



Diagnosing IgG4-Related Sclerosing Cholangitis

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Abstract

Background: IgG4-related disease (IgG4RD) is a systemic autoimmune disorder characterized by tissue eosinophilic infiltration and fibrosis. The biliary system is commonly affected, leading to IgG4-related sclerosing cholangitis (IgG4-SC). The diagnosis of IgG4-SC is challenging due to its histological overlap with primary biliary cholangitis (PBC) and cholangiocarcinoma (CCA).

Methods: We conducted a retrospective analysis of 100 patients with suspected IgG4-SC who underwent liver biopsy. The histological features were compared between the IgG4-SC group and the PBC/CCA group.

Results: The IgG4-SC group showed characteristic features including dense lymphoplasmacytic infiltration, storiform fibrosis, and a high ratio of IgG4-positive plasma cells. In contrast, the PBC/CCA group showed bile duct damage, portal fibrosis, and a lower ratio of IgG4-positive plasma cells.

Conclusions: The histological findings in liver biopsies are crucial for the diagnosis of IgG4-SC. A high ratio of IgG4-positive plasma cells and the presence of storiform fibrosis are key features that distinguish IgG4-SC from PBC and CCA.

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