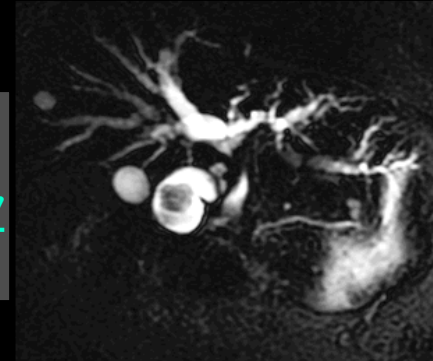


Bilio-pancreatic "tumor-like" lesions



1. Fibrous sclerosing (stenosing) cholangitis (cholangiopathies) and hepato-biliary inflammatory pseudo-tumors

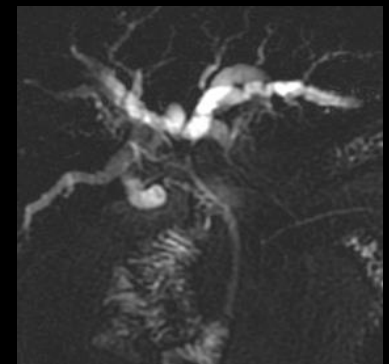
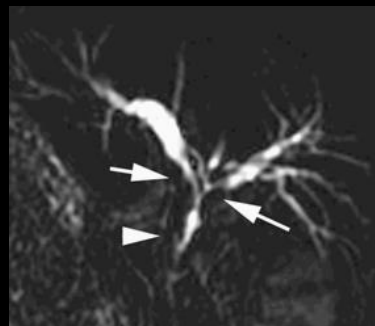


2. Autoimmune pancreatitis and pancreatic inflammatory pseudotumors



Only 2 among these 5 patients with biological cholestasis \pm icterus have a malignant lesion of the biliary tract and should be operated. Can you identify them ?

5 MRCP ; only 2 malignant lesions !!!



1a-Sclerosing (fibrous obliterating) cholangitis (cholangiopathies)

-cholangiocarcinoma (biliary adenocarcinoma)
accounts for **15 % of hepatobiliary cancer**

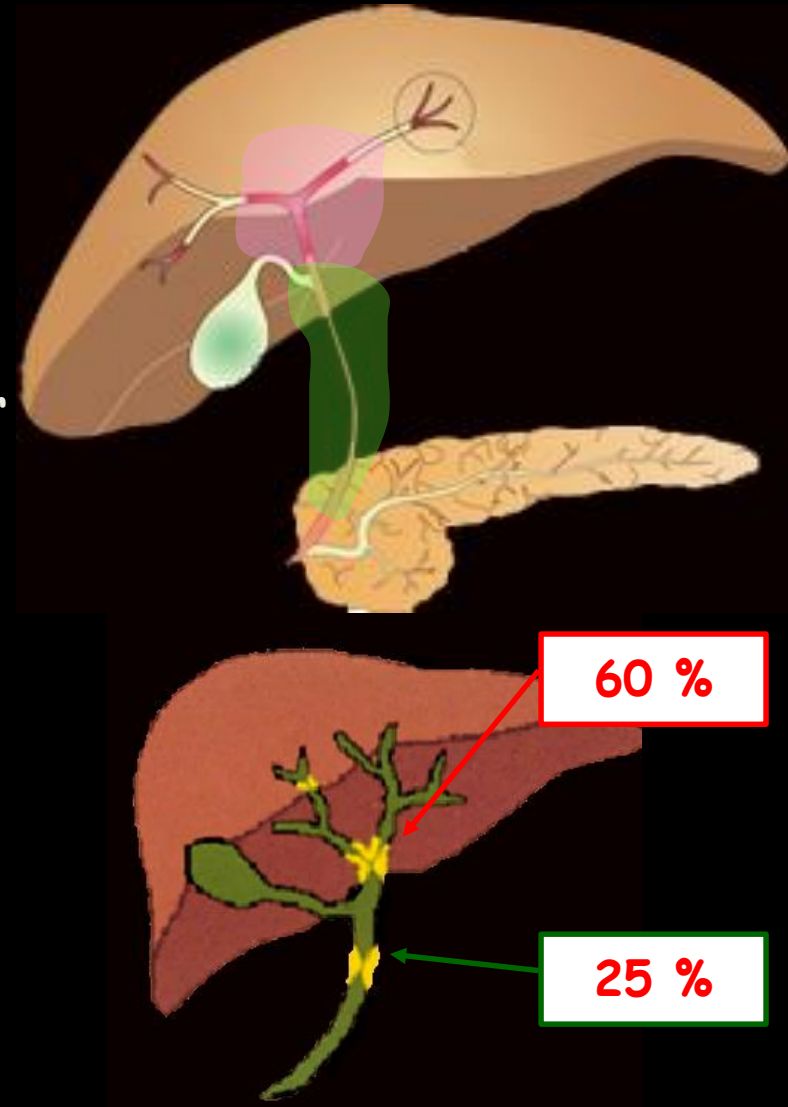
-60 % of extrahepatic cholangiocarcinoma involve
hilar structures (**Klatskin's tumor**) ; 25 % are
main bile duct cholangiocarcinoma

-recent surgical studies have shown that
approximately **10 to 24 %** of patient operated for
biliary obstruction presumed to be hilar or main
bile duct malignant lesion are ultimately proved to
have idiopathic benign stricture on final histologic
review

*Corvera CU et al Clinical and pathologic features of proximal biliary
stricture masquerading as hilar cholangiocarcinoma J Am Coll Surg
2005;201:862-9*

*Erdogan D et al Immunoglobulin G4 related sclerosing cholangitis in
patients resected for presumed malignant bile duct strictures Brit J Surg
2008;95:727-34*

*Clayton RA et al Incidence of benign pathology in patients undergoing
hepatic resection for suspected malignancy Surgeon 2003; 1:32-38*

