

Acute Hepatic Failure

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Medical History

- 49 y-old man, m+7
- Heavy smoker
- History of alcohol & drug abuse

Medical History - cont.

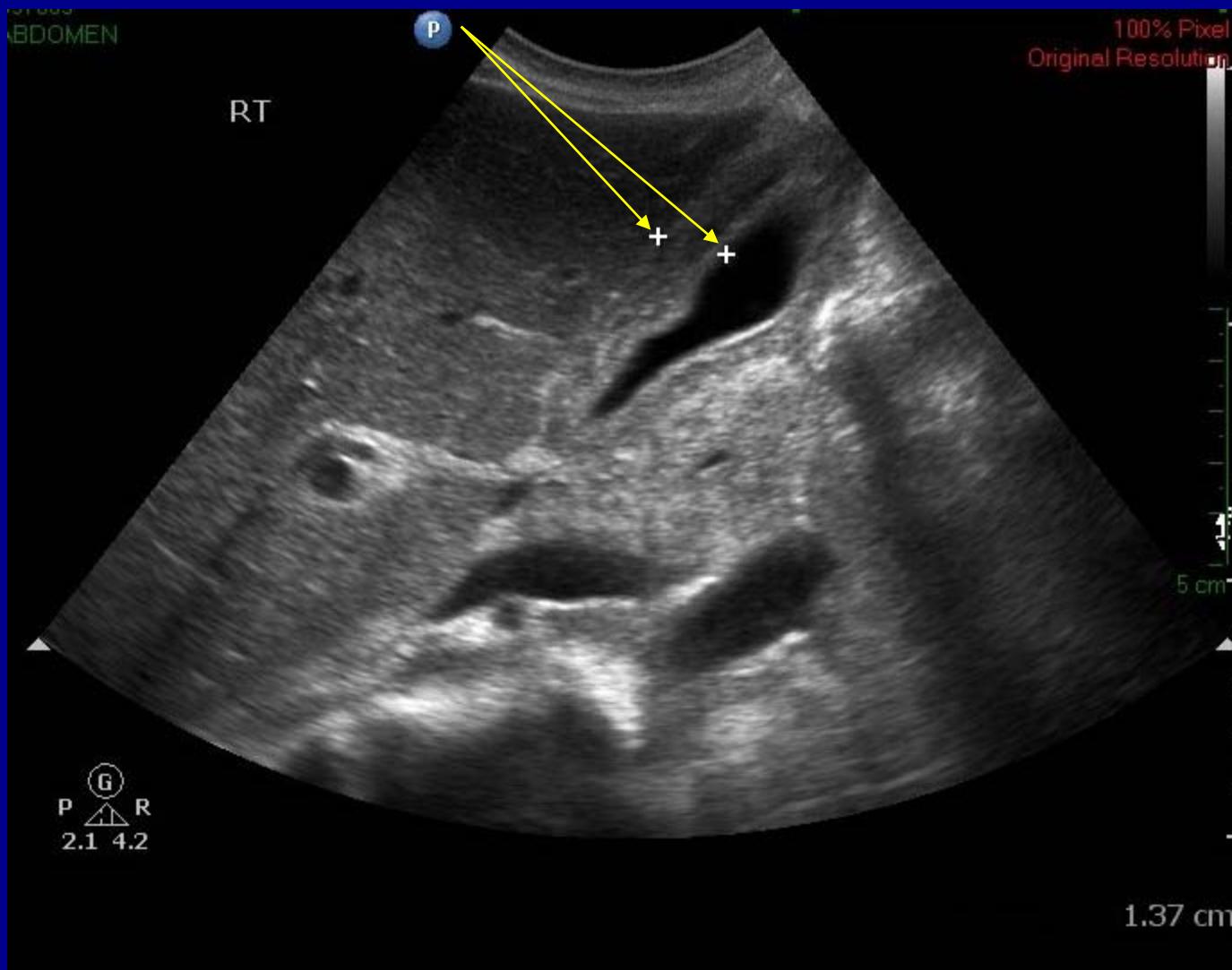
- Family history of IHD
- Normal coronary angiography-08/2006
- Medications:
 - Deralin
 - Micropirin

Current Disease

- Four days prior to admission:
 - Abdominal pain and swelling
 - Vomiting
 - Anorexia
 - Icterus
 - Low grade fever

