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Editorial

Primary Biliary Cirrhosis

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Editorial

Primary biliary cirrhosis (PBC) is an autoimmune liver disease of adults that can progress to liver fibrosis and cirrhosis. PBC mainly affects middle-aged women [1,2]. The mean proportion of female is 92% and the prevalence is increasing over the time [3]. The etiology of PBC is not clearly known thus it is believed that the disease appears in genetic predisposed population in exposure of environmental factors such as infection, chemicals and smoking [4].

The diagnosis is made based on combination of clinical signs and symptoms, abnormal liver function tests presenting for more than six months and detectable AMA (Anti Mitochondrial Antibody) in serum [4]. Ninety to ninety-five percent of patients are AMA positive although AMA is found in less than 1% of normal population [5].

About 60% of patients are asymptomatic at the time of diagnosis. Serum liver function tests and liver histology are used to assess severity of the disease and severities in symptomatic patients are less than symptomatic patients. The most common symptom in symptomatic patients is pruritus, 25% of patients have non-specific symptoms such right upper quadrant pain and lethargy, 20% of cases have jaundice, variceal hemorrhage or ascites. In asymptomatic patients the main reason of death is non-hepatic malignancies thus most important cause of death in symptomatic patient is liver failure or variceal bleeding [6].

Three groups of medications are effective in PBC treatment: Immuno-modulators, anti-fibrotics and anti-cholestatics. Currently, the first-line therapy is ursodeoxycholic acid (UDCA), an anti cholestatic [7]. In end stages especially when serum bilirubin level reaches to 100μmol/ l (5.9mg/dl) the best option is liver transplant [8].

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