GRAND ROUNDS CLINICI DEL MERCOLEDÌ
con il Policlinico San Matteo

Sistema Socio Sanitario

Regione Lombardia

Fondazione IRCCS Policlinico San Matteo

ATS Pavia

Aula Magna "C. Golgi"
& WEBINAR

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Pancreatite autoimmune: una rara causa di ittero ostruttivo

Pancreatite autoimmune: una rara causa di ittero ostruttivo

- Male, 60 y.o., caucasian
- APR: splenectomy during treatment of Hodgkin's disease (1984-85), spine arthrodesis for dorsal kyphosis (2020), arterial hypertension
- Therapy: nebivolol, olmesartan

He referred to our attention for **jaundice with severe weight loss** (- 8 kg) in the last two months. **No pain.**

Laboratory tests:
bilirubin T/D 10.85/8.23 mg/dL
GGT 723 mU/mL, ALP 385 UI/mL
AST/ALT 261/186 mU/mL
amylase 149 mU/L, lipase 249 mU/L
Normal values of CEA, CA 19,9, AFP, viral hepatits markers

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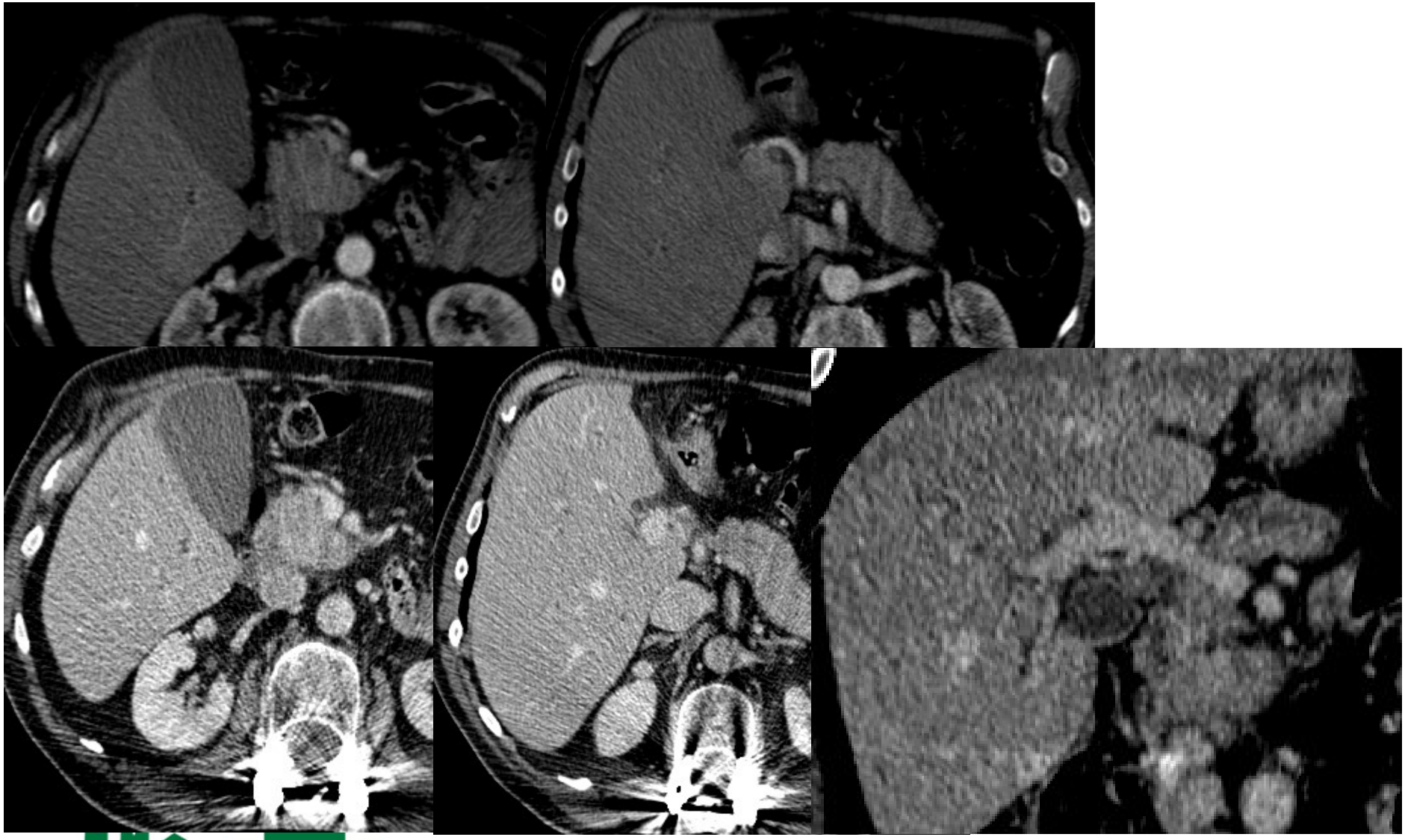

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FEATURES:
No mass in the pancreatic head and normal vascularization of the gland
No dilatation of the MPD
Hydrops of the gadbladder and dilation of the BD
The intrapancreatic BD is thin with minimal parietal enhancement
No biliary stones
No free fluid

RADIOLOGICAL CONCLUSION:
- exclusion of primary pancreatic lesion
- exclusion of choledocholithiasis
- not exclusion of intrapancreatic BD pathology

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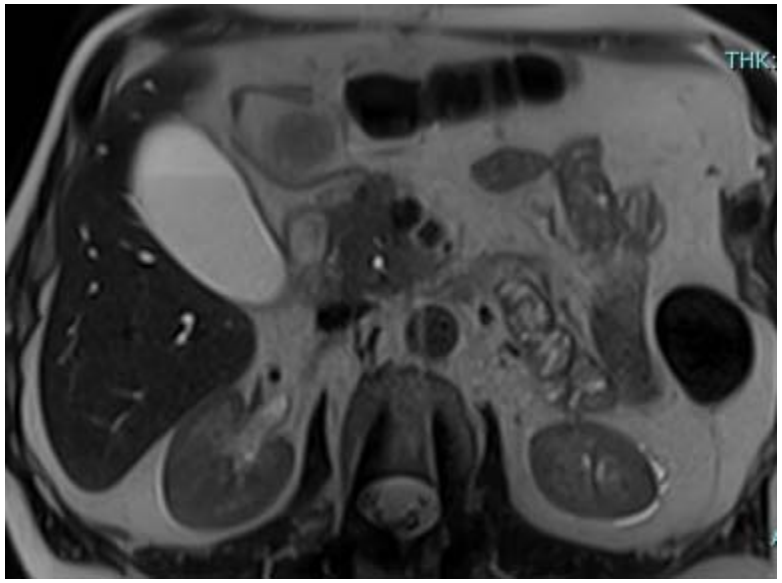

