Autoimmune liver diseases

Autoimmune liver diseases are characterized by histologically inflammatory infiltration of the portal fields by lymphocytes and plasma cells, serologically the presence of non-organ-specific antibodies, high IgG levels, and elevated aminotransferases.

- autoimmune hepatitis (AIH),
- autoimmune sclerosing cholangitis (ASC),
- de novo autoimmune hepatitis after liver transplantation for other liver diseases. [1]

Autoimmune hepatitis

2 types according to the spectrum of autoantibodies:

- Type 1 AIH: antinuclear antibodies (ANA) and/or anti-smooth muscle antibodies (ASMA), event. antibodies against cytoplasmic components of neutrophilic leukocytes (p-ANCA);
  - more common type, starting at puberty or adolescence;
- Type 2 AIH: antibodies against cytochrome P450 type 1 liver and kidneys (LKM), event. concomitant antibodies to hepatic cytosolic type 1 antigen (LC-1);
  - it manifests itself in young children, it is more aggressive;
- in both types, antibodies to liposoluble liver antigen (anti-SLA) may be usually impaired;

AIH mainly affects women (75-85%). The pathogenesis is not clear. About 20% of patients have associated another autoimmune disease.

Clinical picture

- often sudden development - picture of viral hepatitis: fatigue, anorexia, abdominal pain, development of jaundice;
- in type 2 up to the picture of fulminant liver failure with the development of encephalopathy;
- sometimes long-term symptoms: fatigue, headache, loss of appetite, weight loss, intermittent subicterus;
- fluctuating course, without therapy cirrhosis develops rapidly.

Diagnostics

- elevated aminotransferases, IgG (greater than 16 g/l) and some autoantibodies;
- ALP and GGT are usually normal or slightly elevated; reduced C4 complement level;
- liver biopsy - a typical picture (hepatitis interface), often already cirrhotic reconstruction.

Therapy

- immunosuppressants: prednisone, azathioprine. [1]

Autoimmune sclerosing cholangitis

In children, it partially overlaps with primary sclerosing cholangitis (PSC), which is predominantly described in adults. It affects girls more often. Almost half of the children also have chronic inflammatory bowel disease. Autoantibodies ANA and/or ASMA, p-ANCA are usually present. The ALP/AST ratio tends to be higher than that of AIH. It is often difficult to distinguish from AIH. The therapy is the same as for AIH.

Histology

- hepatitis interface image (inflammatory infiltration of portal fields by lymphocytes and plasma cells penetrating the lobes, necrosis of hepatocytes at the lobe edge, destruction of the limiting membrane) and inflammation of the bile ducts with focal dilatations and wrinkles (less pronounced changes than in PSC).[1]

Resources

Related articles

- Hepatitides
- Non-infectious chronic liver diseases

Reference

1. LEBL, J – JANDA, J – POHUNEK, P. *Klinická pediatrie.* 1. edition. Galén, 2012. pp. 362-363. ISBN 978-80-7262-772-1.