Approach to Primary Biliary Cholangitis

Management in a District General Hospital

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Introduction Primary biliary cholangitis (PBC) is a rare progressive immune-mediated liver disease that, if not adequately treated, may culminate in end-stage disease and need for transplantation. Both genetic and environmental influences are presumed relevant to disease initiation. PBC is most prevalent in women and those over the age of 50, but a spectrum of disease is recognised in adult patients globally; male sex, younger age at onset (<45) and advanced disease at presentation are baseline predictors of poorer outcome. According to current guidelines, PBC is diagnosed in the presence of anti-mitochondrial antibodies (AMA) or specific antinuclear antibodies, and of a cholestatic biochemical profile, while biopsy is recommended only in selected cases. All patients receive ursodeoxycholic acid (UDCA) in first line; the only registered second-line therapy is obeticholic acid (OCA) for UDCA-inadequate responders.

Method Retrospective review of clinical records of patients with anti-mitochondrial antibody positive status were reviewed by help of local immunology department. Patients involved were the ones who were AMA positive between 2015 to 2020. 26 patients were included in this review and results are summarised below.

Results Out of 26 patients 11 were AMA positive only without any LFTs abnormality so no treatment was started, were planned to be followed up by yearly LFTs.

14 patients out of 26 were AMA positive and cholestatic abnormality; 6 patients out of 14 were on urso dose of 13 – 15mg/kg. 5 patient out of 14 had record of pruritis and fatigue documented. Only 3 patients out of 14 had DEXA scan done in last 5 years. Only 2 had developed cirrhosis who were not considered for transplant due to multiple co-morbidities but were followed by as per guideline for HCC and varices.

None of these 26 patients had overlapping diagnosis of autoimmune hepatitis.

Conclusion Significant issues were identified in management of patients with PBC when it comes to correct dose of ursodeoxycholic acid and mentioning about symptoms of fatigue and pruritis. Initial management needs to be optimized before consideration of obeticholic acid for these patients.