- confirmation of choledochal stop, without stones, with upstream dilation
- the MPD is visible in the pancreatic head

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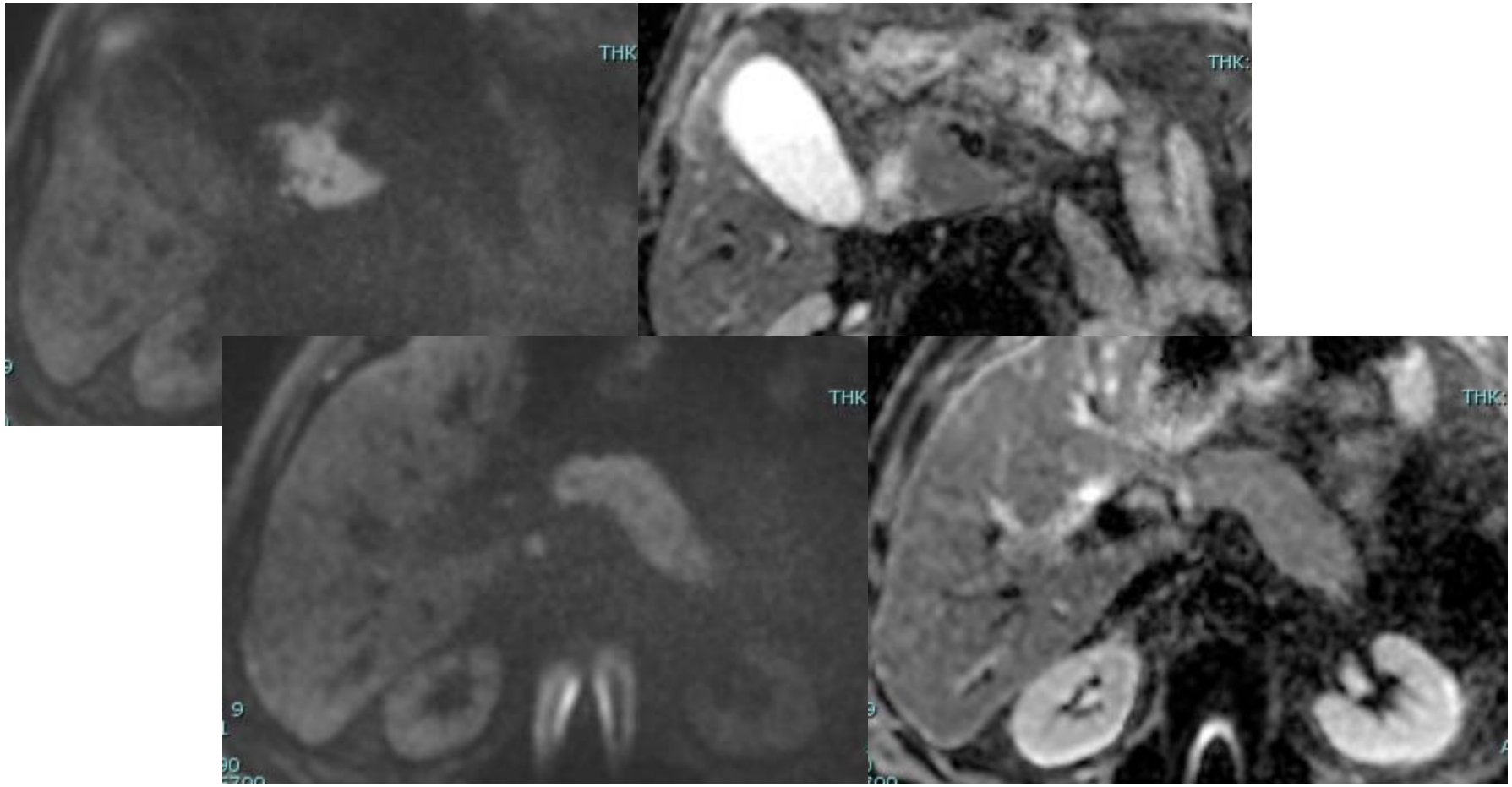

- slightly hyperintensity in DWI and hypointensity in ADC in the pancreatic head compared to the body-tail
- focal peri-ductal hyperintensity in ADC

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RADIOLOGICAL CONCLUSION:
- confirmation of exclusion of primary pancreatic lesion
- confirmation of exclusion of choledocholithiasis
- confirmation of potential intrapancreatic BD pathology due to the stop, without pathological restricted signal in ADC

EUS

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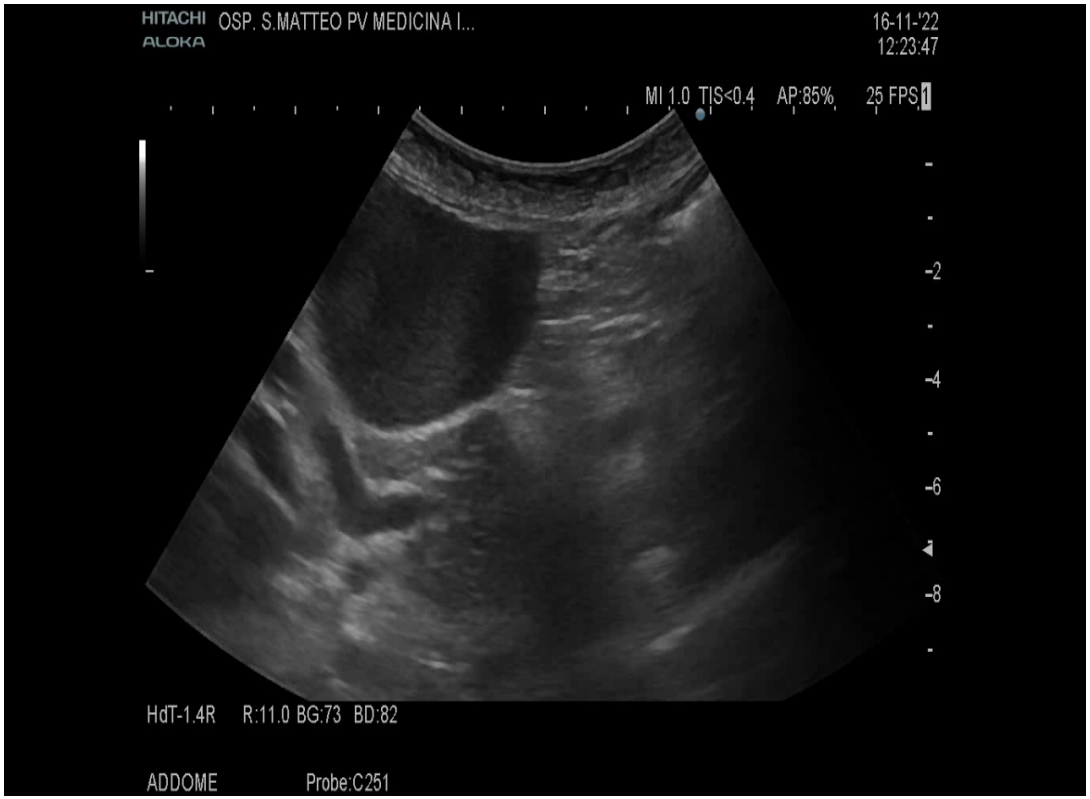
CE-US (SonoVue®, Bracco Imaging)



