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Clinical Image

Hypereosinophilia with IgG4 Associated Cholangiopathy

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Abstract:

IgG4-related sclerosing cholangitis (IgG4-SC) belongs to the spectrum of IgG4-related disease which encompasses many medical conditions. We report a case of a 59-year-old man who presented with clinical and radiologic features of primary sclerosing cholangitis but where hypereosinophilia along with an extremely high blood level of IgG4, liver infiltration with IgG4-positive plasma cell and a spectacular response to corticoids treatment, confirmed the diagnosis of IgG4-SC.

Key words: IgG4 cholangitis; hypereosinophilia; sclerosing cholangitis

Introduction

IgG4-related disease encompasses a variety of conditions, including Mikulicz's syndrome, chronic sclerosing sialadenitis, hypophysitis, Riedel thyroiditis, chronic interstitial pneumonitis, interstitial nephritis, autoimmune pancreatitis, retroperitoneal fibrosis, sclerosing cholangitis, and lymphadenopathy. [1] The present article reports the case of a 59-year old man who presented jaundice and hypereosinophilia in the setting of IgG4-related disease.

Case report

A 59-year-old man from Iraq presented with a 6 month history, of asthenia, progressive weight loss of 20 Kg, jaundice and pruritus. Laboratory tests revealed hypereosinophilia with a total count of 9680, elevated creatinine (212 µmol/l), with cholestasis (GGT = 770 U/L, alkaline phosphatase = 706 U/L, and Bilirubin level at 3.7 mg/dl). A non-enhanced MRCP showed a normal common bile duct with mild dilatation of the intrahepatic bile ducts and a diminished arborization of the biliary tree (FIG 1). Viral, autoimmune and metabolic workup was strictly normal. A bone marrow biopsy was performed to rule out a hypereosinophilic syndrome. It only revealed hypercellularity with eosinophilia. Stool analysis for parasitic infection and serologies for Fasciola hepatica, Distomatosis and Toxacara canis were negative.

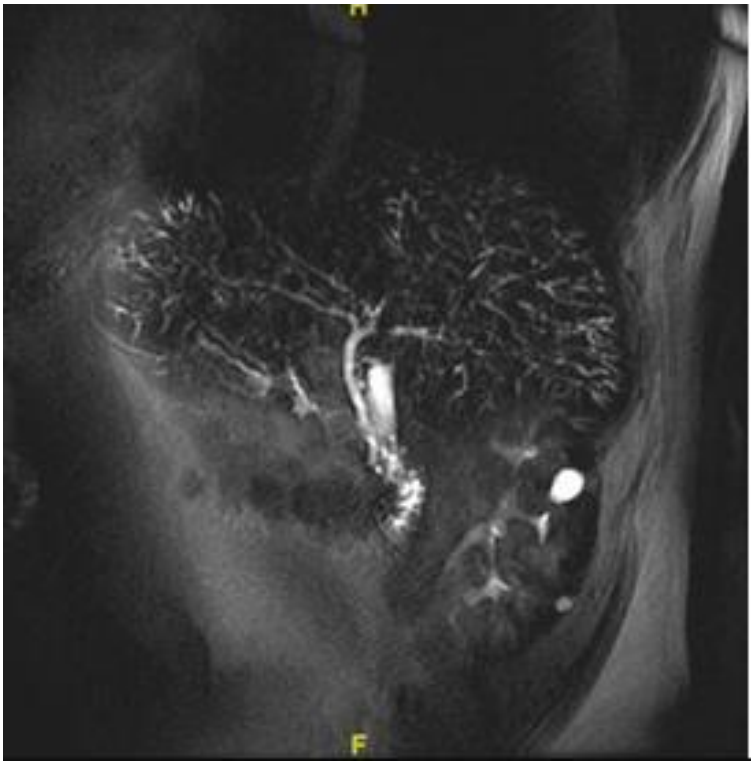

Figure 1: A non-enhanced MRCP: Normal common bile duct with a diminished arborization of the biliary tree

Subsequently, the patient underwent liver biopsy. Microscopic examination of the liver biopsy showed portal tracts edema, mixed inflammatory infiltrate with numerous plasma cells and neoductular proliferation. On immunostaining, the plasma cells were positive for IgG4. (FIG 2). IgG4 blood level was elevated to more than 6640 mg/100 mL, confirming the diagnosis of IgG4-associated cholangiopathy with hypereosinophilia as an associated feature.

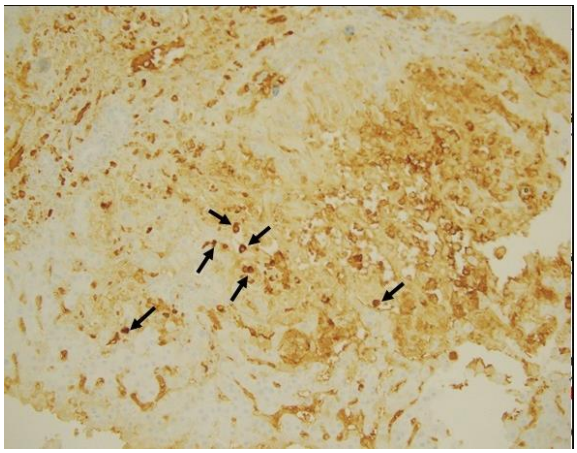

Figure 2: Liver biopsy: Plasma cells infiltrate positive for IgG4 on immunostaining

Treatment with oral prednisone 0.5mg/kg/day has normalized the eosinophil count, as well the liver enzymes and the creatinine levels. Two months after starting treatment, IgG4 level fell to 992mg/100ml and eosinophils count normalized. The patient was kept on maintenance therapy with Azathioprine 50 mg/day and 5 mg/day of prednisone.

Discussion

IgG4-related sclerosing cholangitis (IgG4-SC) belongs to the spectrum of IgG4-related disease which encompasses many medical conditions with the Type 1 autoimmune pancreatitis, the most common associated condition [1]. Five histological patterns are described in IgG4-SC: portal inflammation, large bile duct obstructive features, portal sclerosis, lobular hepatitis and canalicular cholestasis in perivenular areas [2]. More than 10 IgG4 -positive plasma cells/HPF is a commonly used threshold to suggest the diagnosis. A high IgG4 level can be observed in cholangiocarcinoma, primary sclerosing cholangitis (PSC), atopic dermatitis, and other cholangio-pancreatic malignancies. However, a cutoff level fourfold higher than the upper limit of normal had 100% specificity for IgG4-SC. Moreover, Torre et al revealed that peripheral hypereosinophilia and elevated serum IgE levels may be present in 27% and 35% of patients

respectively. These two features are not related to atopy but inherent to IgG4 disease itself (3).

The clinical, biological and MRI features in our patient, couldn't discern between IgG4-SC and PSC especially in the absence of any features of autoimmune pancreatitis. Liver infiltration with IgG4-positive plasma cell as well as the extremely high blood level of IgG4 and the spectacular response to corticoids treatment, confirmed the diagnosis of IgG4-SC.

Conclusion

The present case illustrate well the fact that hypereosinophilia is a feature of IgG4 related disease.

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