Autoimmunerkrankungen anderer Organe: Leber

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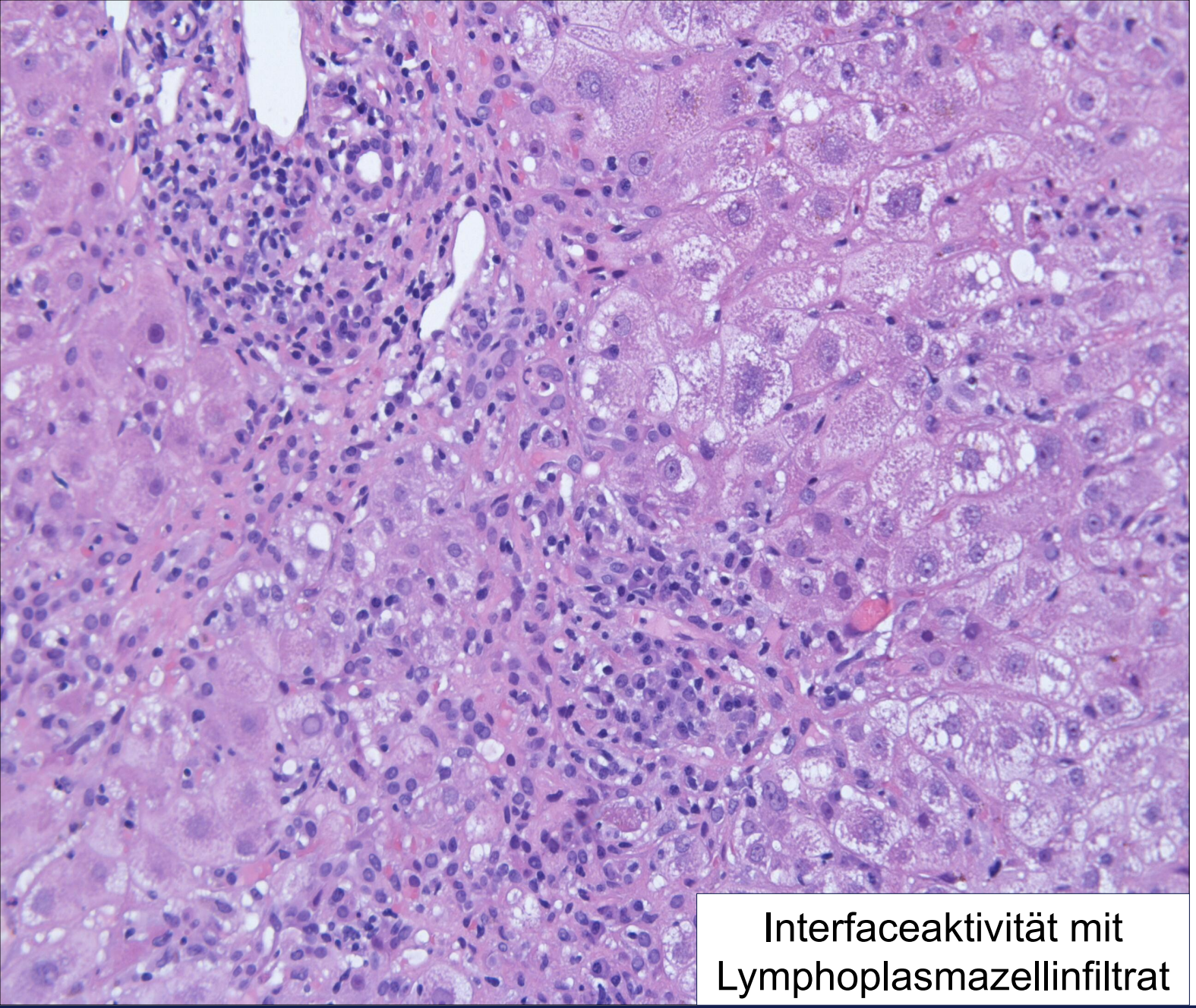
Klinische Präsentation

- 72 Jahre; männlich
- Arterielle Hypertonie; Diabetes mellitus Typ 2

- Schmerzloser Ikterus
- ASAT (GOT): 270 U/l (⊥: <50); ALAT (GPT): 624 U/l (⊥: < 50)
- y-GT: 519 U/l (⊥: < 60)
- Gesamt Bili: 1,4 mg/dl (bis 1,2); direktes Bili: 1,75 mg/dl (bis 0,3)
- alkal. Phosphatase: normal
- x-ANCA 1:160; SMA 1:160