- Diffuse enlargement of the pancreas with a more hypoechoic area in the head
- Main pancreatic duct (PD) duct narrowing without upstream dilation
- CEUS: omogenous and vivid c.e. of all the gland without any parenchymal wash-out in late phase

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Various hypothesis evaluating the CE-US

NET ?

Mass mimicking AIP?

Ductal adenocarcinoma?

Cholangiocarcinoma of the distal CBD?

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IgG4 4783 mg/L (n.v. 39-864 mg/L)
Chromogranin A 58 (n.v. 19-98 ng/mL)

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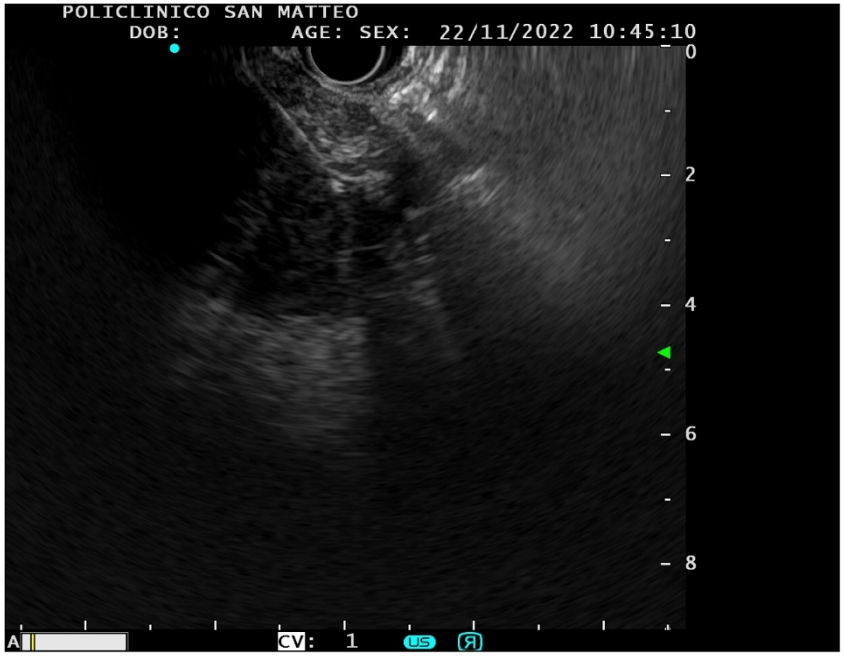
EUS



- Diffuse ‘sausage-like’ shape of pancreas with mass-mimicking hypoechoic area of the head
- No dilation of the PD
- CBD dilation with thickened walls
- Hard parenchyma in elastosonography (not typical !!!!)
- CE-EUS: similiar findings as CE-US

Contestual procedure of:

- FNB-EUS guided for istological diagnosis
- ERCP and stenting of the CBD

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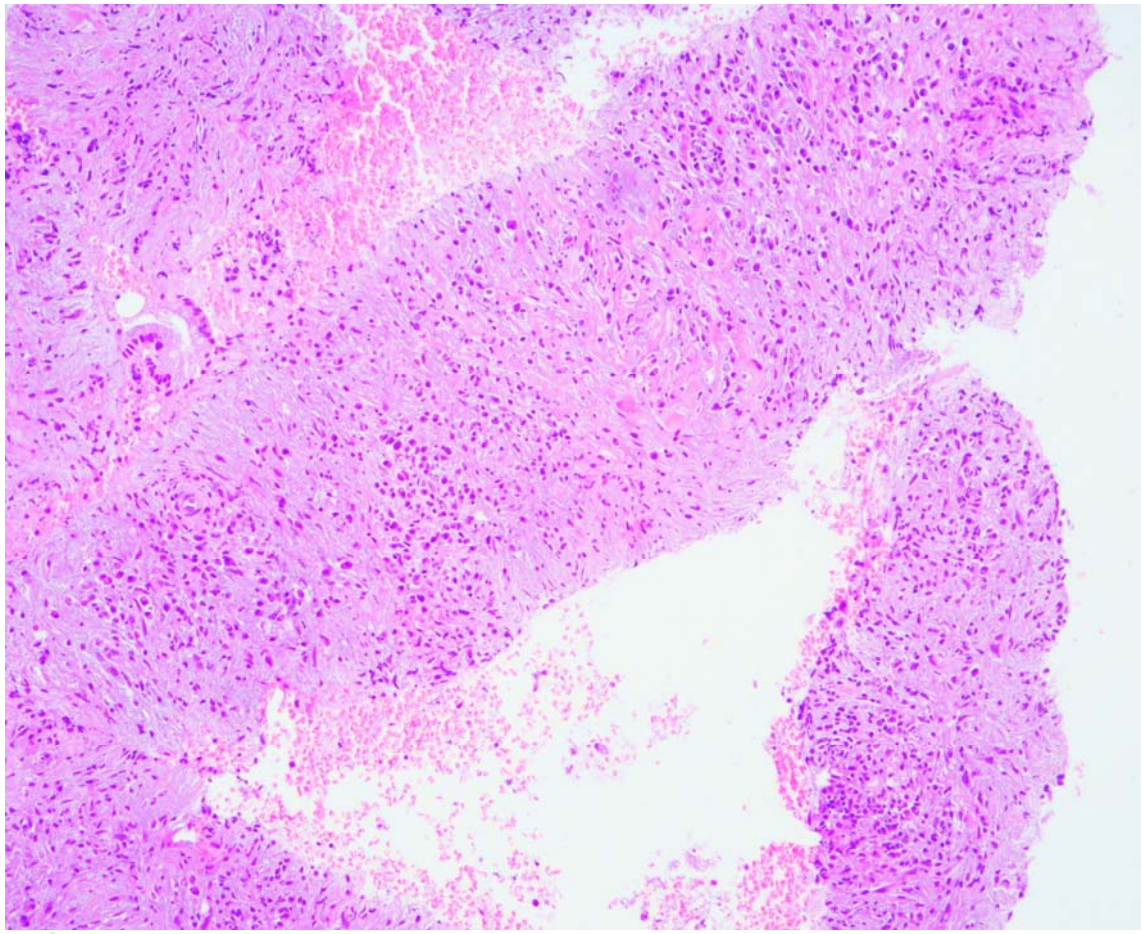

