



Autoimmune Cholangiopathy at a Glance: Diagnosis, Differential Diagnosis and Treatment

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Editorial

Autoimmune cholangiopathy (cholangitis; AIC) is a chronic inflammation of liver that is considered a variant syndrome of autoimmune hepatitis. In 1997 Heathcote coined the phrase "autoimmune cholangitis" to describe anti-mitochondrial antibodies negative primary biliary cirrhosis (PBC). The patient usually suffers from fatigue, pruritus which may be very troublesome; clinical and/or laboratory features of cholestasis including persistent elevated levels of alkaline phosphatase and gamma-glutamyl trans peptidase enzymes combined with positive serological tests for antinuclear and/or anti smooth muscle antibodies and negative anti-mitochondrial antibodies are essential for diagnosis. Like PBC, it is characterized by a female predominance with female: male ratio of 10:1 and it slowly progresses to fibrosis and cirrhosis of the liver if left untreated [1,2]. However, AIC may still be difficult to be distinguished from some cases of sclerosing cholangitis (SC). Noninvasive differentiation between AIC and small duct primary or IgG4 SC as well as some overlap syndromes remains a yet to be settled clinical topic and some researchers consider to expand AIC to include several diseases and transition states of autoimmune hepatitis with bile duct damage. Further, Histological findings may be also indistinguishable. Currently, the only distinguishing clinical feature between small duct primary SC and AIC is the presence or absence of inflammatory bowel disease; respectively [3]. An elevated serum IgG4 level is a characteristic feature of IgG4 SC and the pancreas is the commonest organ involved other than the liver; other manifestations may include sclerosing sialadenitis, retroperitoneal fibrosis, and mediastinal lymphadenopathy [4].

The diagnosis of primary SC is also a challenge; unlike other autoimmune conditions, Primary SC has a male predominance of 2:1 and does not respond to corticosteroids. Patients are asymptomatic or display unspecific symptoms like fatigue, pruritus, and jaundice. Primary SC is frequently associated with inflammatory bowel disease and ulcerative colitis in particular and vice versa. Episodes of fever, chills and jaundice may reflect bacterial cholangitis from biliary obstruction rather than advanced disease [1,5]. The serum alkaline phosphatase and bilirubin may fluctuate, indicating transient biliary obstruction. Persistent jaundice suggests advanced PSC and should raise the suspicion of cholangiocarcinoma. ERCP has traditionally been the gold standard for the diagnosis of primary sclerosing cholangitis; however MRCP is increasingly being used a noninvasive alternative to ERCP [1]. Further, cholangio-NMR has undergone a significant technical evolution and is currently considered a preferred tool to investigate cholestatic disease showing similar sensitivity and specificity

to ERCP in the diagnosis of primary SC with fewer complications [5]. Noteworthy, IgG4 SC can be differentiated from the traditional Primary SC by applying the famous Mayo clinic HISORt criteria; in cases where serum IgG4 is less than 2x upper limit of normal and the ratio of IgG4/IgG1 > 0.24 is indicative for IgG4 SC [6].

Treatment of AIC is empiric and should be reserved mainly for those individuals who are symptomatic with jaundice, pruritus, and/or malaise, it consists of ursodeoxycholic acid which is also used for management of primary SC at a dose 13-15 mg/kg/day, corticosteroids which is also used for management of IgG4 SC and autoimmune pancreatitis starting with a standard initial dose is 40 mg per day of oral prednisolone, for 2-4 weeks. If there is obvious clinical and radiological improvement, the dose is decreased by 5 mg/1-2 week(s) or a combination of both. Steroid sparing immunosuppressive therapy with azathioprine and mycophenolate mofetil and rituximab maybe sometimes indicated for patients with a relapsing course requiring chronic immunosuppressive therapy. Moreover, cholestyramine is used in patients with moderate pruritus and the recommended dose is 4-16 gm per day in divided doses. Further, calcium and vitamin D should be recommended to all patients with chronic cholestatic liver disease in order to prevent metabolic bone disease. Once osteoporosis is established, bisphosphonates are recommended [1,3,7].

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