Interfaceaktivität mit Lymphoplasmazellinfiltrat

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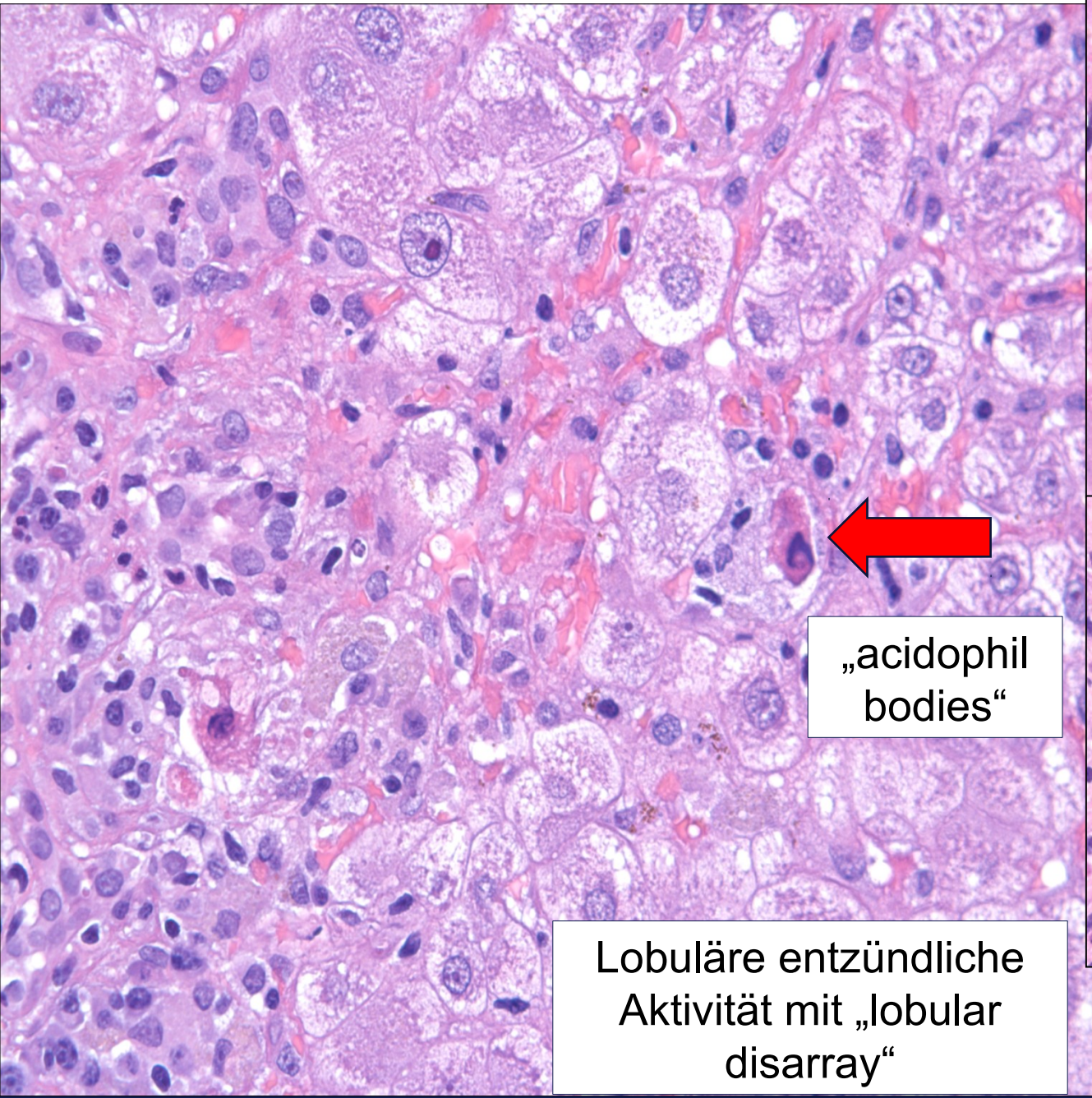
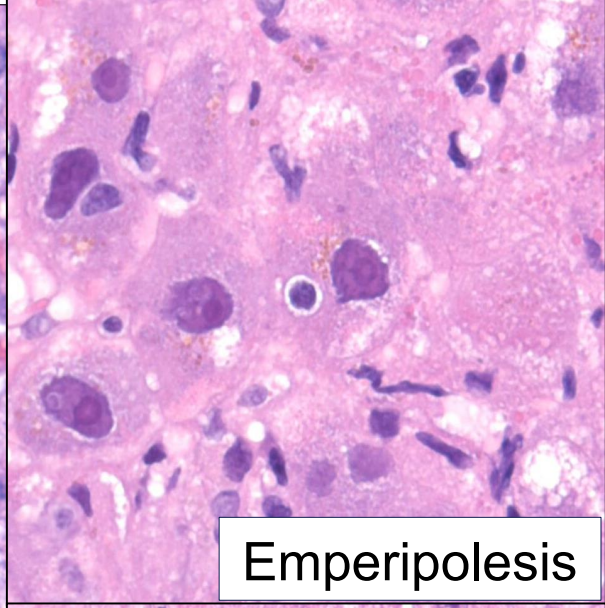
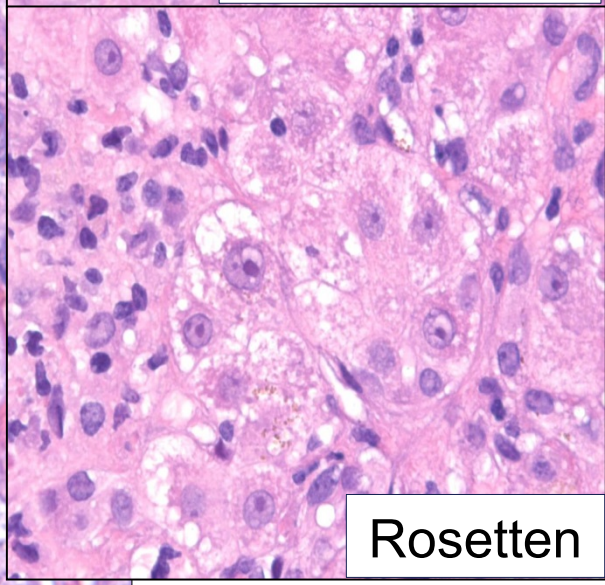


