

Incidence of Cancer and Risk Factors for Complication by Cancer in IgG4-Related Sclerosing Cholangitis

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Objectives:

IgG4-related sclerosing cholangitis (IgG4-SC) is a biliary IgG4-related disease (IgG4-RD) that is characterized by an elevated serum IgG4 concentration and by extensive fibrosis and dense infiltration of the bile duct wall by IgG4-positive plasma cells. There are increasing reports of cancer as a complication of IgG4-RD. We prospectively investigated the incidence of cancer in IgG4-SC patients and attempted to identify risk factors for developing cancer in IgG4-SC.

Method:

We divided 48 IgG4-SC patients (M:F = 38:10, mean age 67 years old, median observation period 5.2 years) into two groups: a group with cancer (Group A, n=14) and a group without cancer (Group B, n=34). IgG4-SC was diagnosed according to the clinical diagnostic criteria for IgG4-SC proposed in 2012 (J Hepatobiliary Pancreat Sci 2012). The organs affected by IgG4-SC (more than one organ in some cases) were biliary tract in all 48 cases, pancreas (type 1 AIP) in 46 cases, a salivary gland in 19 cases, the respiratory tract in 10 cases, and a retroperitoneal organ (kidney and/or retroperitoneal fibrosis) in the 10 cases. To determine the relative risk of cancer in IgG4-SC patients, we calculated standardized incidence ratios (SIR) from the national cancer rates in Japan during the observation period.

Results:

A total of 16 cancers were diagnosed in 14 of the 48 IgG4-SC patients. The prevalence of cancer in the IgG4-SC patients was 29.1%, and their SIR risk of cancer was 4.5 (95%CI=2.7-7.6, p__0.0001). The cancer sites were the stomach, lung and colon in 3 cases each, the pancreas in 2 cases, and the esophagus, duodenum, thyroid gland, liver and urinary bladder in 1 case each. 2. Five of the 16 cancers were detected within one year after the diagnosis of IgG4-SC. The SIR of cancer was 7.8 (CI =3.1-19.6) in the first year after the diagnosis of IgG4-SC and 3.8 (CI=2.1-7.0) thereafter. 3. The results of multivariate analysis showed that the presence of retroperitoneal organ involvement was significant risk factors for developing cancer in IgG4-SC patients (p=0.04), and the serum IgG4 level and whether treated with a steroid were not significant risk factors.

Conclusions:

IgG4-SC patients were found to be at increased risk of developing cancer. The possibility of complication by cancer should be borne in mind when treating IgG4-SC patients in whom multiple organs are affected, especially when there is retroperitoneal involvement.