Autoimmune Hepatitis

Key Points:

- Autoimmune hepatitis is classified into two main types: Type I/Classic (ANA and ASMA) and Type II (ALKM-1 and ALC-1)
- AIH may present at any point on the spectrum of liver disease from asymptomatic elevated transaminases to fulminant hepatic failure
- Laboratory findings frequently include elevated serum immunoglobulins in addition to the autoantibodies
- AIH is strongly associated with other autoimmune diseases
- Treatment usually relies heavily on steroids and azathioprine and is usually fairly successful

I. Classification and Epidemiology: Two types
   a. Type I, a.k.a. “classic”: Characterized by ANA &/or anti-smooth muscle antibodies (ASMA)
      - ASMA titers of > 1:320 are thought to be reflective of anti-actin antibodies (AAA)
      - 10% will have antibodies to soluble liver antigens (anti-SLA)
      - A variety of other auto-antibodies may be present including pANCA, anti-dsDNA, anti-liver-pancreas protein, and many more
      - On occasion, anti-mitochondrial antibodies (AMA) may be present in association with the above autoantibodies; if isolated, however, this represents either primary biliary cirrhosis (PBC) or an overlap syndrome
      - Strong female predominance, but occurs in all age groups
      - Most commonly associated with thyroiditis, Graves’ disease, ulcerative colitis, and RA
   b. Type II: Characterized by the presence of anti-liver/kidney microsomal antibodies (ALKM-1) and/or anti-liver cytosol antibodies (ALC-1).
      - Almost exclusively in girls and young women
      - 22% have an associated autoimmune condition, most commonly type I DM, vitiligo, autoimmune thyroid disease, and polyglandular autoimmune syndrome

II. Clinical Manifestations: Range from asymptomatic to fulminant hepatic failure, from isolated elevated AST to full-blown cirrhosis
   - 6% of U.S. cases of fulminant hepatic failure are due to autoimmune hepatitis
   - Small joint arthralgias are a classic association
   - Associated with other autoimmune diseases as above

III. Laboratory Findings:
   - LFT’s generally show a more hepatocellular than cholestatic picture, but not in all cases
   - Elevated serum IgG levels
   - Autoantibodies as above
   - Non-specific histology with piecemeal necrosis in a periportal distribution

IV. Treatment:
   - Patients with symptoms should be treated; asymptomatic patients with mild inflammation may be left without treatment but should be very carefully monitored
   - Generally steroid-responsive (Prednisone 20-30 mg/day)
   - Azathioprine may be used as a steroid-sparing or combination agent
   - 10 year survival for treated patients is 90%, even in patients with advanced cirrhosis
   - Many patients can have their immunosuppressives reduced or eliminated once “remission” is achieved, though some will require suppressive azathioprine
   - For treatment failure: cyclosporine, tacrolimus, mycophenolate, liver transplant
   - AIH may recur after transplant, usually at the time when immunosuppression is reduced

References