	Baseline	Presentation	Admission
Alb			3.1
Bil T	0.5	1.6	1.7
Alk Phos	85	579	500
GOT	22	164	113
GPT	19	304	250
GGT	20	478	429
LDH	364	1052	499
HB	15.4	15.5	13.6
PLT	305	145	101
INR			1.1

US

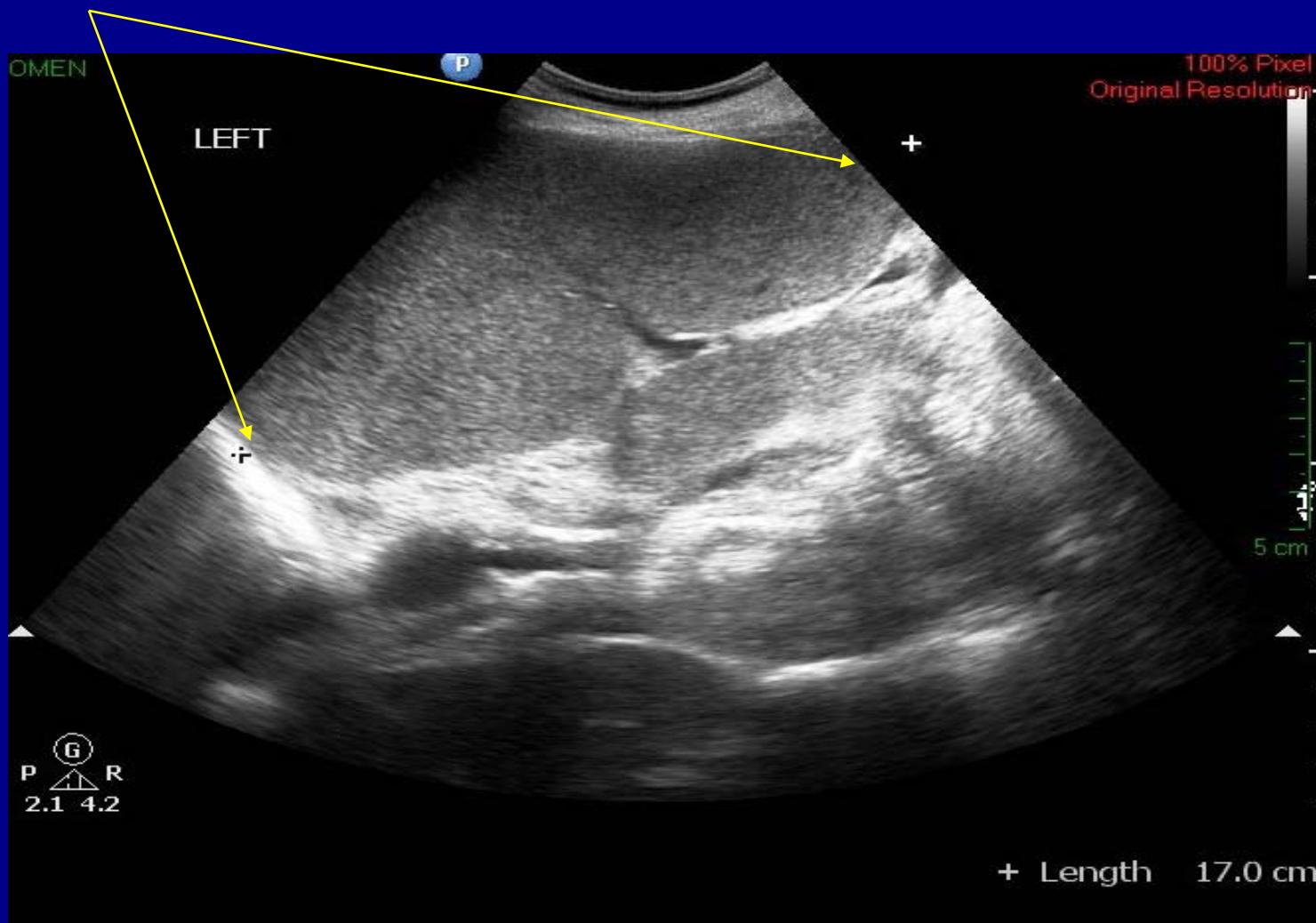


Conclusion

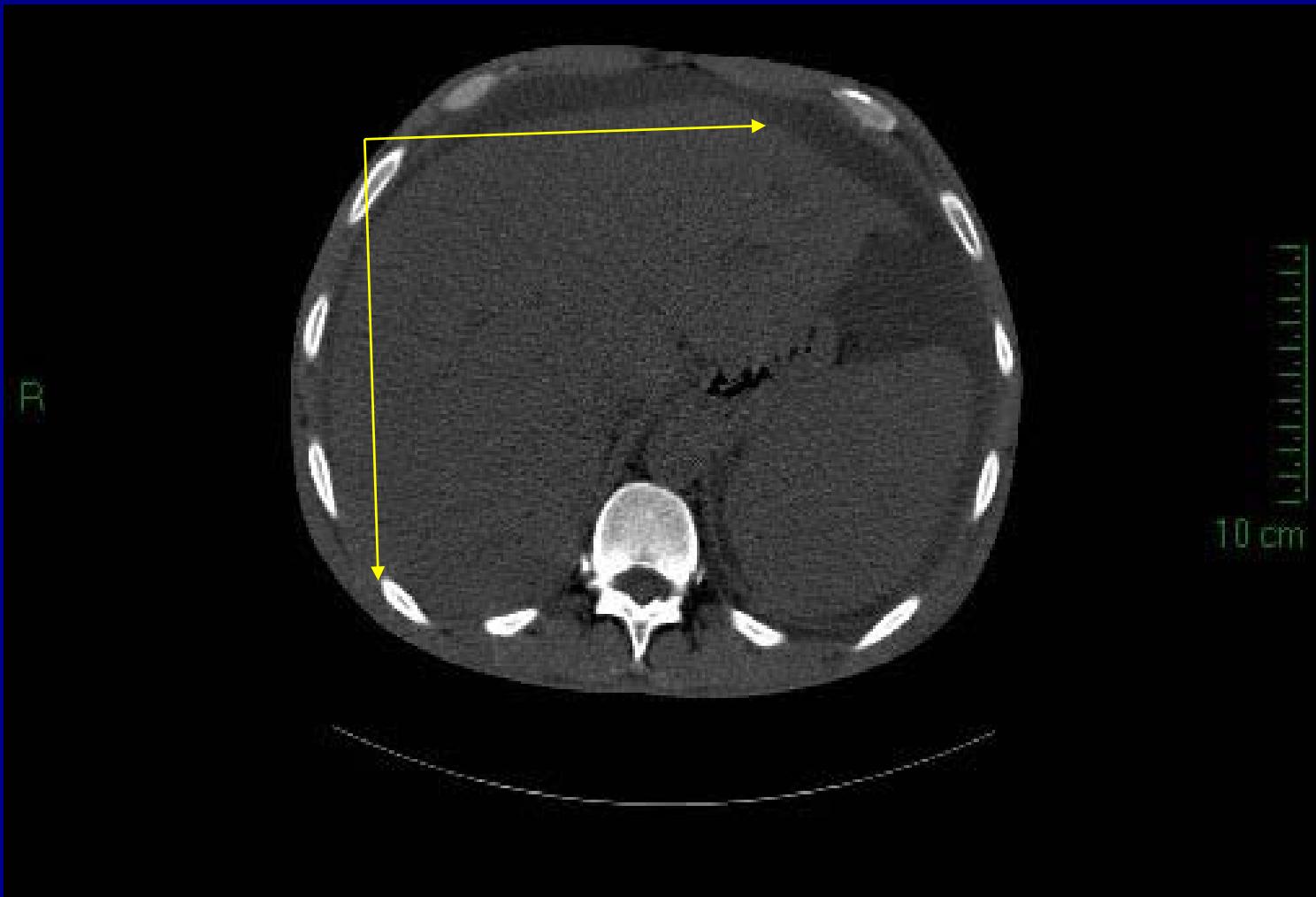
- 49 male
- New onset hepatitis
- Cholecystitis?
- Working diagnosis:
 - acalculous cholecystitis

	Baseline	Presentation	Day 8
Alb			2.5
Bil T	0.5	1.6	7.9
Alk			
Phos	85	579	1065
GOT	22	164	89
GPT	19	304	142
GGT	20	478	741
LDH	364	1052	404
HB	15.4	15.5	9.8
PLT	305	145	46
Amonia			151
INR		1.1	1.36

US - Day 8



CT



Follow Up

- Abdominal distress
- Anorexia
- Swollen abdomen
- Low grade fever

Lab Results - cont.