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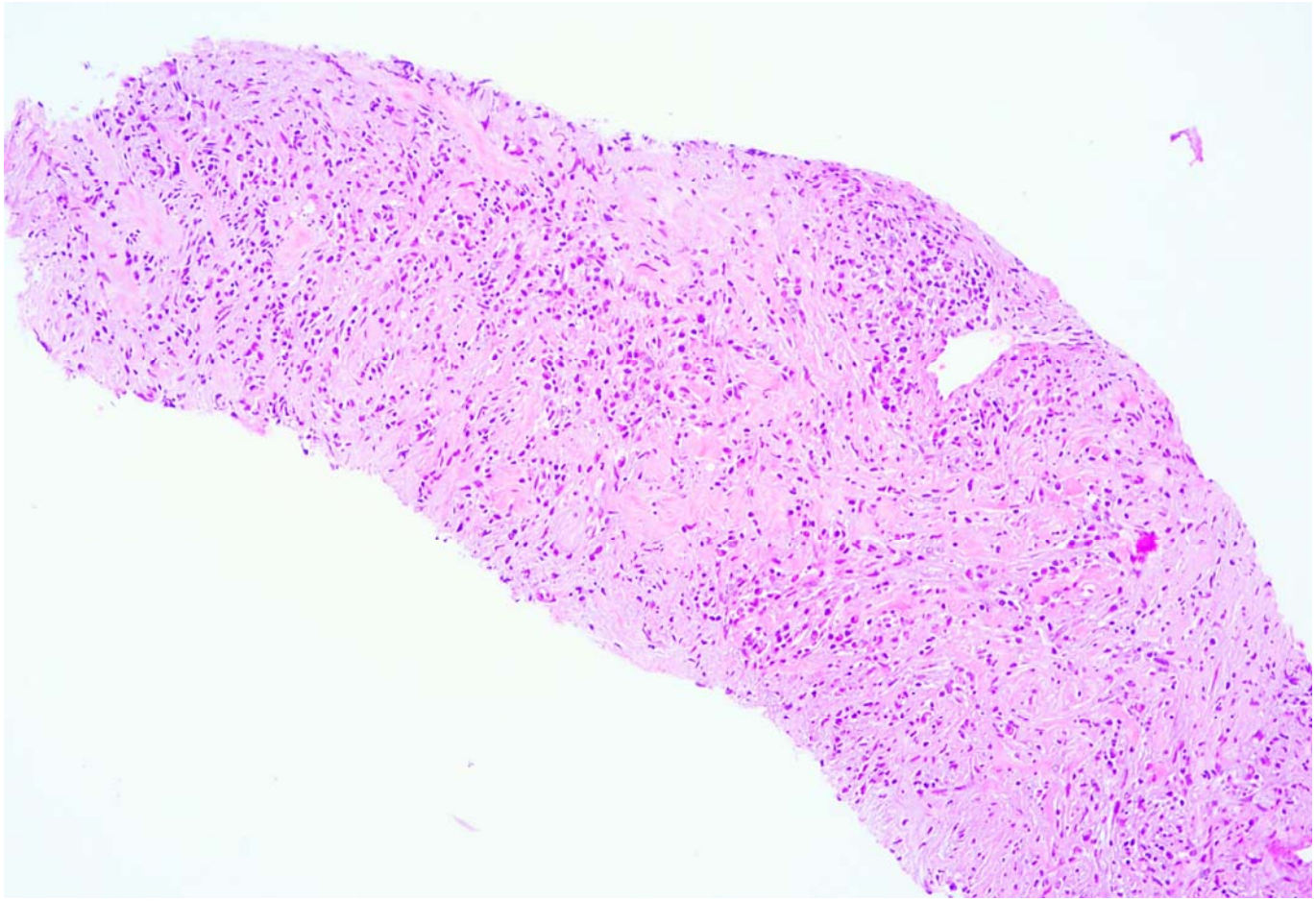

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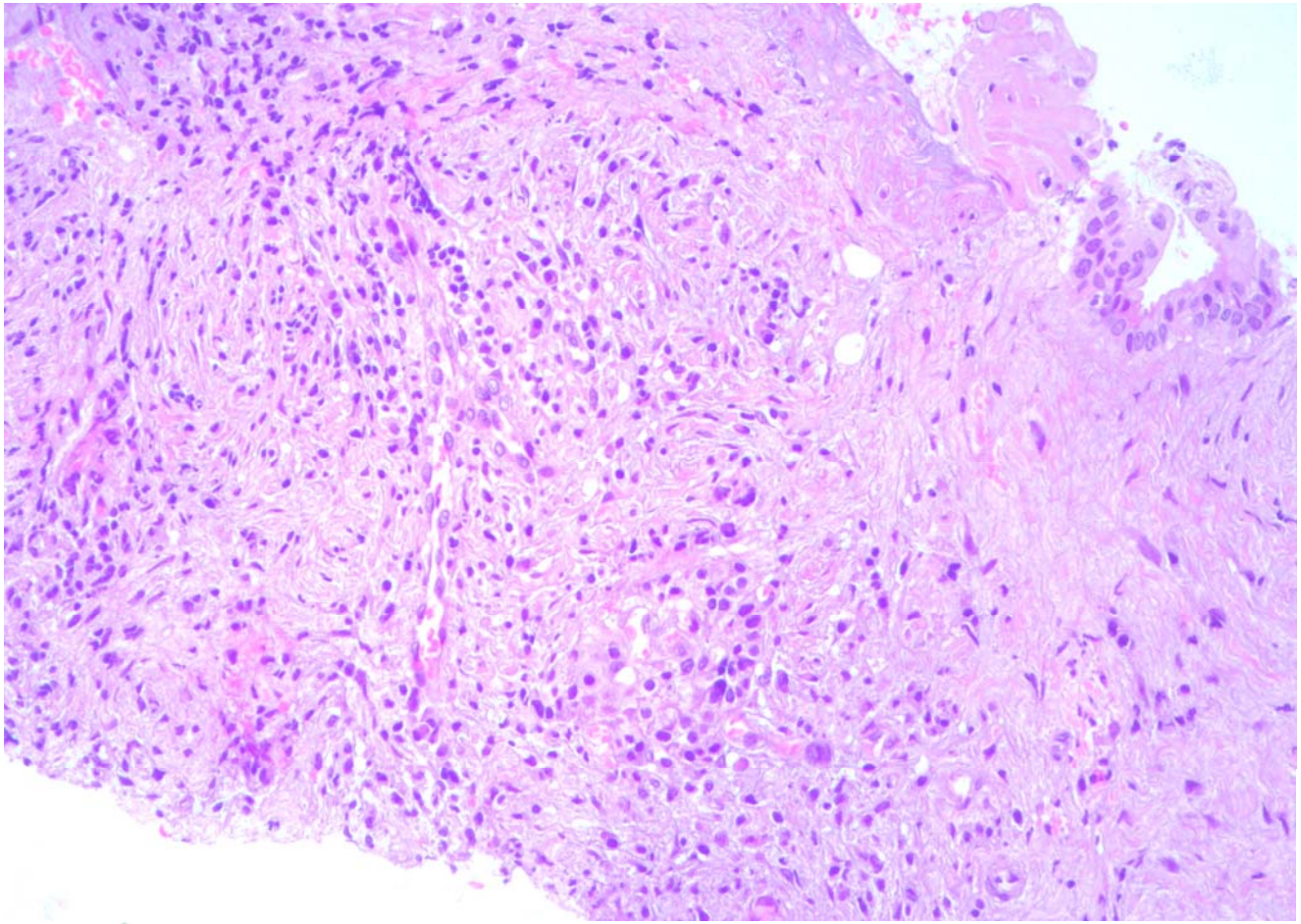