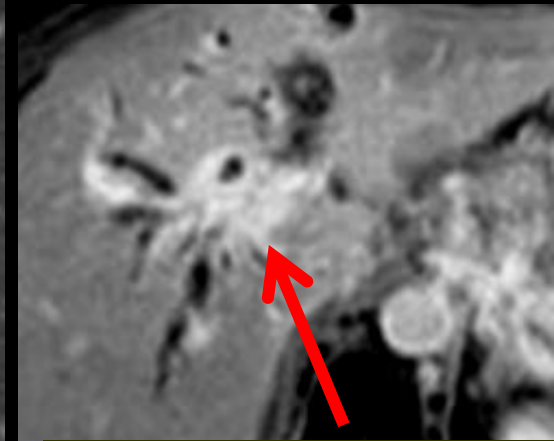


Klatskin's tumors ;classical presentations

mass forming hilar cholangiocarcinoma



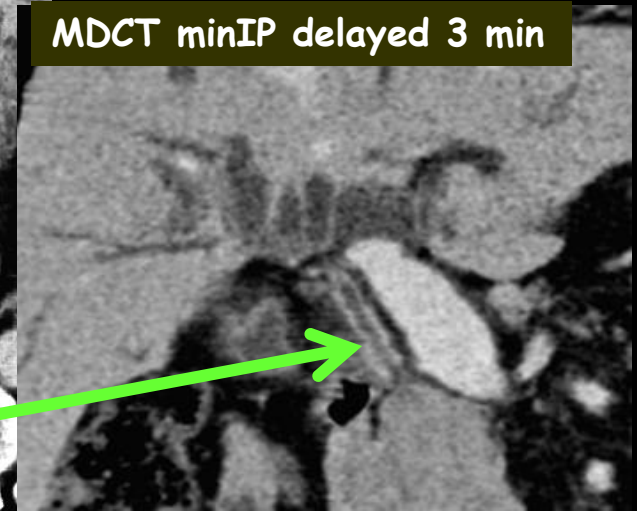
MRCP



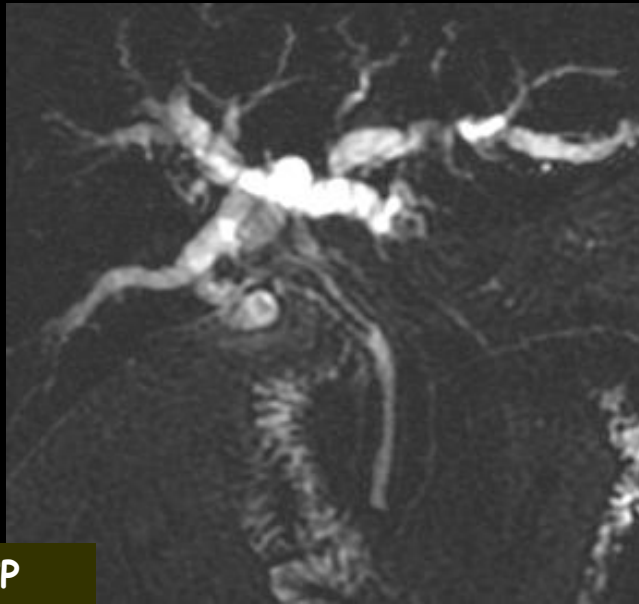
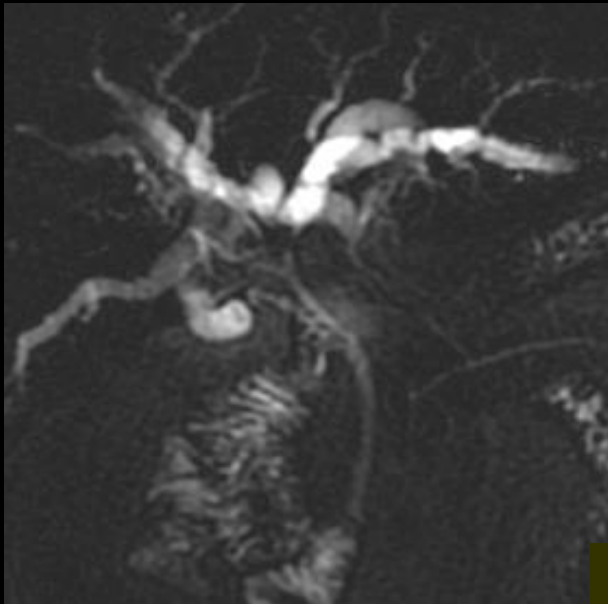
T1W gado Fat sat
delayed : 3 to 5 minutes



MDCT minIP delayed 3 min



infiltrating hilar and main bile duct cholangiocarcinoma



infiltrating
cholangiocarcinoma
of the hepatic duct
with hilar
involvement

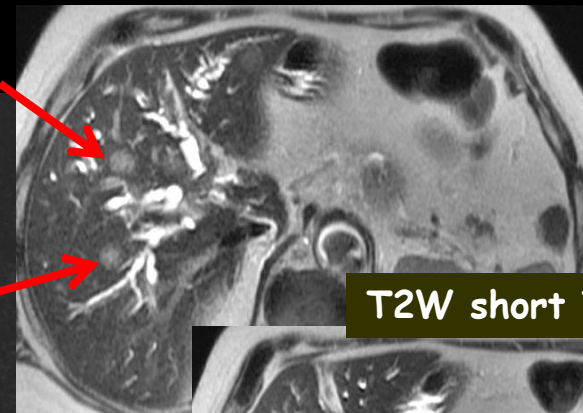
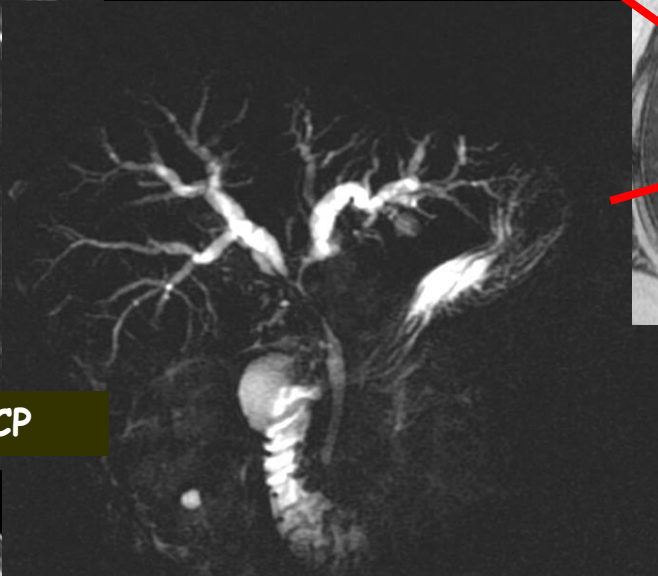
MRCP



