See: Diagnosing autoimmune pancreatitis

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Immunology | Pancreas

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Type 1 and type 2 autoimmune pancreatitis are distinct diseases with different histology, epidemiology, and clinical presentations, requiring systematic evaluation to exclude pancreatic cancer.

KEY POINTS

- Type 1 AIP is a manifestation of IgG4-related systemic disease affecting older men (sixth or seventh decade) and is much more prevalent in East Asia (13–20 per 100,000 in Japan versus below 1 per 100,000 in Western countries), while type 2 AIP affects younger adults and is associated with inflammatory bowel disease.
- Type 1 AIP typically presents with painless jaundice, discomfort, and often exocrine or endocrine insufficiency at diagnosis, frequently accompanied by sclerosing cholangitis, whereas type 2 AIP presents with recurrent acute pancreatitis episodes and epigastric pain between episodes.
- The speaker stated that storiform fibrosis and obliterative phlebitis are histologic hallmarks of type 1 AIP, not merely IgG4-positive cells, and that up to 40% of type 1 AIP patients have normal serum IgG4 levels.
- The speaker emphasized that IgG4 is a poor distinguishing parameter because elevated levels occur in beekeepers, up to 10% of pancreatic cancer patients, and patients with infectious diseases such as Mycobacterium avium, and that not every IgG4-positive serology or histology indicates IgG4-related disease.
- Diagnosis requires integrating five pillars of evidence with the primary goal of excluding pancreatic cancer, and no single test is reliable; the speaker stated that in cases with insufficient histology or persistent diagnostic uncertainty, resection may be necessary.
- The speaker recommended a case-by-case approach to repeat biopsy versus steroid trial, favoring repeat EUS-guided biopsy when AIP diagnosis is uncertain and only a focal pancreatic mass is present without other organ involvement, noting that cancer remains more likely than AIP even after one negative biopsy.

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