„acidophil bodies“

Lobuläre entzündliche Aktivität mit „lobular disarray“



Emperipolesis



Rosetten

Differentialdiagnosen

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DD Autoimmunhepatitis

Typical histology for AIH:

- Interface hepatitis, with lymphocytic/ lymphoplasmacytic in portal tracts and extending into the lobule
- Emperipolesis
- Hepatocyte rosette formation

Histology compatible with AIH:

- Chronic hepatitis pattern of injury with lymphocytic infiltration but lacking some of the features considered „typical“

Table 6. Simplified diagnostic criteria of the International Autoimmune Hepatitis Group [28].

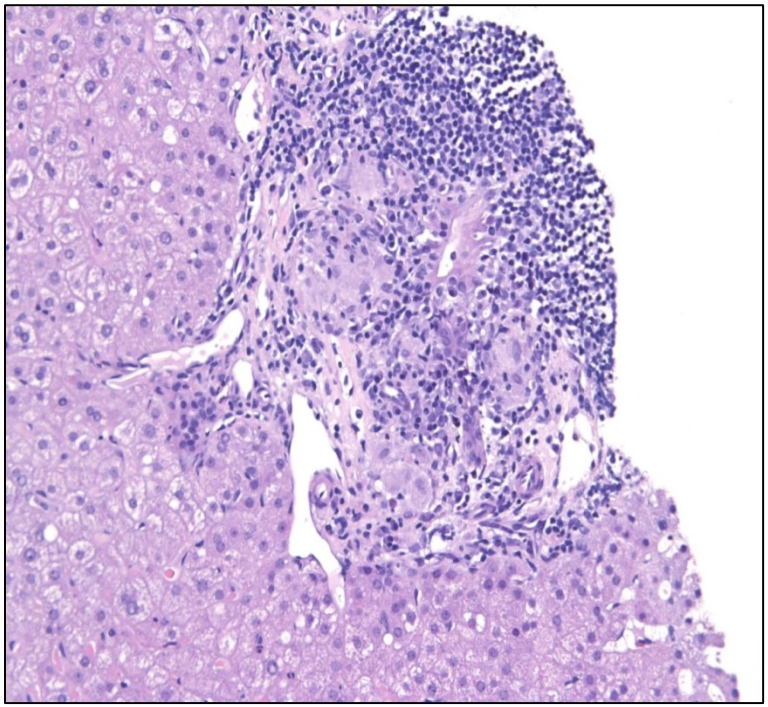
Feature/parameter	Discriminator	Score
ANA or SMA+	≥1:40	+1*
ANA or SMA+	≥1:80	+2*
or LKM+	≥1:40	+2*
or SLA/LP+	Any titer	+2*
IgG or γ-globulins level	>upper limit of normal >1.1x upper limit	+1 +2
Liver histology (evidence of hepatitis is a necessary condition)	Compatible with AIH Typical of AIH Atypical	+1 +2 0
Absence of viral hepatitis	No Yes	0 +2