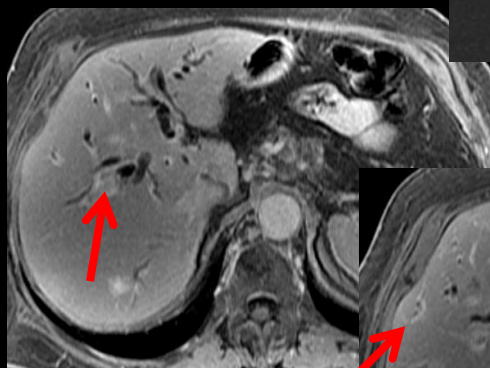
T1W gado Fat sat; delayed acquisition



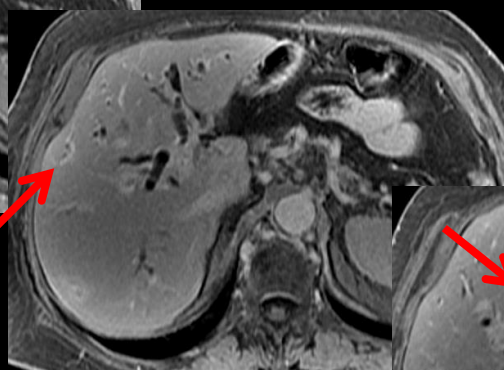
MRCP



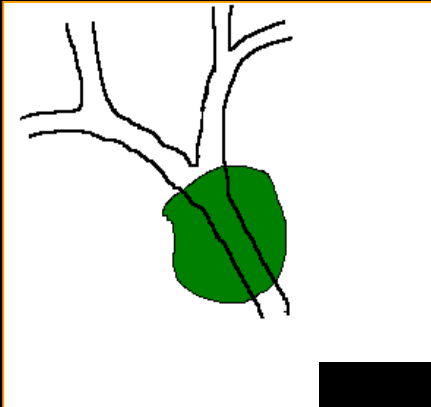
T2W short TE eff



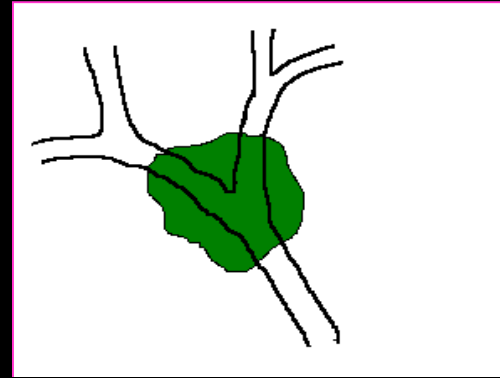
T1W gado FatSat



infiltrating Klatskin's tumor with liver metastases 3T MRI



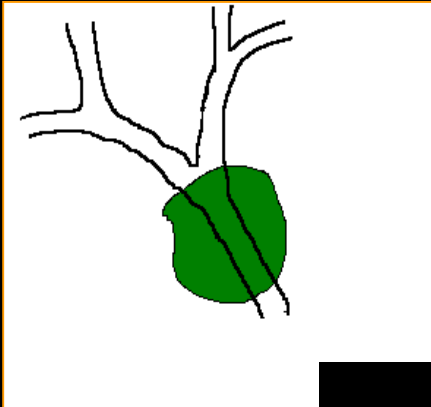
TYPE I



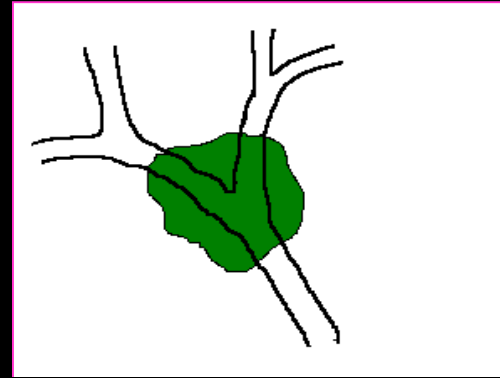
TYPE II

O.Durieux,S.Agostini
Marseille

Bismuth Cornette I or II : main bile or proximal biliary duct resection + biliary intestinal anastomose (Roux en Y hepatico jejunostomy)



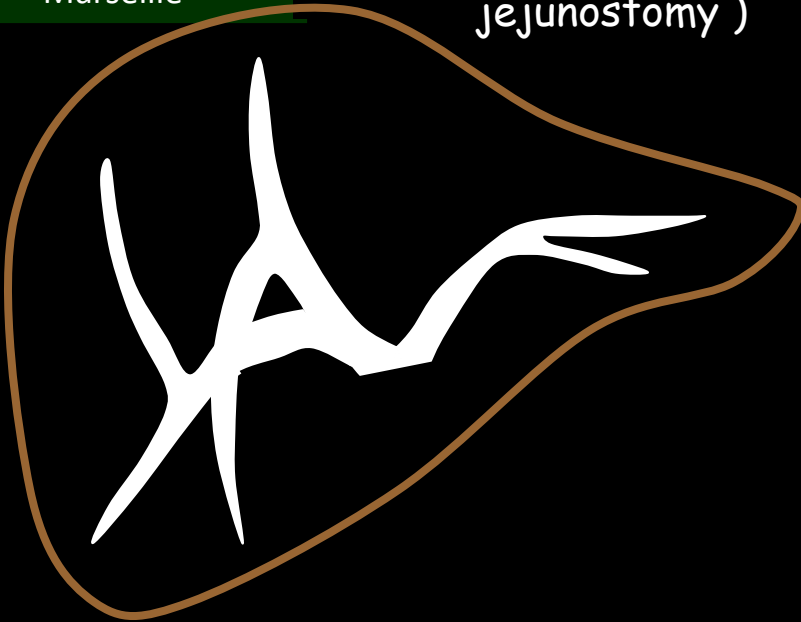
TYPE I

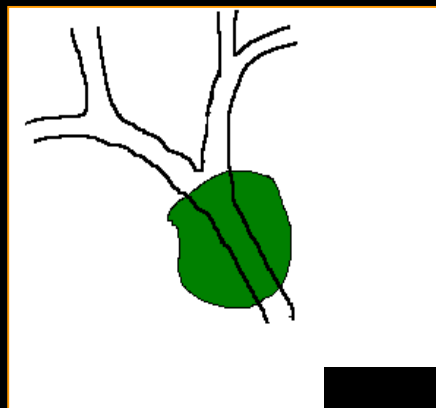


TYPE II

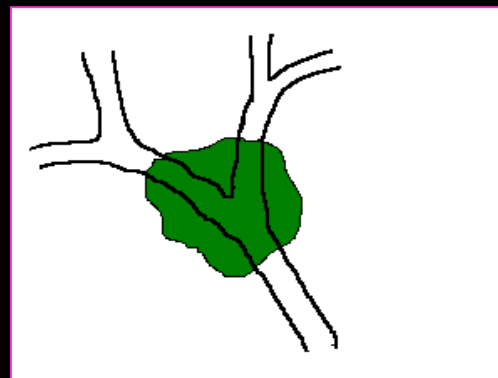
O.Durieux,S.Agostini
Marseille

Bismuth Cornette I or II : main bile or proximal biliary duct resection + biliary intestinal anastomose (Roux en Y hepatico jejunostomy)





TYPE I



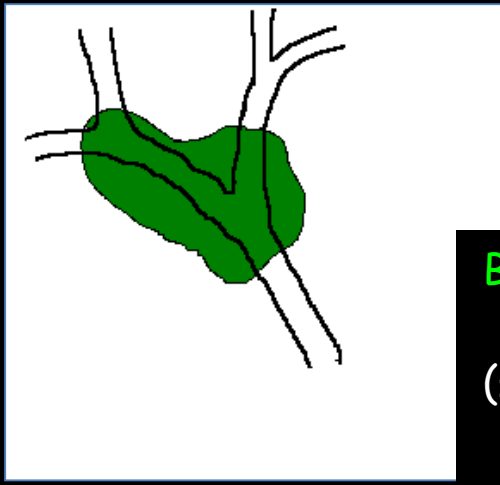
TYPE II

O.Durieux,S.Agostini
Marseille

Bismuth Cornette I or II : main bile or proximal biliary duct resection + biliary intestinal anastomose (Roux en Y hepatico jejunostomy)

