Autoimmune pancreatitis

Autoimmune pancreatitis (AIP) is a rare autoimmune disease. It represents 2-4% of all chronic pancreatitis. It occurs **in two forms** differing in geographical distribution, histological picture, and other characteristics:

- **Type 1 AIP** (lymphoplasmocytic sclerosing pancreatitis) is an IgG4 associated disease located in the pancreas. It rather affects elderly patients.
- **Type 2 AIP** affects younger patients, the risk of relapse is significantly lower than in type 1 AIP. It is often associated with idiopathic intestinal inflammation.

Both types are characterized by painless **jaundice**, sudden **diabetes mellitus**, radiologically detectable pancreatic enlargement, and a history of autoimmune disorders. Both types respond very well to **corticosteroid** therapy.

The HISORt diagnostic criteria from the Mayo Clinic (2006) for autoimmune pancreatitis are determined in such a way that at least one of the symptom groups must be met for the diagnosis of AIP.

Histopathological finding: at least one of the following must be met:

1. lymphoplasmacytic infiltrate with storiform fibrotization and obliterative phlebitis
2. lymphoplasmacytic infiltrate with storiform fibrotization and at least 10 IgG4 + plasma cells at the highest magnification

Clinical finding: all criteria must be met

1. CT resp. MRI displays diffusely enlarged pancreas with delayed and peripheral gain
2. diffusely enlarged and irregular ductus pancreaticus
3. the IgG4 level exceeds 1.4 g/l

Therapeutic experiment: all criteria must be met

1. unexplained pancreatic disease after excluding other known causes
2. elevation of serum IgG4 or histologically confirmed the involvement of another organ with IgG4 + cell infiltration
3. remission or significant improvement in pancreatic and/or non-pancreatic disorders after corticosteroid therapy

References

Related Articles

- Chronic pancreatitis
- IgG4 associated disease

References

- DITE, P. – NECHUTOVA, H. – UVIROVA, M.. *Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub* [online]. [cit. 2021-12-9]. <http://biomed.papers.upol.cz/pdfs/bio/2014/01/03.pdf>.
- SUGUMAR, A. – CHARI, S.. *Expert Rev Gastroenterol Hepatol.* [online]. ©2009. [cit. 2021-12-9]. <https://www.medscape.com/viewarticle/708921_1>.