- HCV Ab - nonreactive
- HBsAg - nonreactive
- HAV IgM – nonreactive
- ANA, AMA, ASMA – negative
- IgG, IgM, IgA – normal
- Alpha 1 Antitrypsin – normal
- Ceruloplasmin – normal

Summary

- Acute hepatic failure
- Combined cellular and cholestatic abnormalities
 - mainly cholestatic
- Thrombocytopenia, elevated LDH
- Hepato-splenomegaly
- No peripheral lymphadenopathy



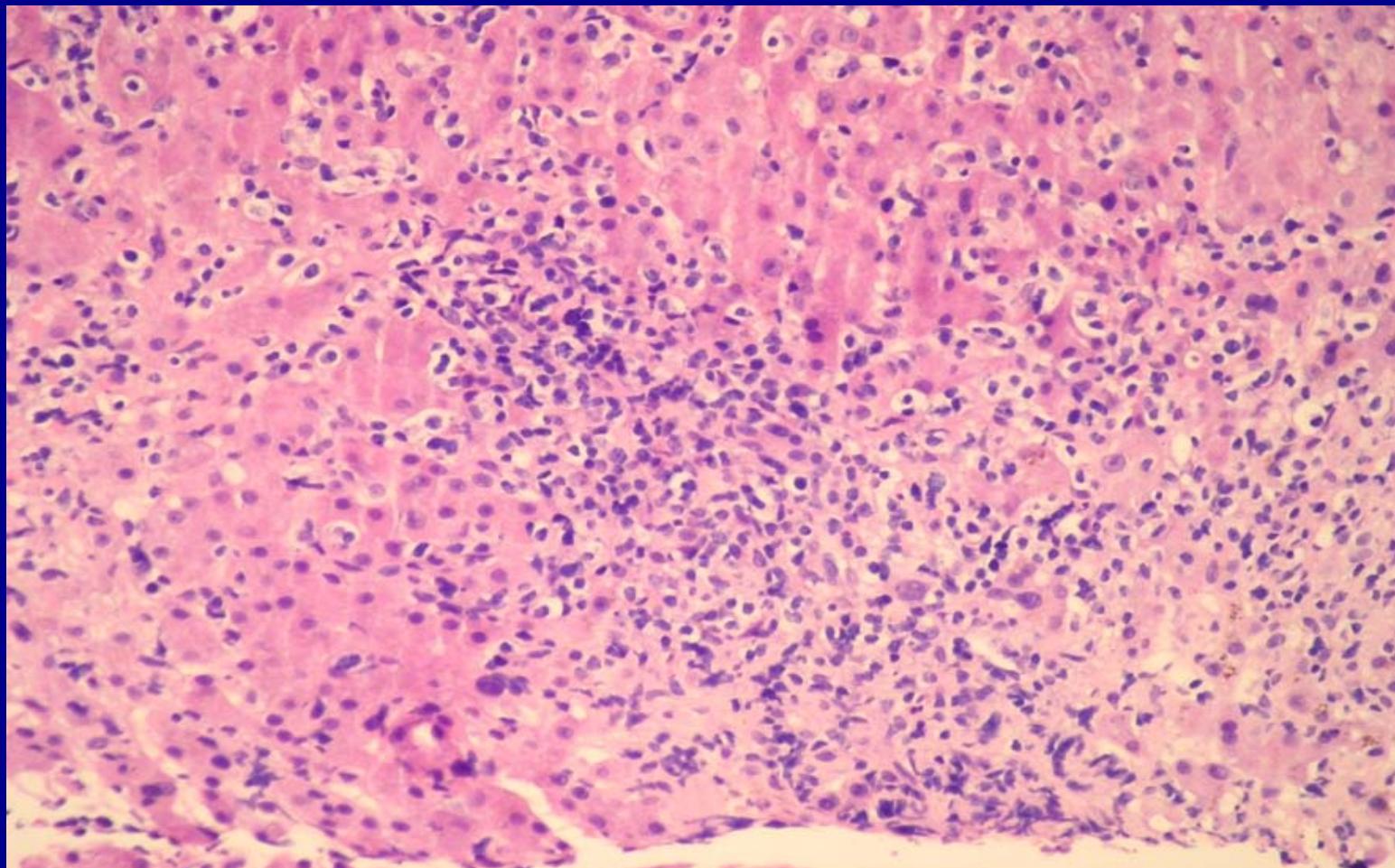
Further Investigation

- Trans-jugular liver biopsy