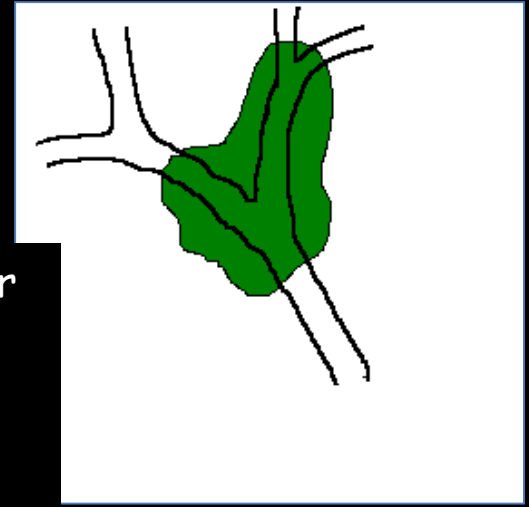


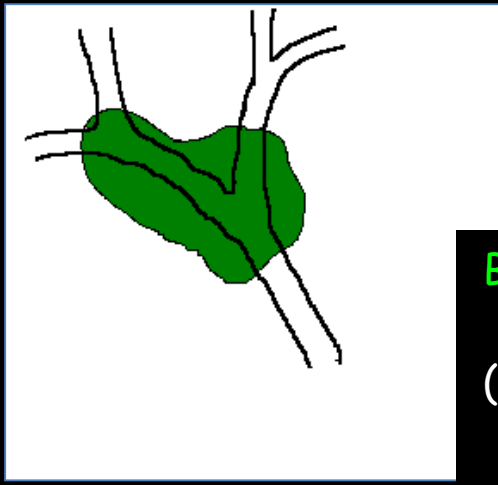
PJ Valette Lyon



TYPE III droit et III gauche

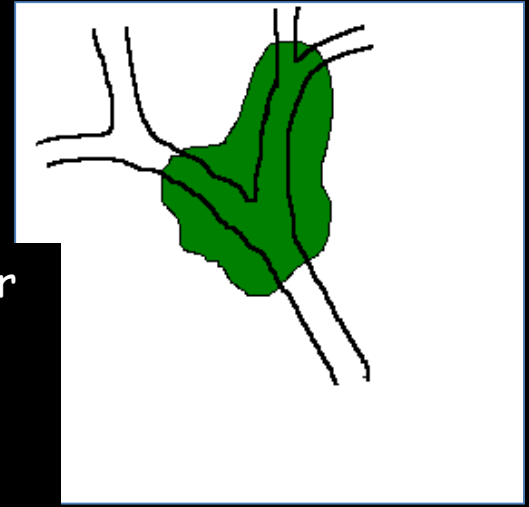
Bismuth-Cornette III : extended right or left hepatectomy
(± previous right lobe portal embolisation for extended right hepatectomy)

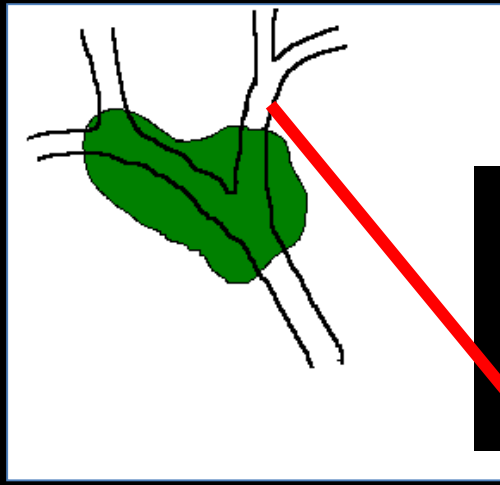




TYPE III droit et III gauche

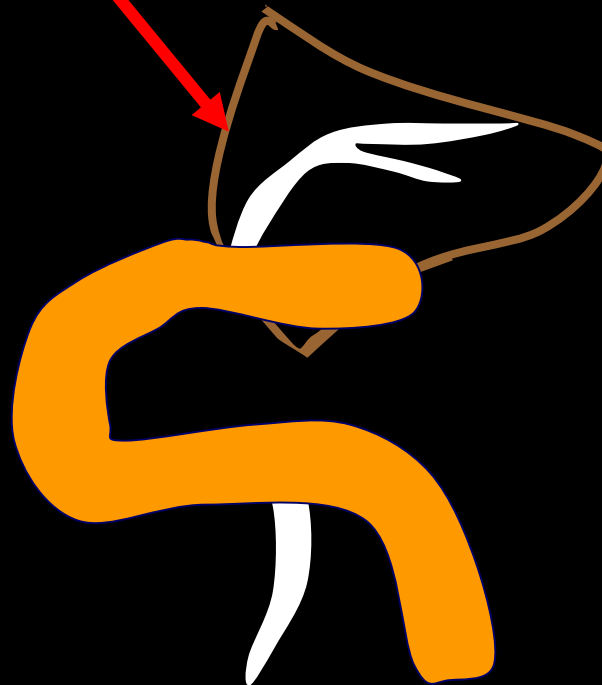
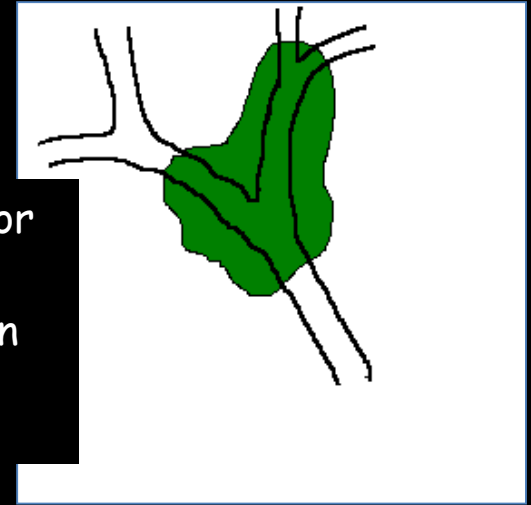
Bismuth-Cornette III : extended right or
left hepatectomy
(± previous right lobe portal embolisation
for extended right hepatectomy)

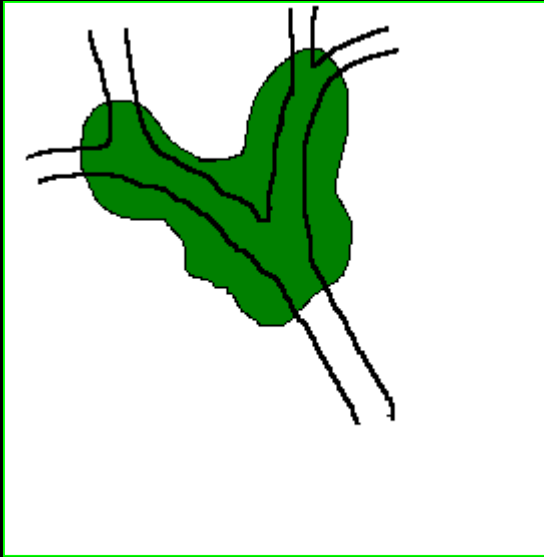




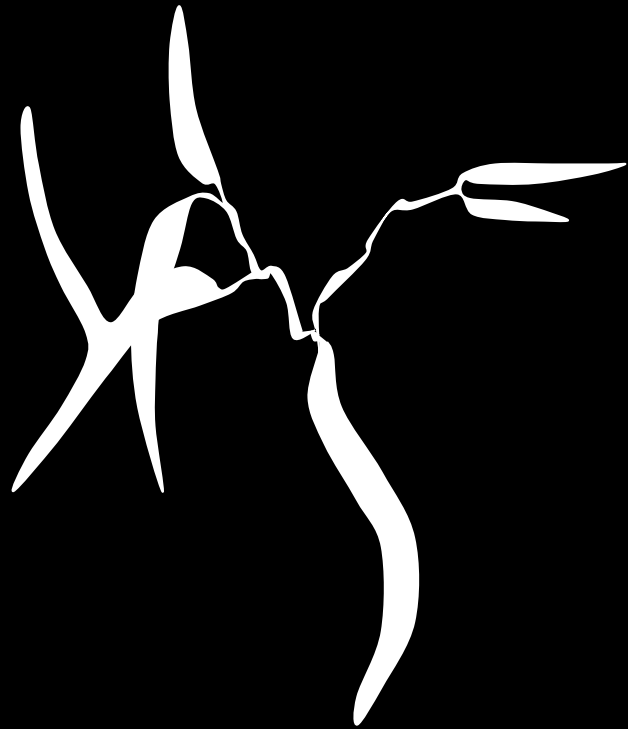
TYPE III droit ou III gauche

Bismuth-Cornette III : extended right or
left hepatectomy
(± previous right lobe portal embolisation
for extended right hepatectomy)





