Summary

IgG4 – associated cholangitis or IgG-4 related sclerosing cholangitis (IgG4-SC) is a rare disease characterized by fibrosis and obliterative phlebitis in the bile duct wall, infiltration of IgG4 plasma cells, increased serum levels of IgG4 and a good response to steroids [1-2]. IgG4– associated cholangitis is part of the IgG4-related disease. Multiple organs can be affected such as the pancreas, kidneys, salivary glands, thyroids, lacrimal glands and arterial vessels. The prevalence is still unclear in Europe [3]. In Japan a prevalence of 0,9% per 100.000 individuals has been reported [4]. It is more frequent in men and usually presents between fifth and sixth decades of life. There is a strong association between IgG4 cholangitis and autoimmune pancreatitis (AIP) 2. IgG4-SC occurs in 20 – 80% of AIP cases 1. IgG4– associated cholangitis may mimic other biliary diseases as primary sclerosing cholangitis or cholangiocarcinoma. Moreover, it can lead to fibrosclerotic progressive disease and cirrhosis [5].

The diagnosis is based on clinical, laboratory and radiological findings [6-7]. The most common symptoms are jaundice and weight loss. 80% of the patients had elevated serum levels of IgG4 (cutoff value 135 – 140 mg/dl). Other abnormalities reported in the laboratory are hypergammaglobulinemia, IgG elevation and antinuclear antibody and positive rheumatoid factor [1, 2, 5]. Imaging studies allow confirming the presence or absence of biliary obstruction. The ultrasound could show biliary dilation, pancreas enlargement or both. The Computed Tomography of the Abdomen (Abdomen CT) or Magnetic Resonance cholangiogram (MRCP) are performed to obtain a better information of pancreatobiliary abnormalities. IgG4-SC could affect any part of the biliary tree. It is classified in: Type 1, low biliary duct stenosis; Type 2: intrahepatic and extrahepatic stenosis; Type 3: hilar and lower biliary common duct stenosis; Type 4: isolated hilar stenosis [1]. The pathological evaluation and immunostaining for IgG4 confirm the diagnose.

The treatment is based on high dose steroids, obtaining in the most cases remission of the disease. Endoscopic retrograde cholangiopancreatography (ERCP) could be necessary in cases of extrahepatic dominant stricture [1,2,5,8].

The aims of this study is create a Swiss registry of patients with IgG4- related Biliary Disease, to obtain data about epidemiology, diagnoses, clinical manifestations, treatment and evolution that provide a comprehensive description of clinical aspects of this disease.

References:

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