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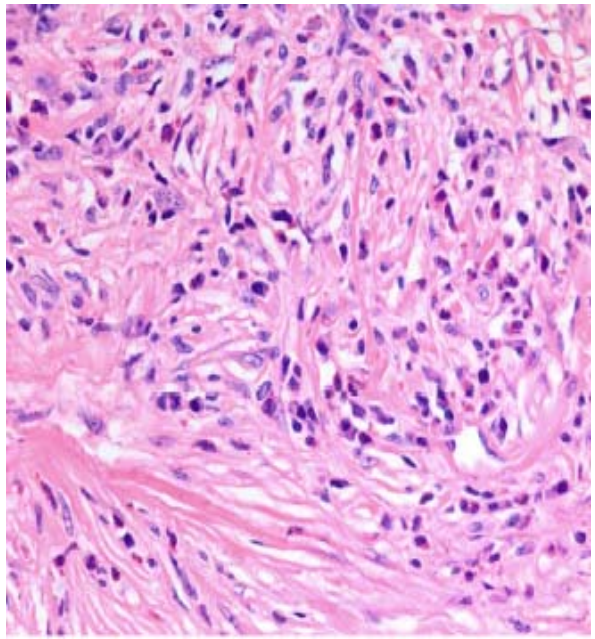
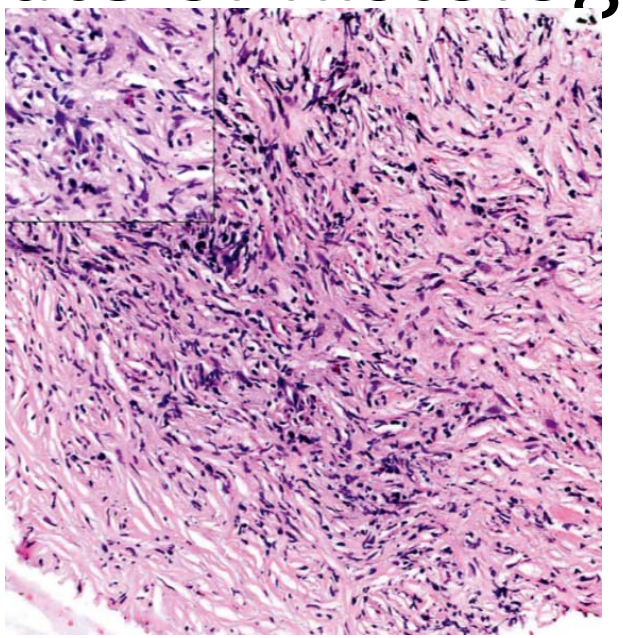
Pancreatite autoimmune: una rara causa di ittero ostruttivo

TYPE 1 AUTOIMMUNE PANCREATITIS

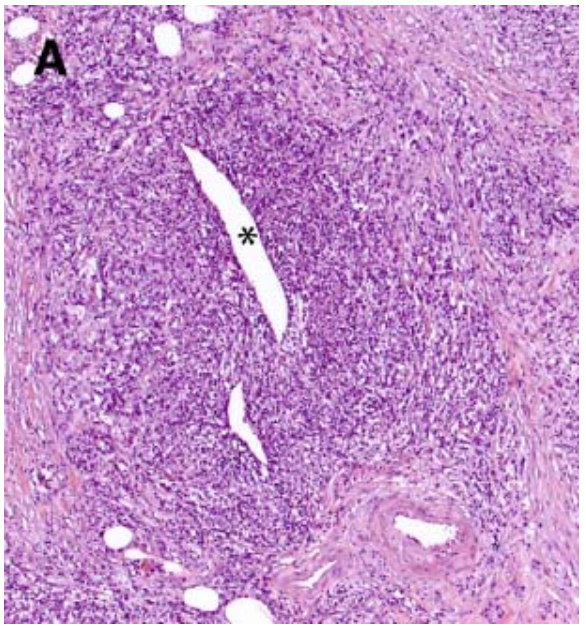
a triumvirate of histologic features



1) **Dense lymphoplasmacytic infiltrate**



2) **storiform-type fibrosis (short intersecting fascicles)**



3) **obliterative phlebitis**

TYPE 1 AUTOIMMUNE PANCREATITIS

- Virtually every case of type 1 AIP shows a markedly elevated number of IgG4+ plasma cells
- A minimum requirement of **50 IgG4+ plasma cells per HPF on a resection specimen** and **10/HPF on a biopsy sample.**

