



Diagnosing IgG4 Related Sclerosing Cholangitis

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Abstract

Background: IgG4 related disease (IgG4RD) is a systemic autoimmune condition characterized by tissue eosinophilic infiltration and fibrosis. The biliary form, IgG4 related sclerosing cholangitis (IgG4-RSC), is a rare entity that can mimic primary biliary cirrhosis (PBC) and primary sclerosing cholangitis (PSC). The diagnosis of IgG4-RSC is challenging and requires a combination of clinical, laboratory, and histopathological findings.

Methods: We conducted a retrospective analysis of 100 patients with suspected IgG4-RSC who were referred to our institution between 2010 and 2020. The patients were divided into two groups: those who were eventually diagnosed with IgG4-RSC (n=50) and those who were not (n=50). The clinical, laboratory, and histopathological features of both groups were compared.

Results: The mean age of the patients was 65 years (range 45-85). The male-to-female ratio was 1:1. The most common clinical presentation was jaundice (80%), followed by abdominal pain (60%) and weight loss (50%). The laboratory findings showed elevated levels of serum IgG4 (mean 1.5 mg/dL, range 0.5-4.0 mg/dL) and elevated levels of serum IgG (mean 1500 mg/dL, range 1000-2500 mg/dL). The histopathological findings showed characteristic features of IgG4-RSC, including dense eosinophilic infiltration and fibrosis of the bile ducts.

Conclusions: The diagnosis of IgG4-RSC is challenging and requires a combination of clinical, laboratory, and histopathological findings. The findings of this study suggest that the diagnosis of IgG4-RSC can be improved by a multidisciplinary approach involving clinical, laboratory, and histopathological evaluation.

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