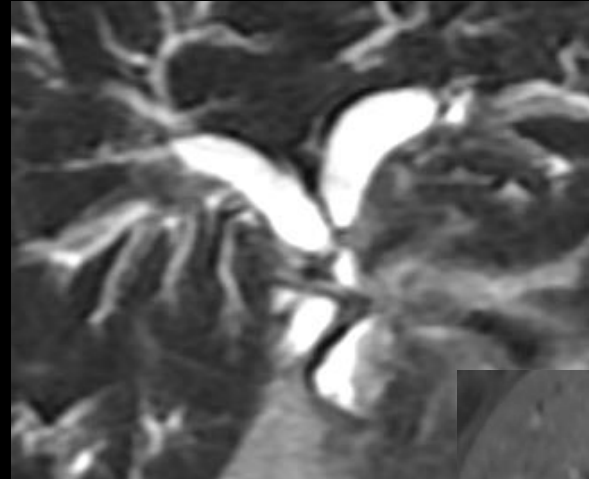
TYPE IV



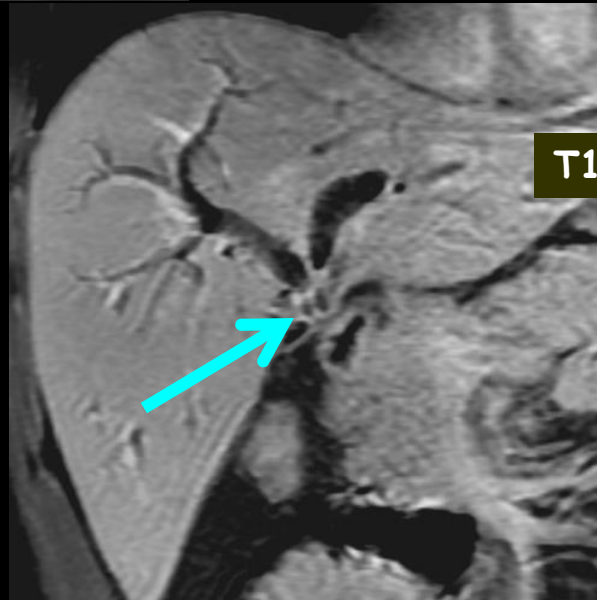
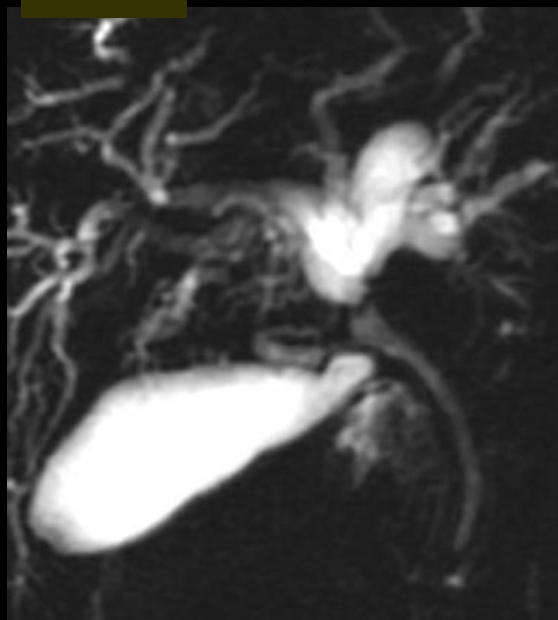
Bismuth-Cornette IV : no resection surgery ; palliative biliary diversion :
endoprosthesis with preferred percutaneously transhepatic approach

33 y.o man , subicterus for ten days , intense asthenia ;anorexia .

Mild and fluctuating biological cholestasis (total serum bilirubin between 50 and 80 mg/L)



SSFSE short eff TE

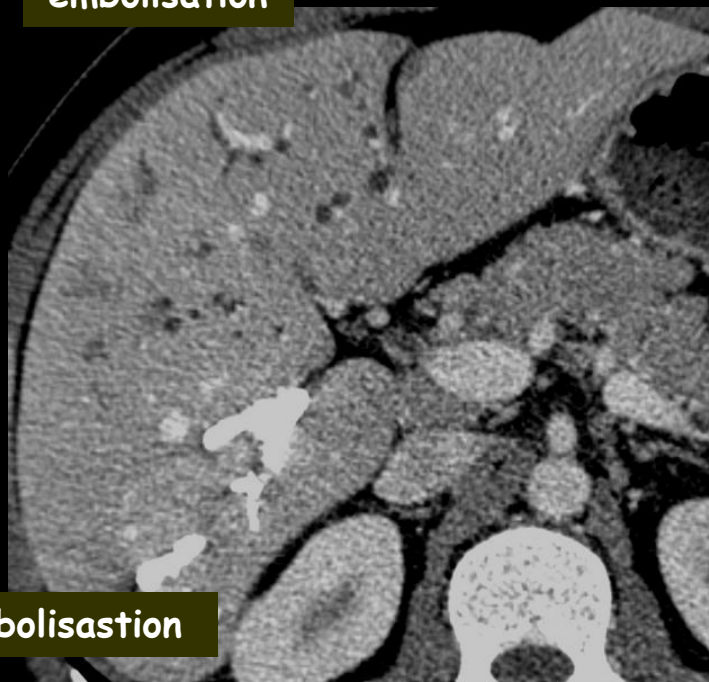


T1W gado FS

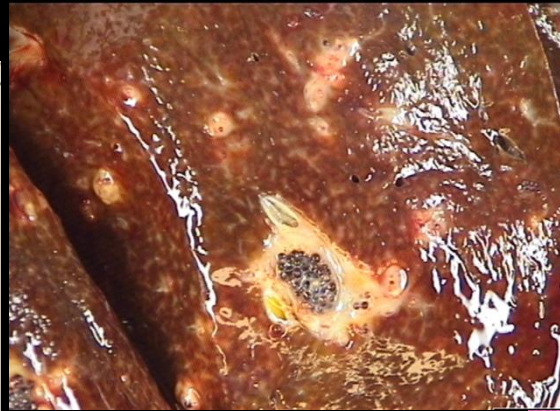
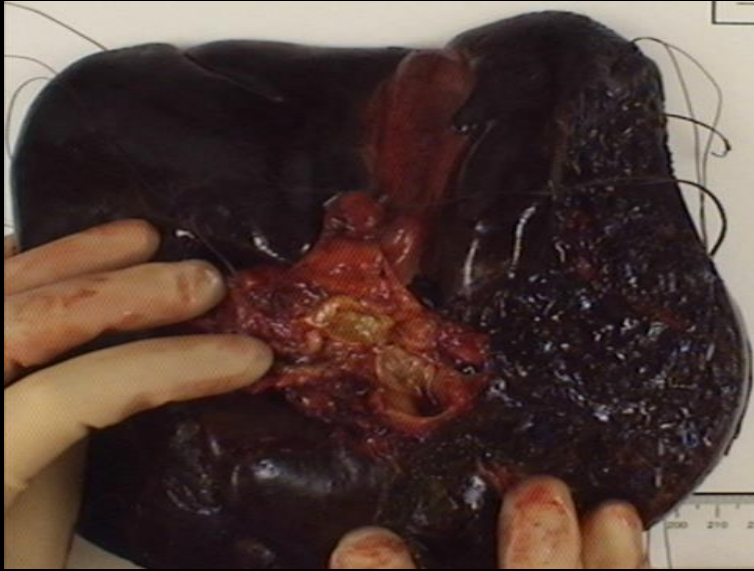