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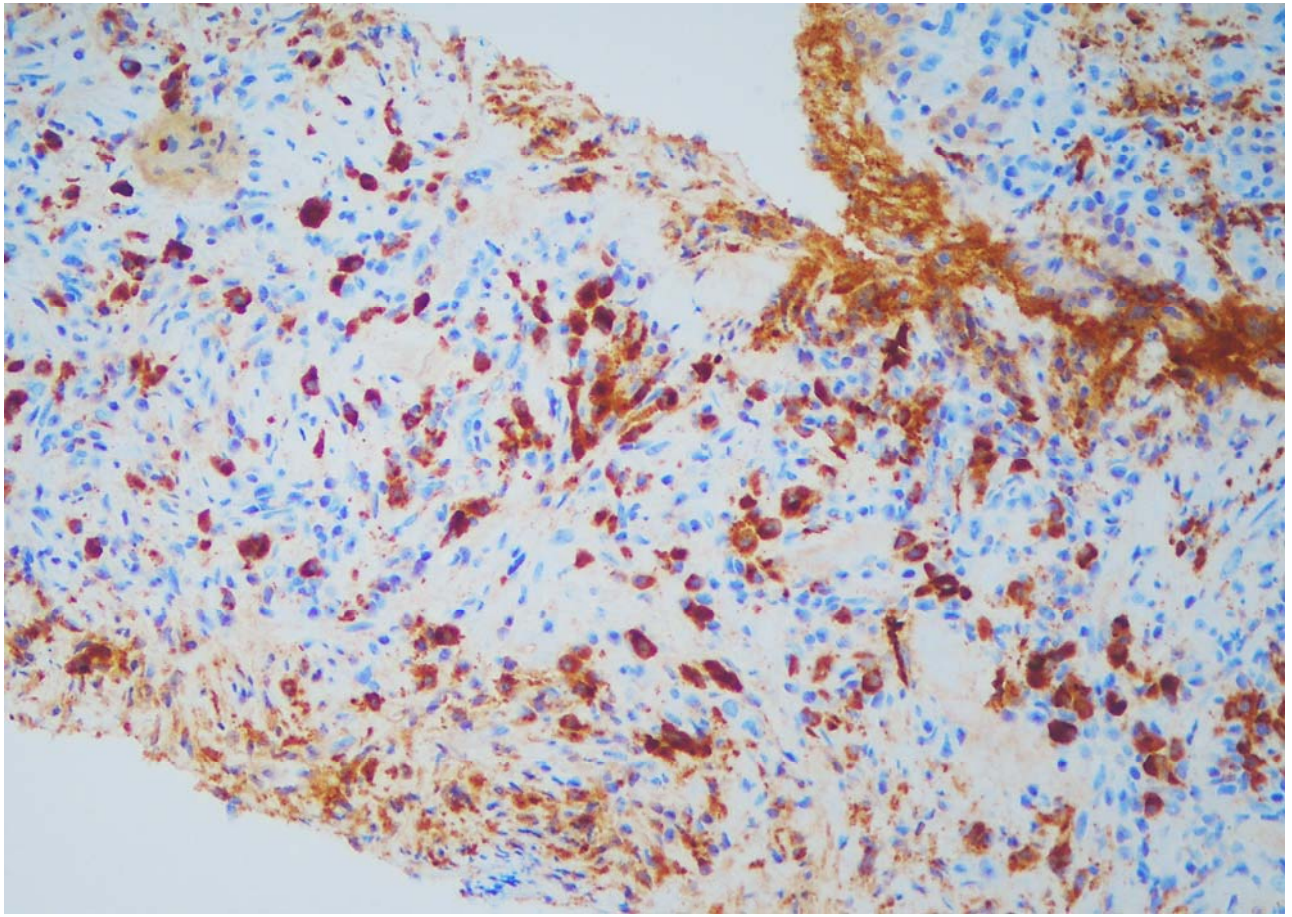

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	Type 1 AIP	Type 2 AIP
Age	Elderly, seventh decade of life	Middle age, fifth decade of life
Sex	Predominantly male	Males and females equally affected
Presentation	Jaundice (75%), acute pancreatitis (15%)	Jaundice (~50%), acute pancreatitis (~33%)
Systemic disease	Yes	No
Elevated serum immunoglobulin G4	80%	Uncommon
Inflammatory bowel disease	No association	Present in 16-30% of cases
Histopathology	Periductal inflammation with one or more of the following features: 1. Storiform fibrosis 2. Obliterative phlebitis	Periductal inflammation with one or more of the following features: 1. Ductal/lobular abscesses 2. Ductal ulceration with neutrophils
Long-term outcome	Frequent relapses	No relapses

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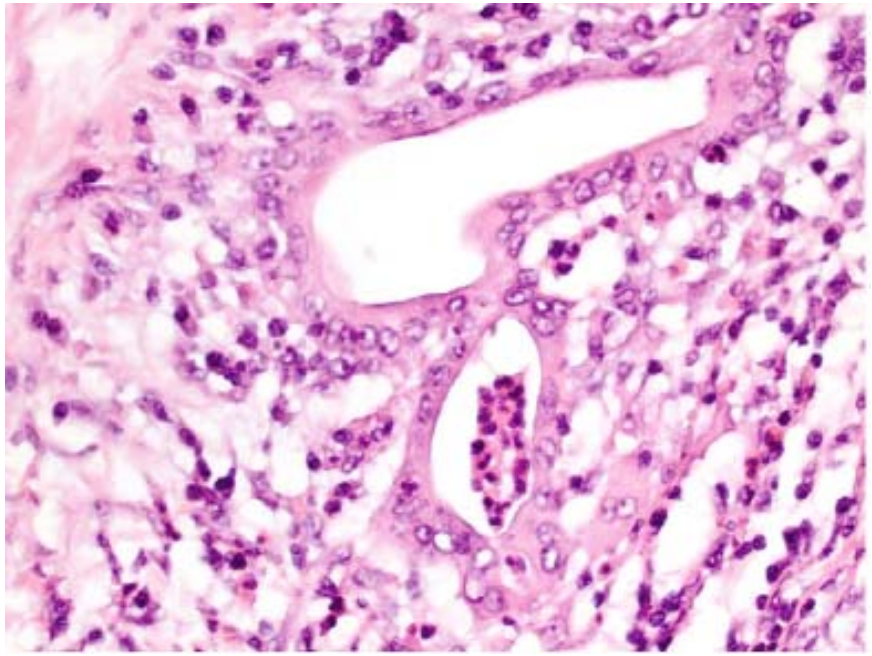
TYPE 2 AUTOIMMUNE PANCREATITIS

The most distinctive feature of this variant is a dense **periductal collar of lymphocytes** and plasma cells, accompanied by **neutrophilic microabscesses within the lumen of the duct**: the so-called **granulocytic epithelial lesion**



Erosion and ulceration of the duct lining are frequently seen, occasionally accompanied by complete destruction of the duct.