Definite autoimmune hepatitis: ⩾7; Probable autoimmune hepatitis: ⩾6.
*Addition of points achieved for all autoantibodies (maximum, two points).

Hennes EM, Zeniya M, Czaja AJ, Pares A, Dalekos GN, Krawitt EL, et al. Simplified criteria for the diagnosis of autoimmune hepatitis. Hepatology 2008;48:169–176.

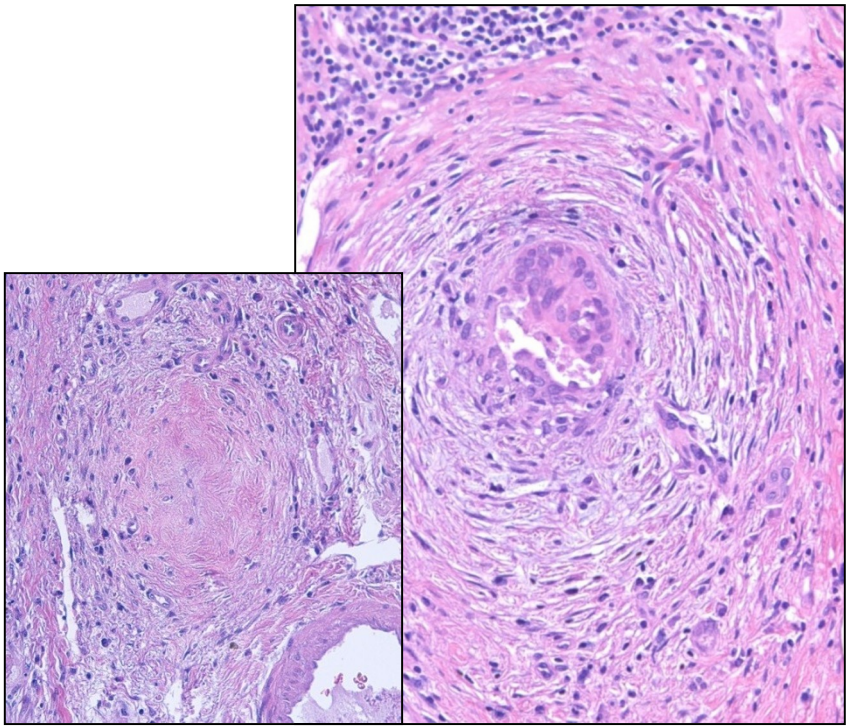
DD Primär biliäre Zirrhose

- Floride Gallengangsläsionen; Ductopenie
- AMA; Alkal. Phosphatase;
- γ-GT; IgM



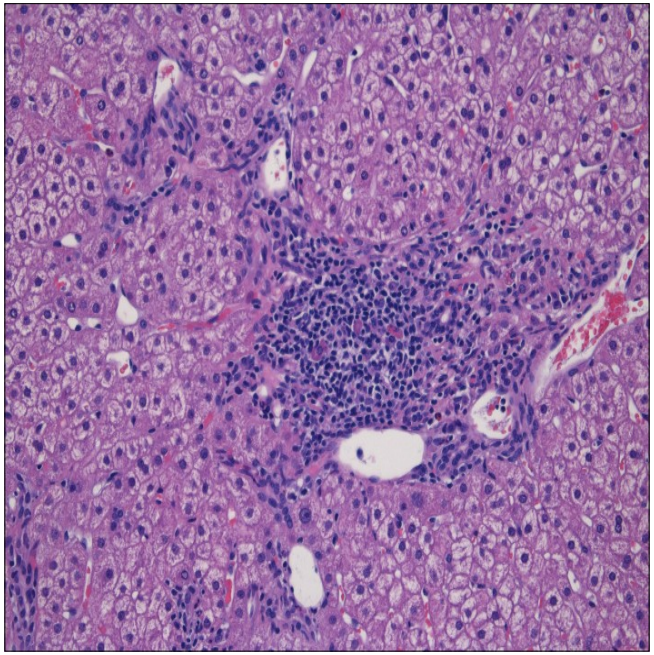
DD Primär sclerosierende Cholangitis

- Obliterative Fibrose; Ductopenie
- ANCA; Alkal. Phosphatase; γ-GT; IgM; IgG



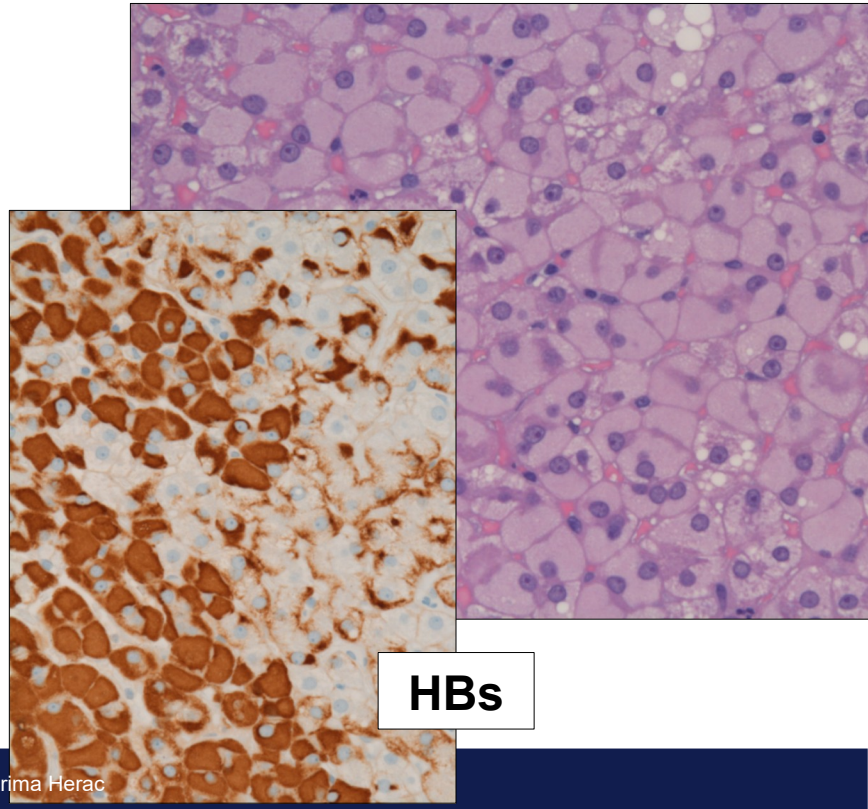
DD Virushepatitis: HCV

- Eher portales und periportales Infiltrat
- Milde Steatose
- HCV RNA im Serum



DD Virushepatitis: HBV

- Milchglashepatozyten
- HBV DNA, HBsAg und HBeAg im Serum



DD Med.-toxisch

- Medikamentenanamnese (auch Tee?)
- AIH Probability Score
- http://livertox.nih.gov/

Medikamente, die eine AIH triggern können:

