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Immunosuppressant treatment for IgG4-related sclerosing cholangitis: A case report

Jong-Sun Kim, Won Ho Choi, Kyung-Ann Lee, Hyun-Sook Kim

Abstract

BACKGROUND

Immunoglobulin G4-related disease (IgG4-RD) is a multi-system fibroinflammatory disorder that can involve any organ, including the salivary glands, pancreas, and biliary tree. Treatment of immunoglobulin G4-related sclerosing cholangitis (IgG4-SC) is similar to that for IgG4-RD, but progression is irreversible in some cases. We present a

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Background: Even though **IgG4-related** disease has gained increased attention worldwide, the diagnosis remains challenging. **IgG4-related sclerosing cholangitis** (IgG4-SC) is not well described in the western hemisphere and may mimic cholangiocarcinoma (CC), especially when occurring without other symptoms such as, e.g. concurrent pancreatitis or retroperitoneal fibrosis.

Author: Anke Mittelstaedt, Peter N. Meier, Eva D... **Publish Year:** 2018

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Background: **IgG4-related sclerosing cholangitis** (IgG4-SC) is the biliary manifestation of **IgG4-related** disease (IgG4-RD) but the presence of IgG4-SC in the porta hepatis is difficult to differentiate from hilar cholangiocarcinoma (HCCA). **IgG4-related** autoimmune hepatitis (**IgG4-related** AIH) is extremely rare and it is not fully clear whether **IgG4-related** AIH is a hepatic manifestation of IgG4 ...

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1. Introduction. Immunoglobulin G4-related **sclerosing cholangitis** (IgG4-SC) is a relatively uncommon but increasingly recognized entity. It was first described in 2001 by Hamano et al. [1] with a landmark study demonstrating elevated levels of serum IgG4 in patients with **sclerosing cholangitis** [1–4]. It is characterized by systemic inflammatory and **sclerosing** lesions with massive infiltration ...

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