Storiform-type fibrosis is typically not prominent.

Obliterative phlebitis is uncommon.

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Following guidelines the patient was treated with biliary drainage and high dose steroids for 6 weeks

“Prednisone with the initial dose of 0.6–1.0 mg/kg/day should be started. ” (level A)

Okazaki K et al. International consensus for the treatment of autoimmune pancreatitis. Pancreatology. 2017 Jan-Feb;17(1):1-6. doi: 10.1016/j.pan.2016.12.003 PMID: 28027896.

IgG4 976 mg/L (n.v. 39-864 mg/L)
Bilirubin T 0,49 mg/dL, GGT 28 mU/mL, ALP 30 UI/mL

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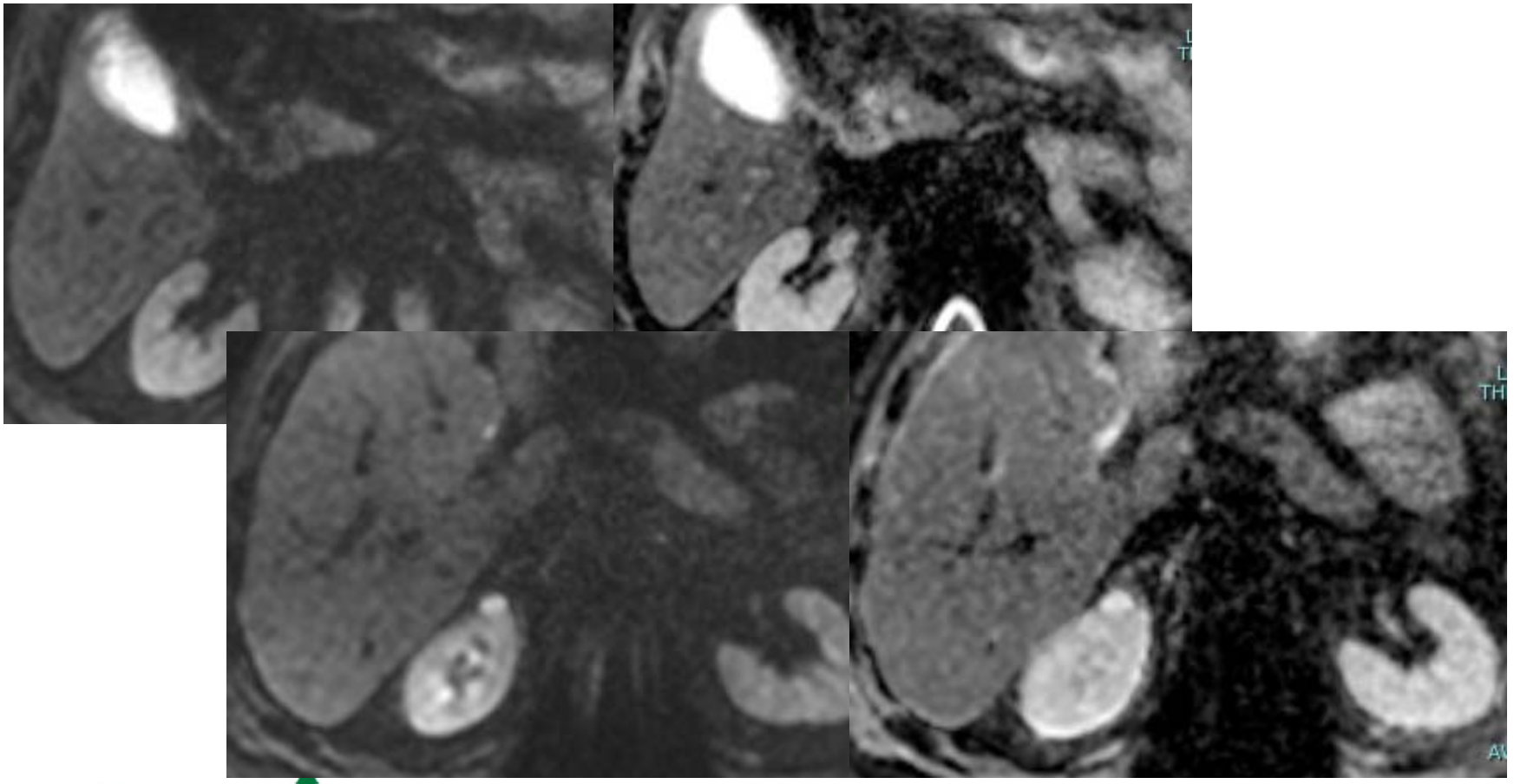
RM POST-THERAPY



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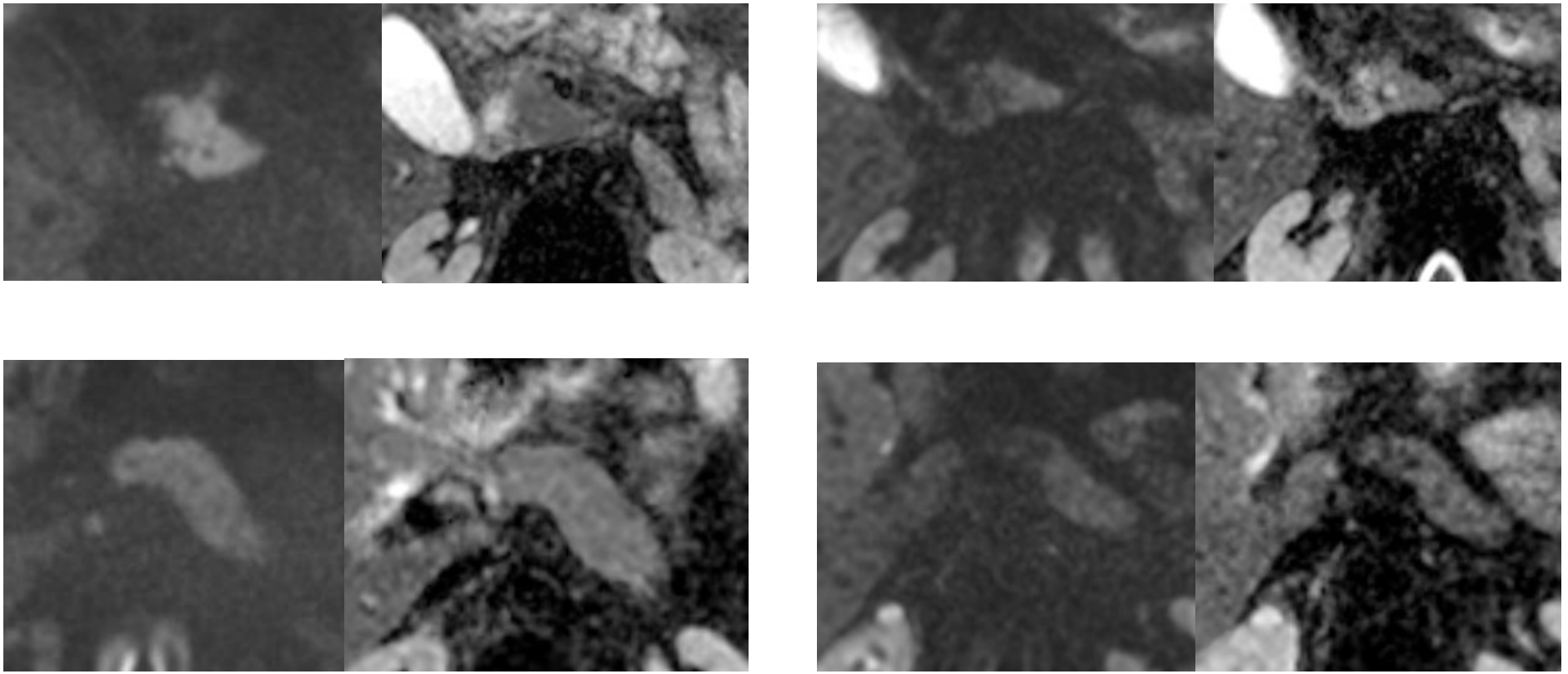
PRE