Atomoxetine, Diclofenac, Imatinib, Inflixamab, Methyldopa, Methylphenidate, Minocycline, Nitrofurantoin, Ornidazole, Penylpropyluracil, Rifampin und Pyrazinamide, Statine,...

Tees, die eine AIH triggern können:

Black cohosh, Echinacea

Patient

- x-ANCA 1:160; SMA 1:160 **(2P)**
- Y-Globuline oberes Limit von Normal **(1P)**
- Histo typisch **(2P)**
- Abszenz einer viralen Hepatitis: ja **(2P)**
- **Summe: 7P**

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Autoimmunhepatitis

- Prävalenz: 15-25 Fälle/ 100.000 Einwohner in Europa
- eher Frauen betroffen; jedes Alter

Charakterisiert durch:

- Hypergammaglobulinämie, Autoantikörper (ANA, SMA, LKM1, SLA, LP, ANCA)
- Human leukocyte antigens (HLA) DR3 oder DR4
- Interfacehepatitis (Histo)
- Gutes Ansprechen auf Immunsuppressiva

Danke für Ihre Aufmerksamkeit!

Quellen

MC Sween`s Pathology of the Liver, 6th Edition

Surgical Pathology of the GI Tract, Liver, Biliary Tract and Pancreas, Robert Odze, 2nd Edition

EASL Clinical Practice Guidelines: Autoimmune hepatitis; Journal of Hepatology 2015 vol. 63; 971-1004