AUTOIMMUNE HEPATITIS


Take home points:
1. Think of autoimmune hepatitis, when more common causes of liver disease have been ruled out.
2. Think of autoimmune hepatitis in young women with cirrhosis and hypergammaglobulinemia.
3. Antibody tests in autoimmune hepatitis include anti-smooth muscle antibody and anti-LKM antibody.
4. Prognosis is generally good and treatment should be reserved for patients with active hepatitis and/or symptoms.

Epidemiology:
- 70% of patients with classic (type 1) autoimmune hepatitis (AIH) are females under the age of 40.
- However, AIH can affect patients of all ages and has a global distribution.
- Associated with other autoimmune diseases, including (in rare cases) antiphospholipid antibody syndrome.

Making the diagnosis:
- Before thinking of AIH, rule out other causes of chronic liver disease:
  - Hereditary (Wilson’s, hemochromatosis, alpha-1-anti-trypsin deficiency)
  - Viral (hepatitis A, B, C, etc)
  - Drug-induced (minocycline, INH, hydralazine, nitrofurantoin, etc)
  - Alcoholism
- A clue to the diagnosis is hypergammaglobulinemia (elevated total protein, decreased albumin, and decreased anion gap).
- A cholestatic picture on LFTs in AIH is extremely rare and should lead you to consider other diagnoses.
- Antibodies can help delineate the type of autoimmune hepatitis.
  - Type 1 AIH: anti-smooth muscle antibody; ANA also frequently positive.
  - Type 2 AIH: anti-LKM antibody, and LC1 antibody.
  - Type 3 AIH: anti-SLA/LP antibody.
- Liver biopsy is gold standard for diagnosis, but both biopsy and antibodies can be abnormal in other liver diseases (especially viral hepatitis).

Types of autoimmune hepatitis:
- Type 1 (classic) AIH: all ages, female predominance, caucasian predominance; > 30% have other autoimmune diseases (thyroiditis, RA, ulcerative colitis); can present abruptly, rarely leading to fulminant hepatic failure. Associated with anti-smooth muscle antibodies (ASMA).
- Type 2 AIH: usually presents during childhood and is associated with the picture of an autoimmune polyendocrine syndrome (type 1 DM, vitiligo, autoimmune thyroiditis, premature ovarian failure, mucocutaneous candidiasis); may be associated with low serum IgA concentrations and a higher frequency of progression to cirrhosis. Associated with anti-LKM antibody.
- Type 3 AIH: presents similarly to type 1 AIH but with a different antibody (anti-SLA/LP antibody).

Indications for treatment:
- Absolute indications: AST > 10× upper limit of normal; AST > 5× upper limit of normal + gamma globulin level > 2× upper limit of normal; bridging necrosis or multilobular necrosis in liver tissue.
- Relative indications: symptoms (fatigue, arthralgia, jaundice), serum AST less than absolute criteria but elevated; pathology of “interface hepatitis”.
- Don’t treat patients who have no symptoms or who have inactive cirrhosis.

Treatment regimens:
- Combination of prednisone + azathioprine or prednisone alone; liver transplantation always an option.