POST



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RADIOLOGICAL CONCLUSION:

TYPICAL FOCAL PAI
- **isovascular** in all dynamic phases (NB: if late diagnosis, a slight hypovascularization is possible)
- **MPD not dilated and not visible**
- **potential dilatation of the CBD** in PAI of the pancreatic head

DIFFERENTIAL DIAGNOSIS:
1) DUCTAL ADENOCARCINOMA OF THE PANCREAS
- hypovascular mass
- typically MPD upstream dilation with “double duct sign” in head pancreatic mass

2) CBD CHOLANGIOCARCINOMA
- parietal enhancement of the CBD, with stop like “coda di topo”
- upstream dilation of the BD

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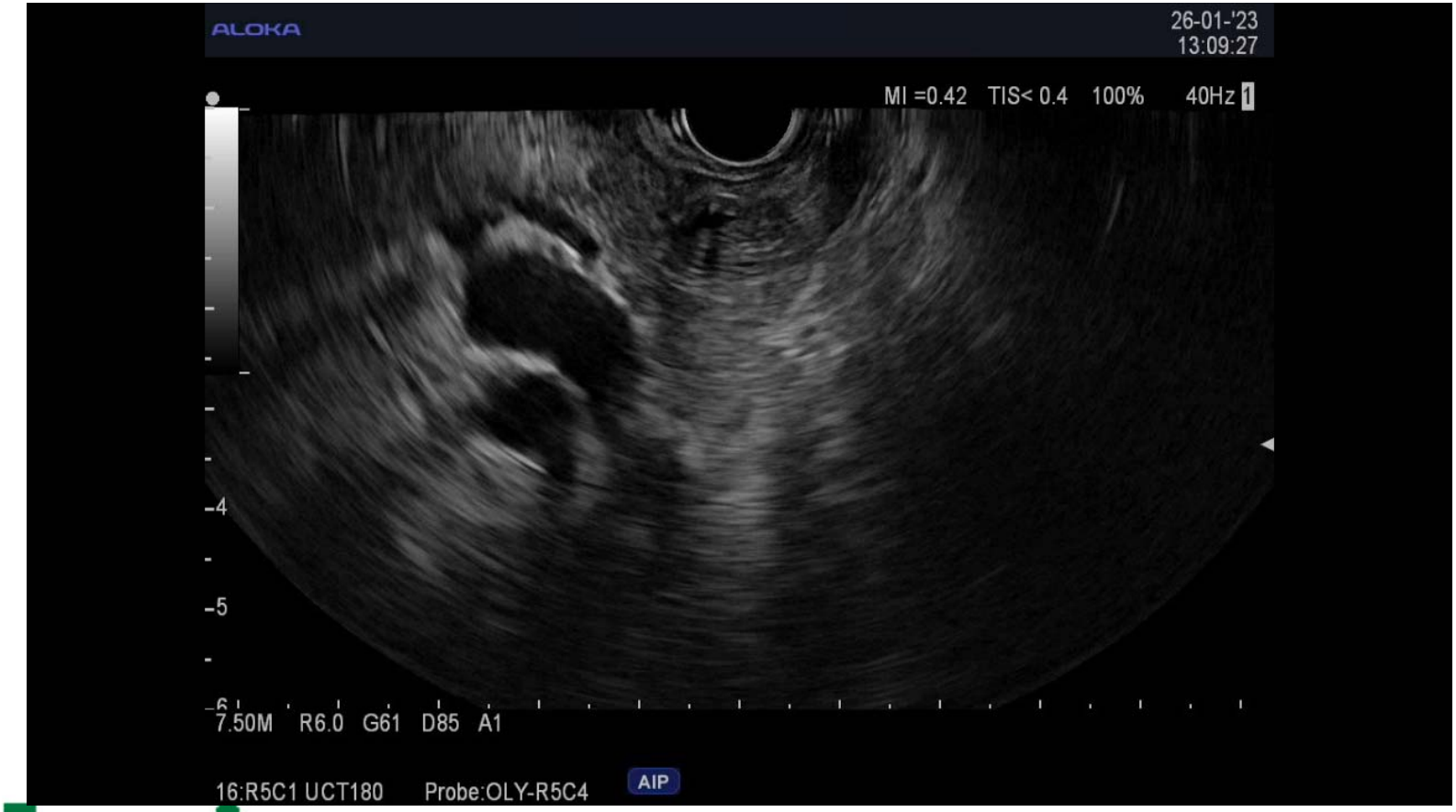
Post teraphy CE-EUS



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Take Home Messages

- Consider AIP when approching a patient with obstructive jaundice of not clear diagnosis
- Clinical, radiological and serological features often allow the achievement of a definitive diagnosis of AIP
- If in doubt, tissue sampling (FNB EUS-guided) is very accurate to differentiate between AIP and cancer
- CE-US and CE-EUS are very useful tools during differential diagnosis and imaging follow-up

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Grazie per l'attenzione!