatitis. Although the 2 entities share common histopathologic features (periductal lymphoplasmacytic infiltration and peculiar periductal fibrosis), expert pathologists can accurately distinguish them based on other unique histopathologic features. Clinically, the 2 entities have similar clinical presentation (obstructive jaundice/pancreatic mass and a dramatic response to steroids) but differ significantly in their demography, serological characteristics, other organ involvement, and disease relapse. While LPSP is associated with elevation in titers of nonspecific autoantibodies and serum IgG4 levels, IDCP does not have definitive serological autoimmune markers. All experts agreed that the clinical phenotypes associated with LPSP and IDCP should be nosologically...
distinguished; however, their terminology was debated. Whereas most experts agreed that the entities should be referred to as type 1 and type 2 AIP, respectively, others had concerns regarding use of the term “autoimmune” to describe IDCP.