Primary biliary cholangitis

Primary biliary cholangitis (PBC) is an autoimmune disease that primarily affects the liver with chronic destructive bile duct inflammation. The disease is also known as primary biliary cirrhosis, but this name was changed in 2014 because the disease does not necessarily lead to cirrhosis.

Young to middle age women are most likely to be affected (9 times more often than men). The incidence in Europe is approximately 5/100 000 inhabitants per year. Genetic predisposition in combination with external factors plays the main role in the etiology of this disease. Subsequently, small **intrahepatic** (interlobular and septal) bile ducts are attacked by cytotoxic T-lymphocytes.

Clinical picture

PBC is usually asymptomatic in early stages of the disease. As the time progresses, **pruritus**, **fatigue** and sometimes also **pain in the right hypochondriac region** typically occur. Dry mouth is another common subjective complaint.

During physical examination, pruritus-induced skin excoriations may be noticeable at first. There are also common signs of liver damage, including jaundice, hepatomegaly, and symptoms of advanced liver cirrhosis (e.g., ascites or spider naevi). In advanced stages, dry skin or xanthomata is observed.

Diagnostics

PBC is diagnosed by a combination of laboratory examination of **biochemical cholestasis markers** (serum ALP and GGT), immunological determination of **AMA antimitochondrial antibodies** (positive result is highly sensitive and specific) and histological verification using a **liver biopsy**.

Primary sclerosing cholangitis as a differential diagnosis

Primary sclerosing cholangitis (PSC) appears to be related to the PBC. The clinical picture of pruritus and fatigue is also very similar, but it is necessary to mention some principal differences:

1. PSC, unlike PBC, is often associated with inflammatory bowel disease, especially **ulcerative colitis**.
2. Men are affected by PSC more often than women.
3. PSC is diagnosed mainly using MRCP.
4. The prognosis of PBC in patients responding to UDCA treatment is better than with PSC.[1]

Treatment

First-line treatment involves the use of **ursodeoxycholic acid (UDCA)**. Patients' prognosis is very good if symptoms alleviate or disappear and serum ALP decreases. **In the second-line treatment**, **obeticholic acid (OCA)** is used in case of insufficient response.

At the same time, **symptomatic** therapy of pruritus and bone disease prevention are important. In the case of terminal stages progressing to liver cirrhosis or refractory pruritus, **liver transplantation** is the treatment of choice.

Complications

Metabolic bone disease is a common complication that increases the morbidity of patients with PBC. It includes osteoporosis and osteomalacia due to impaired enterohepatic bile acid circulation, often in combination with hepatic insufficiency disrupting vitamin D metabolism.[2] In addition to vitamin D substitution, regular monitoring by dual-energy X-ray absorptiometry is recommended.

The disease is also complicated by the coexistence of other autoimmune diseases, the most serious being the so-called **overlap syndrome** with autoimmune hepatitis.

Prognosis

PBC progresses slowly without any treatment and leads to liver failure in about 10 years. Administration of ursodeoxycholic acid significantly slows down the progression of the disease and allows the doctors to perform liver transplantation. Transplant treatment shows good results, 1-year survival is higher than 90%, 5-year survival is reported to be over 80%.

References

Related articles

- Primary sclerosing cholangitis

Literature

- ČEŠKA, Richard, et al. *Interna.* 2. edition. Triton, 2015. pp. 472. ISBN 978-80-7387-885-6.
- FEJFAR, Tomáš – VAŇÁSEK, Tomáš – HŮLEK, Petr. Guidelines of the Czech Society of Hepatology for diagnosis and treatment of primary biliary cholangitis. *Gastroenterologie a hepatologie.* 2018, vol. 2, no. 72, p. 109-118, ISSN 1804-7874. DOI: 10.14735/amgh2018109 (http://dx.doi.org/10.14735%2Famgh2018109).

Citations

1. SAFER, Ricky. *Distinguishing between PSC and PBC after a name change* [online]. [cit. 2020-07-06]. <https://www.healio.com/news/hepatology/20170320/guest-commentary-distinguishing-between-psc-and-pbc-after-a-name-change>.
2. SHIBATA, Hidetaka. Bone Disease in Primary Biliary Cirrhosis. *Clin Calcium.* 2015, y. 25, no. 11, p. 1, ISSN 0917-5857.