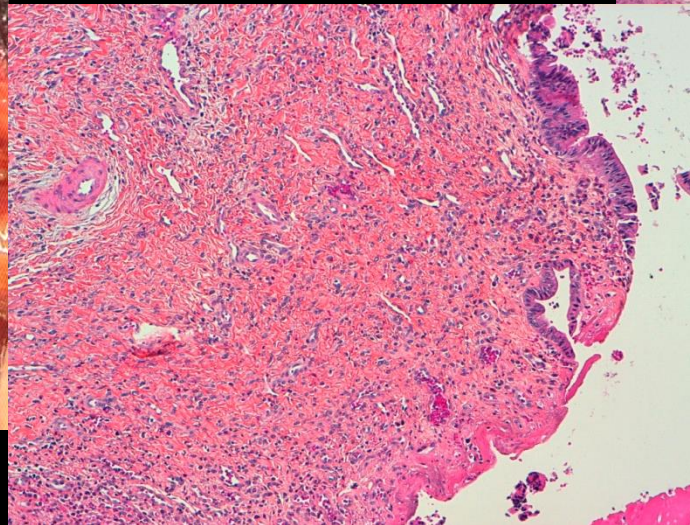
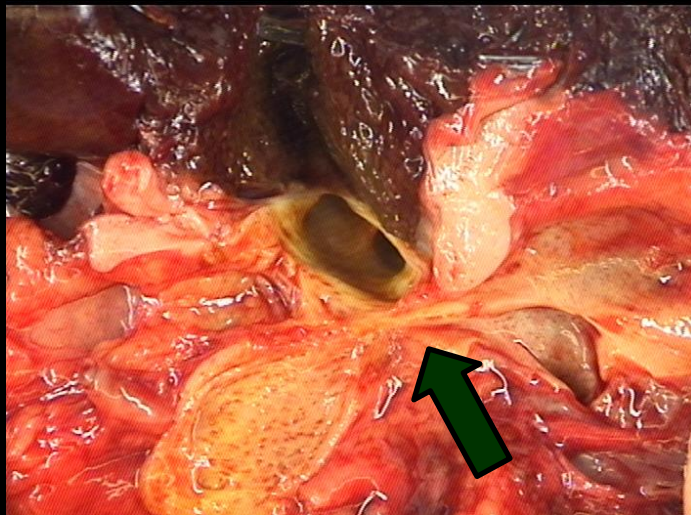
follow up ;2 months after right portal vein embolisation + external biliary drainage of the left hepatic lobe to maximise its hypertrophy



enlarged right hepatectomy

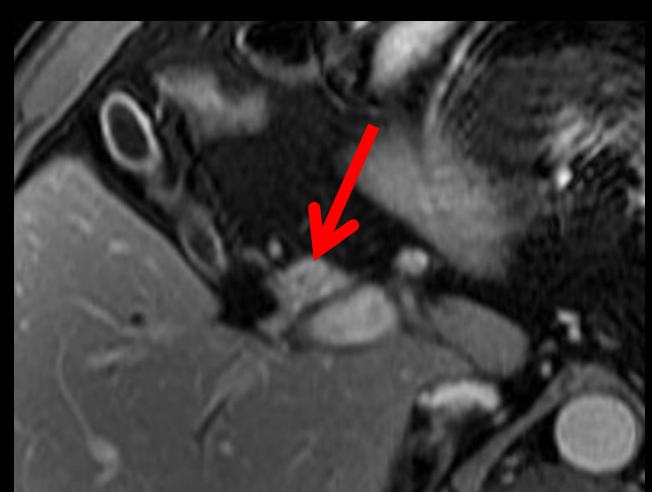
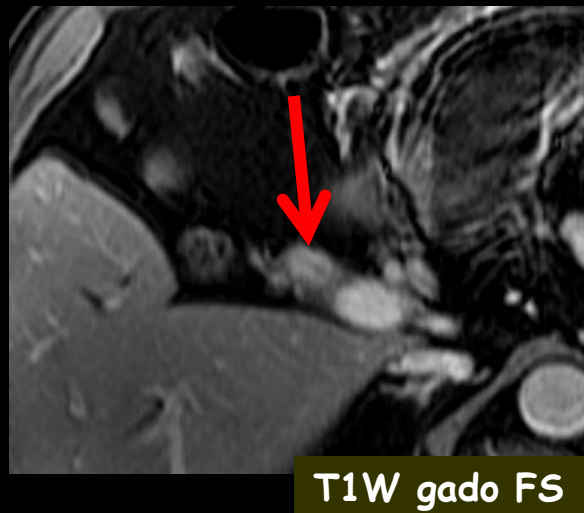
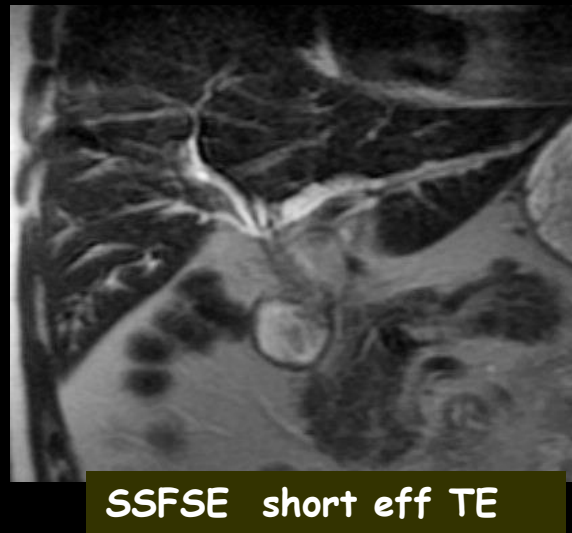
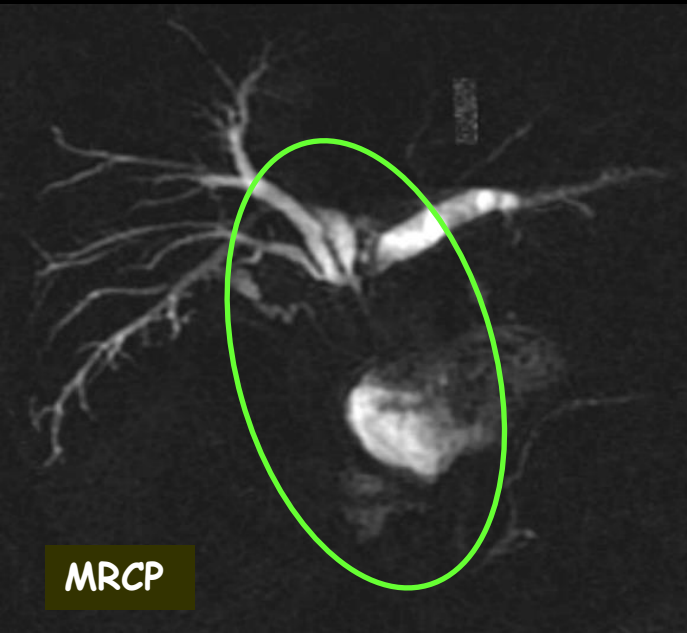