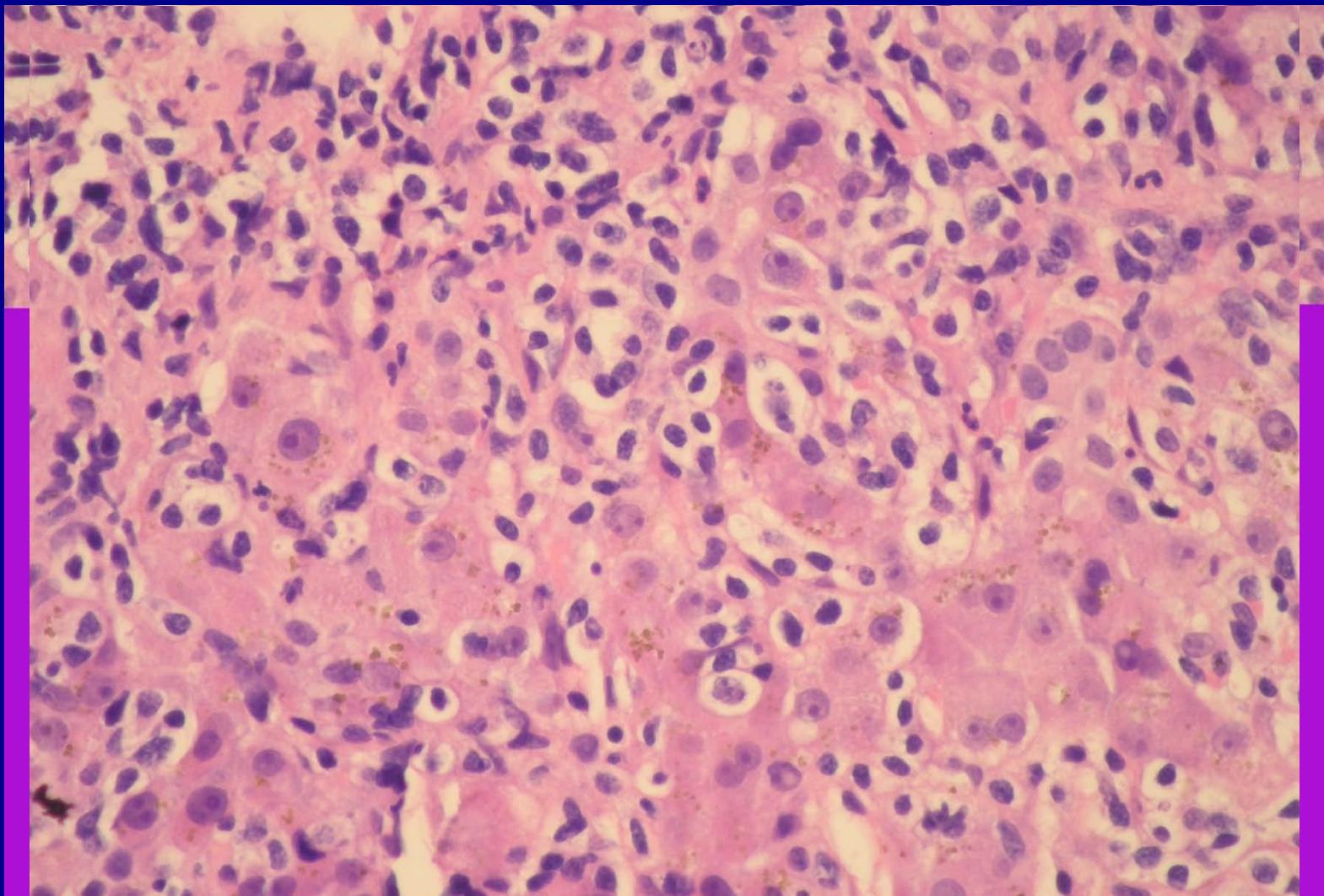


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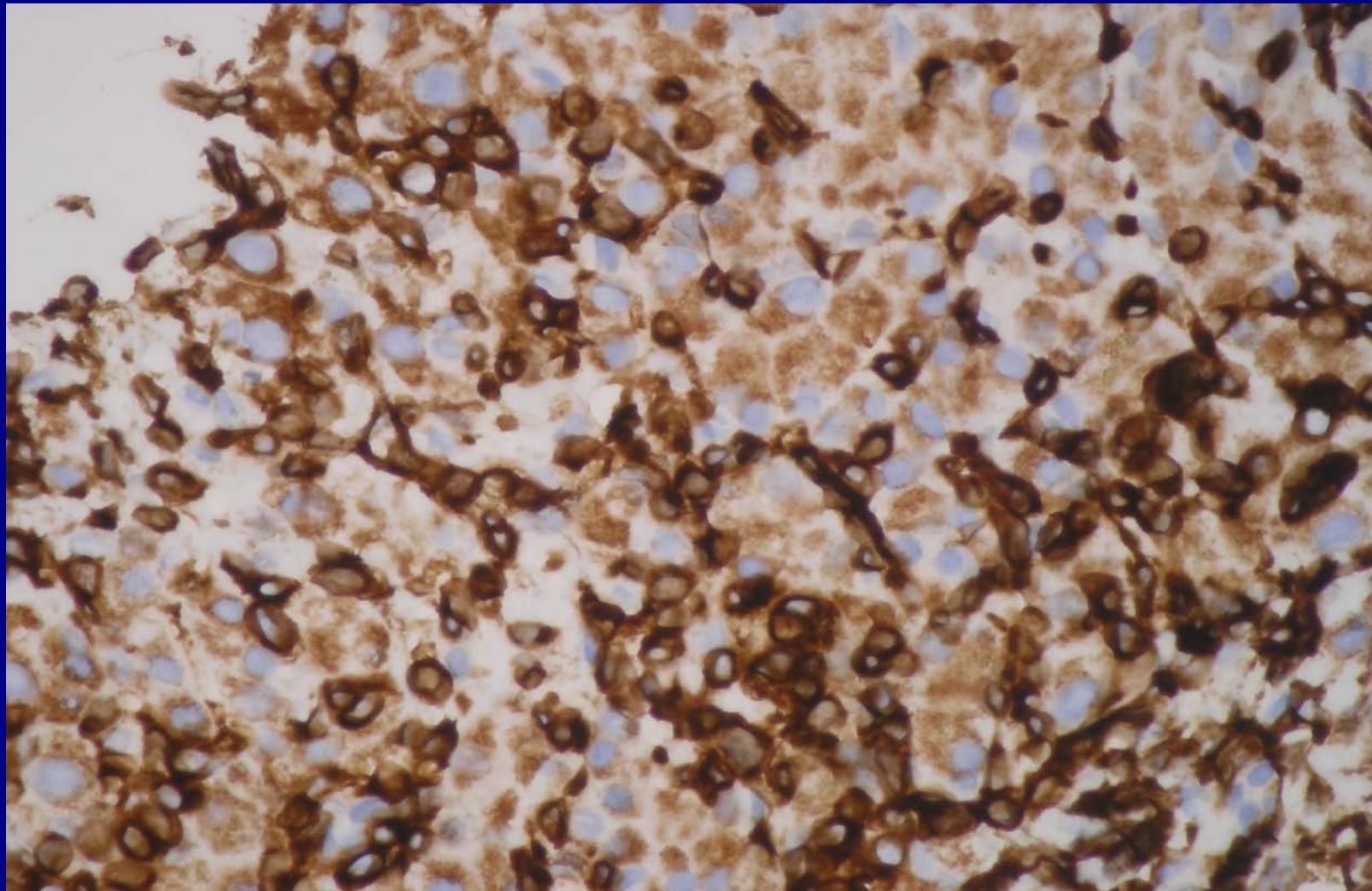
Pathology



Pathology



CD3 immuno-staining



Diagnosis

- Massive T cell lymphocytic infiltration
- Normal liver background
- Malignant T cell lymphoma of the liver

T- cell lymphoma

- PTCL, not otherwise specified (NOS)
- Anaplastic large cell lymphoma
- Angioimmunoblastic T –cell lymphoma
- Extranodal NK/T – cell lymphoma , nasal type
- Subcutaneous panniculitis-like T-cell lymphoma
- Enteropathy associated T – cell lymphoma
- Hepatosplenic T- cell lymphoma

Acute liver failure secondary to hepatic infiltration - DD

- Haematological malignancies - most common
 - Hodgkin's disease
 - Non-Hodgkin's lymphoma
 - Malignant histiocytosis
 - Leukemia
- Other infiltrative metastatic malignancies - rare
 - Adenocarcinoma
 - Melanoma
 - Anaplastic tumor of various primary sites

