
Abstract:
Autoimmune pancreatitis (AIP) has been extensively reported from Japan, Europe, and the USA. While the descriptions of AIP from Japan have predominantly been based on the presence of a distinct clinical phenotype, reports from Europe and the USA describe at least 2 histopathologic patterns in patients diagnosed with AIP, namely lymphoplasmacytic sclerosing pancreatitis (LPSP) and idiopathic duct-centric pancreatitis (IDCP) or granulocytic epithelial lesion-positive pancreatitis. While the 2 entities share common histopathologic features (periductal lymphoplasmacytic infiltration and peculiar periductal fibrosis), expert pathologists can accurately distinguish them on the basis of other unique histopathologic features. Clinically, the 2 entities have a similar presentation (obstructive jaundice/pancreatic mass and a dramatic response to steroids), but they differ significantly in their demography, serology, involvement of other organs, and disease relapse rate. While LPSP is associated with elevation of titers of nonspecific autoantibodies and serum IgG4 levels, IDCP does not have definitive serologic autoimmune markers. All experts agreed that the clinical phenotypes associated with LPSP and IDCP should be nosologically distinguished; however, their terminology was controversial. While 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. and IAP.