right portal vein
thrombosis

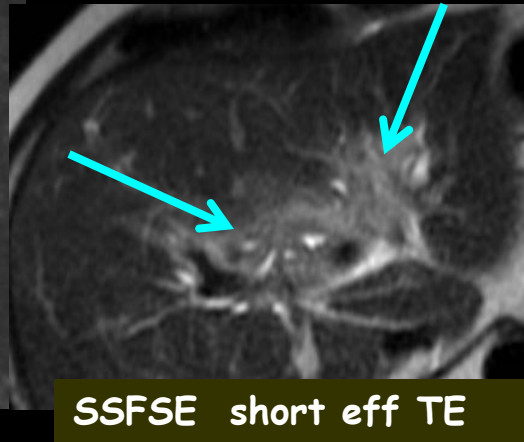
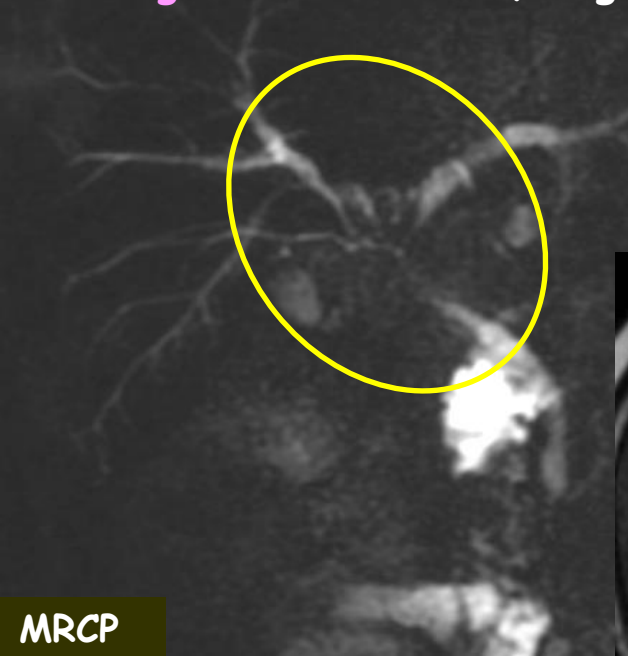


lymphoplasmocytic
infiltration and short
(>5mm)
circonfereential
fibrosis of the
proximal right biliary
duct
;fibrous
transformation of the
adjacent portal vein

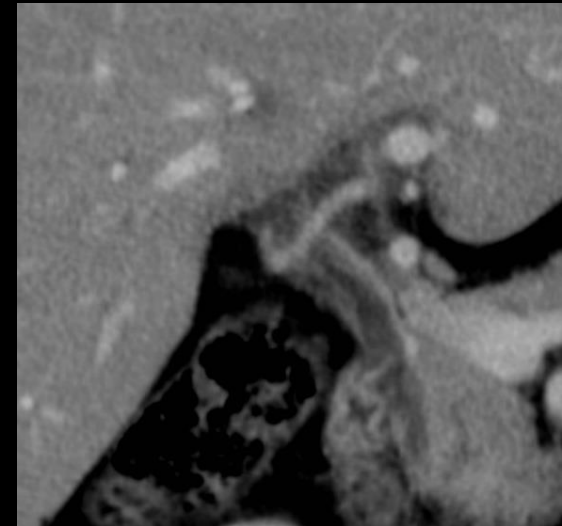
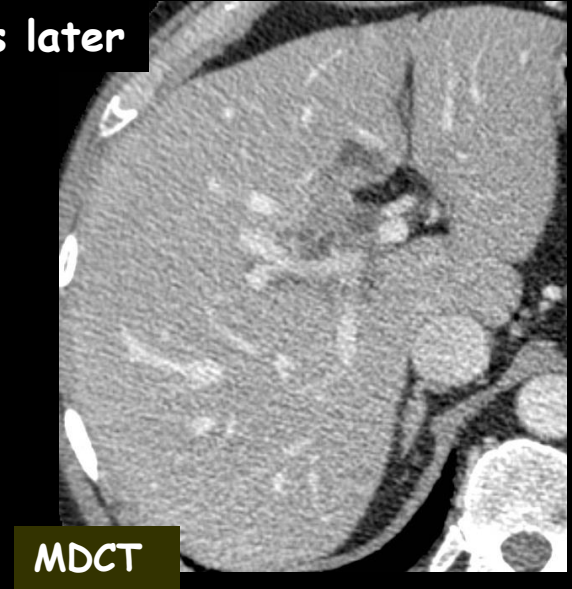
62 yo man , intense **jaundice for 2 weeks** , asthenia , severe biological cholestasis (bilirubin 252 mg/L)



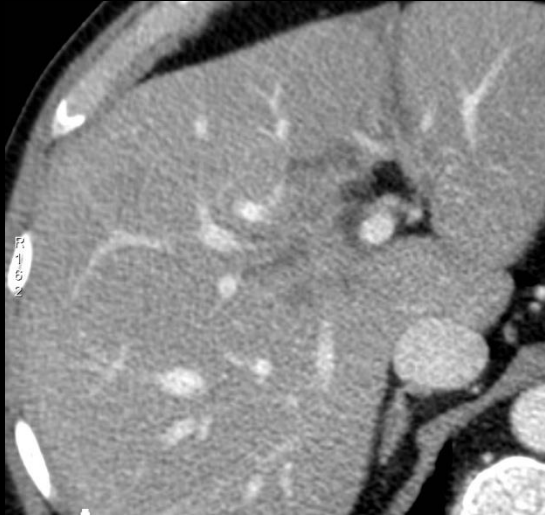
spontaneous slow regression of the icterus with **progressive normalisation of the biological cholestasis** , begining at the end of the first month .



