**Risk of malignancy and pathological study of malignant neoplasms in autoimmune pancreatitis patients**

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**Objective:**

The occurrence of various malignant neoplasms has been reported in autoimmune pancreatitis (AIP) patients. Several studies have demonstrated the infiltration of IgG4-positive plasma cells in various cancer lesions without AIP. However, the association between malignant neoplasms with AIP and the role of IgG4-positive cells in malignancies is not yet fully understood. We retrospectively investigated the incidence and pathological features of malignancies in type 1 AIP.

**Method:**

83 AIP patients in our hospital between April 2000 and February 2015 were enrolled in this study. In the first study, we investigated the clinical features of malignancies in AIP including the types of malignancies, time of diagnosis, and the association with steroid therapy. In the second study, we investigated tumor stage and distribution of IgG4-positive plasma cells in various cancer tissues.

**Results:**

The incidence of malignancies was 17 out of 83 AIP patients, and a rate of 22.7%. The largest number of patients had colon cancer, followed by lung cancer, pancreatic cancer, and gastric cancer. Among the 17 AIP patients with malignancies, 41.2% had their malignancies discovered within a year after the diagnosis of AIP. The 10 patients who developed malignant neoplasms underwent surgery or EMR, and there have been no AIP relapses after these procedures. Microscopic examination showed that IgG4-positive plasma cells had infiltrated pancreatic cancer tissues and colon cancer tissues with AIP. One pancreatic cancer patient had not received steroid treatment, the other patient had undergone steroid therapy when pancreatic cancer was discovered. On the other hand, there were only a few IgG4-positive cells in cancer lesions, and the number of IgG4-positive cells in patients without AIP was much less than in patients with AIP. There were no IgG4-positive cells in gastric cancer and liver cancer.

**Conclusion:**

Tumor cells may cause AIP by activating IgG4-related immune responses. Careful follow-up is recommended within one year after the diagnosis of AIP because of the high incidence of malignant neoplasms in AIP patients.