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Summary

Autoimmunity in HPB (Complete Session)

UEG Week Barcelona 2026 | Presentation

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Three cases illustrate diagnostic and therapeutic challenges in autoimmune hepatopancreatobiliary diseases, including steroid-refractory autoimmune hepatitis managed with tocilizumab, a suspected PBC-AIH variant that proved to be PBC responding to second-line therapy, and type 1 autoimmune pancreatitis mistaken for pancreatic cancer leading to unnecessary pancreatectomy.

KEY POINTS

- A 48-year-old woman with biopsy-proven severe autoimmune hepatitis developed azathioprine-induced cholestatic hepatitis (occurring in approximately 1 in 1000 patients as stated by the speaker), was switched to mycophenolate, but remained steroid-dependent at 10 mg prednisone for three years with multiple flare attempts at reduction.
- The speaker added tocilizumab for concomitant rheumatoid arthritis and reported the patient was subsequently tapered to 2.5 mg prednisone without flares, contrasting with the prior three years of frequent relapses; the speaker positioned this as a potential option in difficult-to-treat cases though no randomized trials exist for this indication.
- A 40-year-old woman with PBC (AMA-positive, elevated alkaline phosphatase) and persistently elevated transaminases underwent liver biopsy showing features suspicious for PBC-AIH variant, but the speaker reported transaminases normalized with obeticholic acid addition rather than immunosuppression, concluding the patient had PBC alone and that optimizing PBC-specific therapy can resolve hepatitic features.
- A 59-year-old man with a pancreatic head mass, normal serum IgG4, and brushing negative for malignancy underwent Whipple procedure complicated by bile leak requiring completion pancreatectomy; histology revealed type 1 autoimmune pancreatitis with storiform fibrosis and IgG4-positive plasma cells, illustrating that the speaker stated up to 5% of Whipple operations are performed for AIP mistaken as adenocarcinoma.
- The speakers emphasized that EUS-guided fine needle biopsy with cutting-edge needles has 80% diagnostic accuracy for autoimmune pancreatitis as stated, and that CA 19-9 and serum IgG4 are unreliable for distinguishing AIP from pancreatic cancer since 40% of type 1 AIP is IgG4 seronegative and both markers can be elevated in adenocarcinoma.

– For suspected PBC-AIH variant, the speakers recommended liver biopsy is mandatory when the predominant phenotype is PBC, and immunosuppression should be reserved for cases with severe interface hepatitis, as mild-to-moderate interface activity may reflect PBC alone and resolve with optimized PBC-directed therapy.

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