Hepatosplenic T- cell lymphoma

- An extranodal and systemic neoplasm derived from cytotoxic T cells

Epidemiology of HSTL

- Very rare neoplasm
 - < 1 % of NHLs
 - reported <100 patients
- Usually fatal
- Median age at diagnosis - 35 y
- Male predominance

Diagnosis

- Biopsies:
 - Accumulation of malignant cells
 - Hepatic sinusoids
 - Bone marrow
 - Spleen red pulp

Prognosis

- Very poor
 - aggressive course
- The average survival time from diagnosis
 - 8 months

Treatment

- Hematopoietic stem cell transplant - standard of care
- Autologous transplant - not a good option for HSTL
- Allogeneic bone marrow transplant - several reports
- All patients eventually relapse



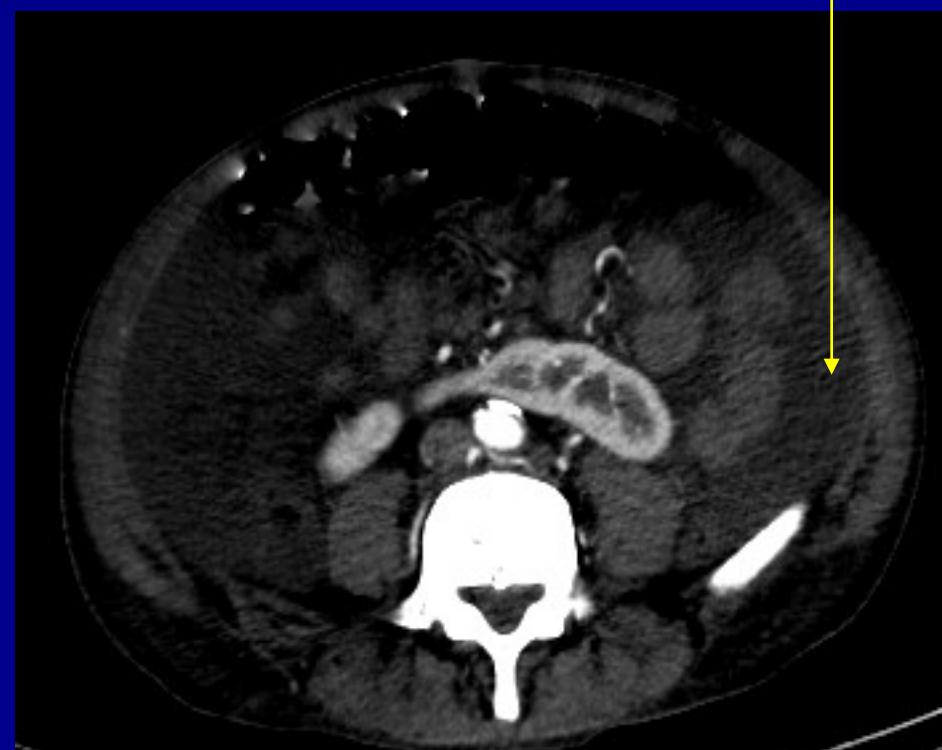
The most aggressive treatment strategy is not curative in hepatosplenic T-cell lymphoma !!!

Post Biopsy...

