Autoimmune liver disease

Autoimmune liver diseases are characterized *histologically* by inflammatory infiltration of the portal fields by lymphocytes and plasma cells, *serologically* by the presence of organ-nonspecific antibodies, high IgG values and elevated aminotransferase values.

- autoimmune hepatitis (AIH),
- autoimmune sclerosing cholangitis (ASC),
- *de novo* autoimmune hepatitis after liver transplantation for other liver disease.

Content

Autoimmune hepatitis

2 types according to the spectrum of autoantibodies:

- 1st type of AIH: antinuclear antibodies (ANA) and/or anti-smooth muscle antibodies (ASMA), possibly antibodies against cytoplasmic components of neutrophil leukocytes (p-ANCA);
  - more common type, starts in puberty or adolescence;
- 2nd type of AIH: antibodies against cytochrome P450 of the 1st type of liver and kidney (LKM), possibly. simultaneously antibodies against liver cytosolic antigen type 1 (LC-1);
  - it manifests itself in young children, it is more aggressive;
- in both types, antibodies against liposoluble liver antigen (anti-SLA) may be present, usually worsening the course;

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

Clinical picture

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

Diagnostics

- elevated aminotransferases, IgG (greater than 16 g/l) and some autoantibodies;
- ALP and GGT usually normal or slightly elevated; reduced level of complement C4;
- liver biopsy – typical image (hepatitis interface), often also cirrhotic remodeling.

Therapy

- immunosuppressants: prednisone, azathioprine.

Autoimmune sclerosing cholangitis

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

Histology

- a picture of interface hepatitis (inflammatory infiltration of the portal fields by lymphocytes and plasma cells with penetration into the lobules, necrosis of hepatocytes at the edge of the lobule, destruction of the limiting membrane) and inflammation of the bile ducts with focal dilatations and puckering (less pronounced changes than in PSC).

Related Articles

- Hepatitis • Non-infectious chronic liver diseases

References

1. ↑Jump up to:a b c LEBL, J, J JANDA and P POHUNEK, et al. *Clinical Pediatrics.* 1st edition. Galén, 2012. 698 pp. pp. 362-363. ISBN 978-80-7262-772-1 .