Authors’ reply

We thank Hirano et al for evincing interest in our study and giving us an opportunity to clarify some specific issues regarding the subtypes of autoimmune pancreatitis (AIP).

We agree with Hirano and colleagues that lymphoplasmacytic sclerosing pancreatitis (LPSP) and idiopathic ductal-centric chronic pancreatitis (IDCP) show some differences in their clinical and histological patterns. As compared to patients with IDCP, patients with LPSP are older, have elevated serum IgG4 levels and a strong association with inflammatory bowel disease. Histologically, LPSP shows periductal lymphoplasmacytic infiltration, storiform fibrosis and obliterator phlebitis with infiltration of abundant IgG4-positive plasma cells, whereas IDCP shows prominent lobular inflammation consisting of neutrophils, lymphocyte, and plasma cells with characteristic granulocytic epithelial lesions (GELs) in pancreatic ductal epithelium. IDCP usually shows IgG4 immunostaining negativity. In terms of geographical distribution, IDCP with typical GELs is rarely found in east Asia, while it is common in Europe and the United States.

By definition, “autoimmune” pancreatitis is a distinct type of chronic pancreatitis that occurs due to an autoimmune mechanism. Although target autoantigens and the effector cells in LPSP and IDCP remain speculative, clinical and histological findings of LPSP and IDCP are both suggestive of autoimmune process and respond to steroid therapy. We differ from Hirano et al in their opinion that IDCP should be excluded from AIP. We believe that LPSP and IDCP should be regarded as two subtypes of AIP rather than as independent disease entities.

In our study population, some patients were suspected of having IDCP based on clinical manifestation; however, the histology of pancreatic biopsy specimen did not reveal GELs and inflammatory bowel disease was not associated in any patient. Currently, present diagnostic criteria for AIP may not fully cover the presence of IDCP because there is no serological marker for IDCP and current diagnostic criteria were made originally based on the findings of LPSP. As our study demonstrated, however, “assessment of steroid responsiveness after a 2-week trial” may have great diagnostic implication in the setting of suspected AIP with the continued need for differentiation from pancreatic cancer because both LPSP and IDCP have almost identical pancreatic imaging and dramatic response to steroid therapy.

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