LETTERS TO THE EDITOR

IgG4-related sclerosing cholangitis should be included as an exclusion criterion for the diagnosis of primary sclerosing cholangitis.

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TO THE EDITOR:

This year's study by Mendes et al. presented invaluable results regarding the frequency of high serum IgG4 concentrations in patients with PSC. PSC is a chronic cholestatic liver disease characterized by progressive destruction of the bile ducts and the eventual development of biliary cirrhosis. However, some PSC patients seem to respond to corticosteroid therapy while others do not. This suggests that PSC may be a heterogenous condition. To diagnosis PSC, the Mayo Clinic's criteria are now widely used (3). However, researchers at the Mayo Clinic have shown that nearly 10% of PSC patients have elevated IgG4 and that as many as half of this select group of patients may need to undergo a liver transplantation.

The role of IgG4 in patients with PSC has been used to differentiate clinical syndromes of atypical PSC cases. In 1991, Kawaguchi et al. first described clinical and pathological features of variant cases of PSC which was later known as sclerosing cholangitis complicated with autoimmune pancreatitis (AIP) (4). In 1995, Takikawa et al. analyzed 192 cases of Japanese PSC and found two peaks in the age distribution. Some cases in elderly patients were complicated with chronic pancreatitis, which was regarded as sclerosing cholangitis complicated with autoimmune pancreatitis (AIP-related sclerosing cholangitis) (5). Later, Nakazawa et al. reported atypical PSC, which corresponded to AIP-related sclerosing cholangitis (6). In 2004, Takikawa et al. analyzed 269 additional cases of Japanese PSC and showed that 7% of these cases had AIP (7).

In the present study by Mendes et al., 9% of PSC patients had an elevated serum IgG4. This study reveals that AIP-related sclerosing cholangitis may have been included among PSC cases in the United States. In addition, the study shows that patients who are suspected of having PSC may respond to corticosteroids could also meet the Mayo Clinic's criteria for PSC. In order to exclude such patients from the diagnosis of PSC, we propose adding IgG4-related sclerosing cholangitis with a high serum IgG4 concentration or abundant IgG4+ cell infiltrates as an exclusion criterion to the Mayo Clinic's diagnostic criteria (8). In Japan this was the consensus of an expert panel following a workshop at Digestive Disease Week Japan 2003 (7).

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