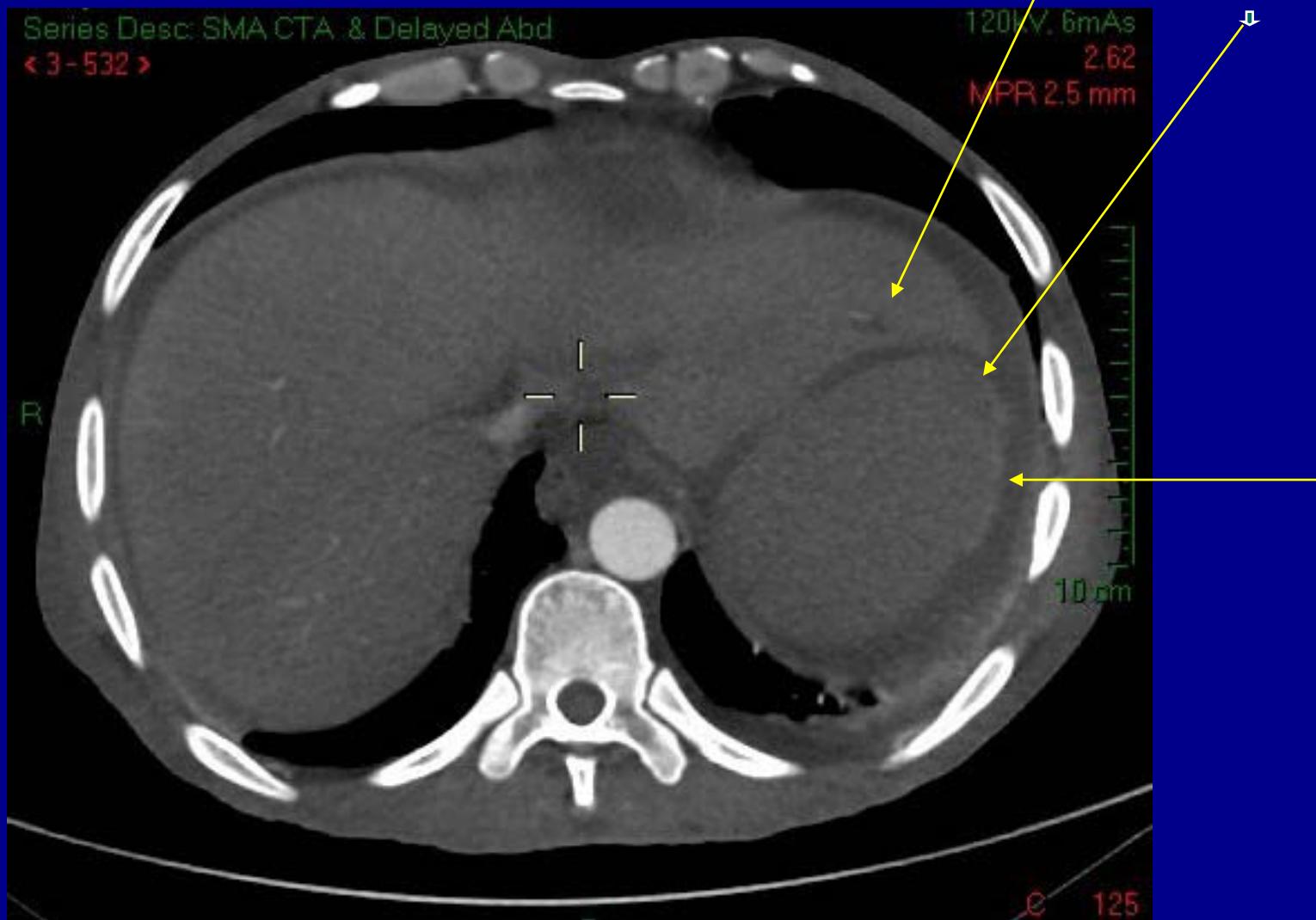
- Within 3 hour post biopsy - shock
 - Hb 3.7g% !!!



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Epilogue

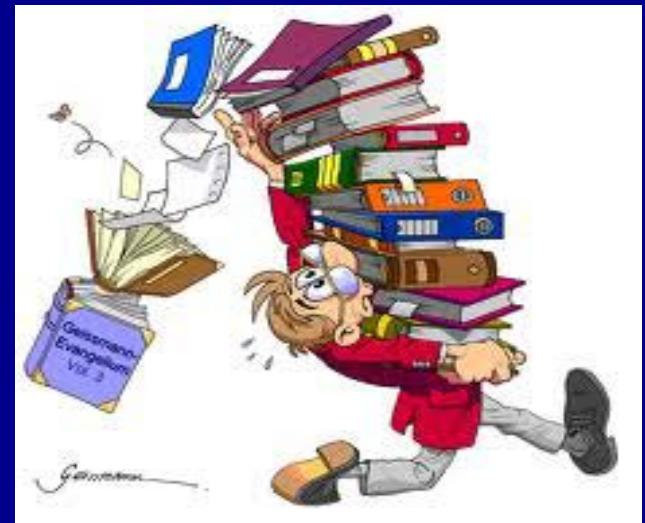
- Exitus
 - Hemorrhagic shock

Pathologic rupture of the spleen in hematologic malignancies

- Rare
- 136 cases of pathologic splenic rupture have been described:
 - 34% acute leukemias
 - 34% in non-Hodgkin's lymphomas
 - 18% in CML
- A male-to-female ratio of 3:1

Take home message

- In case of an acute liver failure
 - thorough rapid work up
 - high index of suspicion of a malignant infiltrating disease (m/p lymphoma)
- Urgent liver biopsy is essential



Thank You