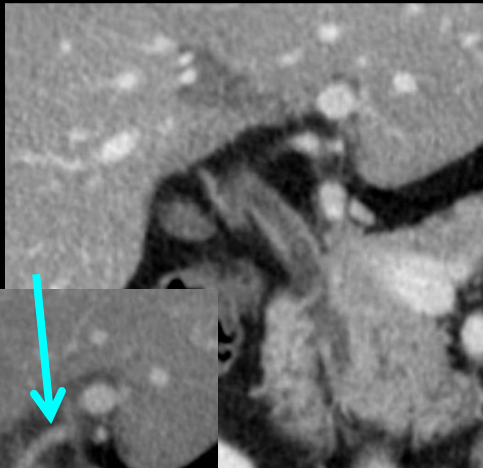
10 months later



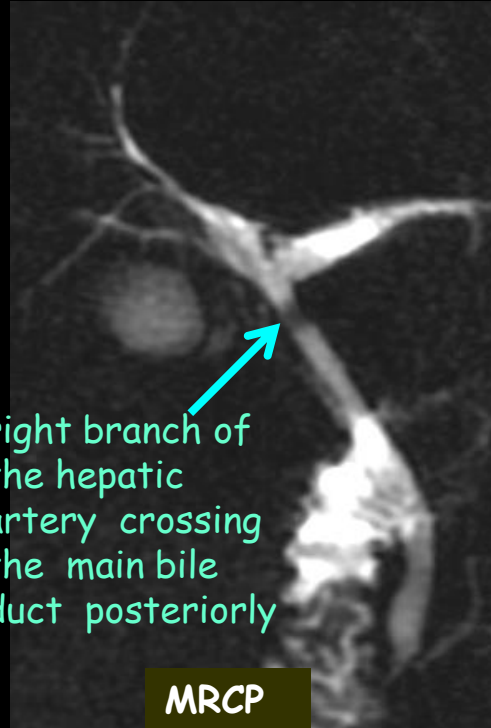
3 years after the beginning



CT

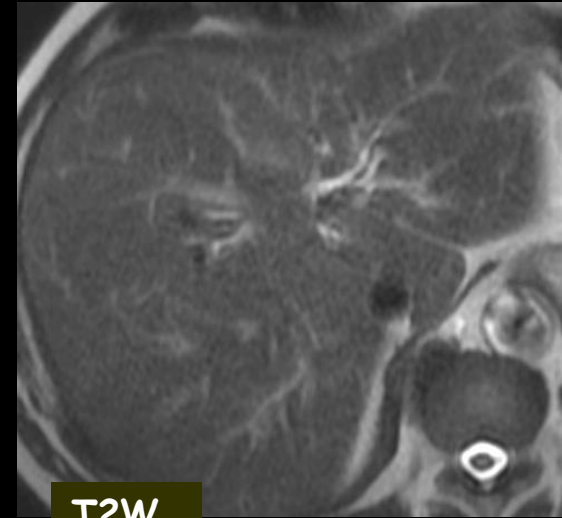


4 years



right branch of
the hepatic
artery crossing
the main bile
duct posteriorly

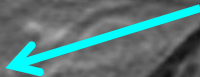
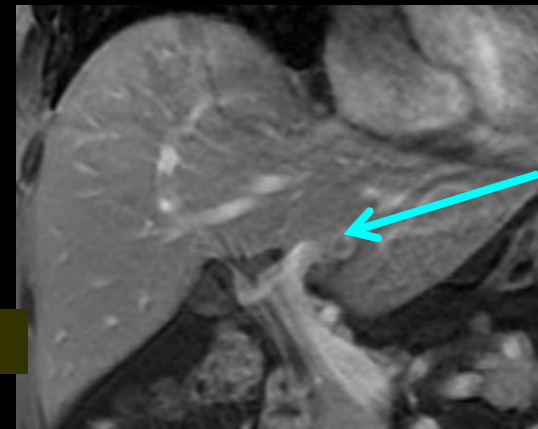
MRCP



T2W



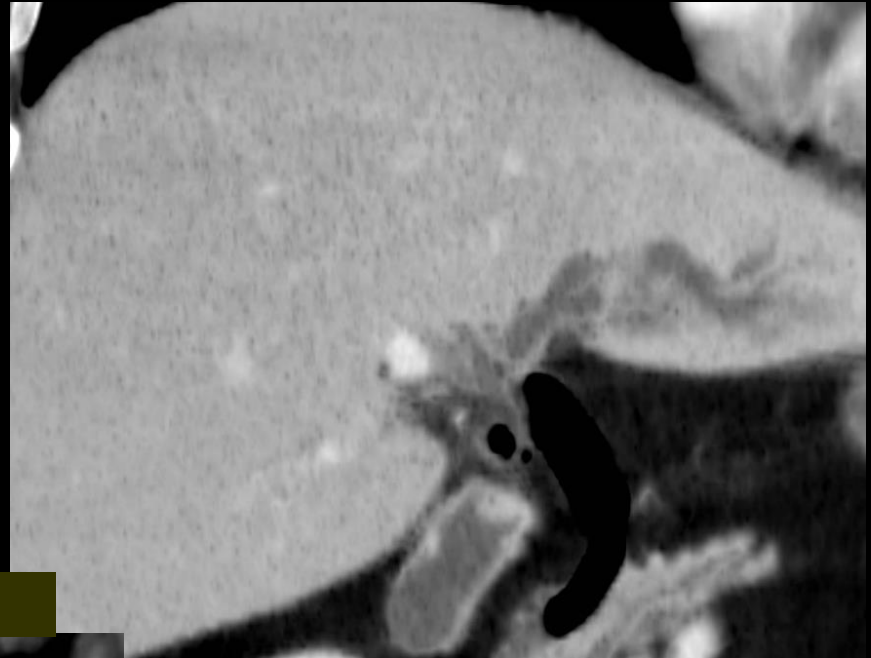
T1W gado FS



78 yo man ; left lobe biliary dilatation with intestinal subocclusion; mild elevation of bilirubinemia



MDCT



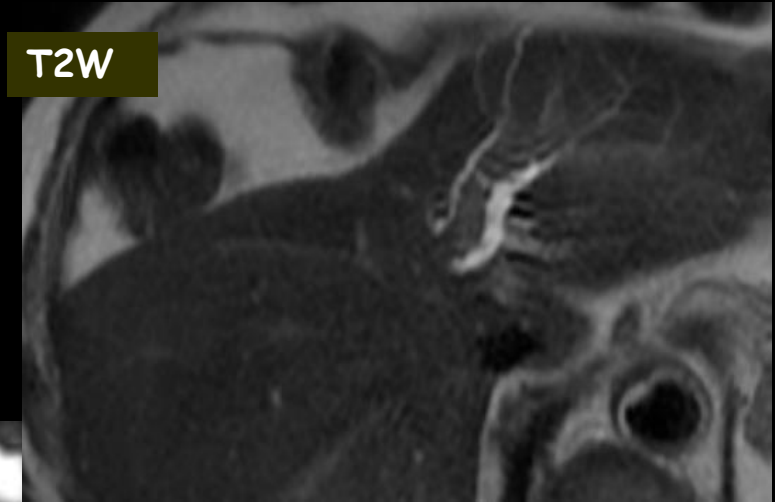
segmental dilatation of the left
hepatic lobe biliary ducts

indication of MRI !

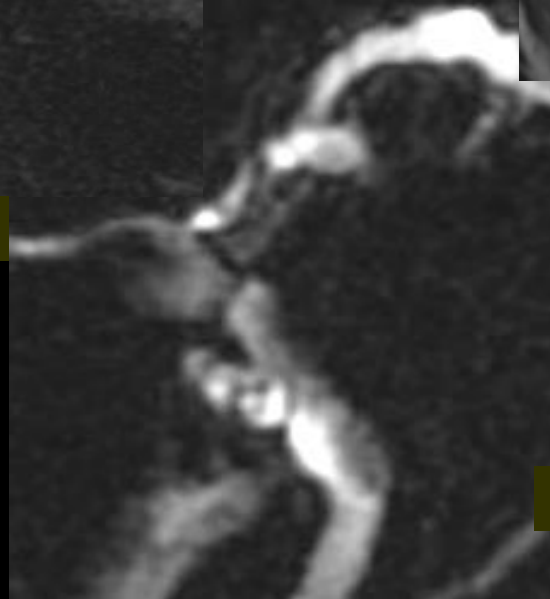
MRI confirm left lobe bile ducts dilatation and thrombosis of adjacent portal branches !



CPIRM



T2W



T1W gado FS

Left hepatic lobectomy , 4 months after the beginning of the symptomatology:
concentric stenosing fibrosis of large as of small bile ducts
inflammatory lympho plasmocytic infiltration
thrombosis of left lobe portal veins ,partially repermeabilised

Sclerosing cholangitis ; associated diseases

Idiopathic

Inflammatory bowel disease
 Crohn's disease
 Ulcerative colitis

Idiopathic fibrosis
 Retroperitoneal fibrosis
 Mediastinal fibrosis
 Peyronie's disease
 Idiopathic lobular panniculitis
 Reidel's thyroiditis

Pseudotumour of the orbit

Autoimmune & connective tissue disorders
 Systemic lupus erythematosus
 Rheumatoid arthritis
 Systemic sclerosis
 Sjögren's syndrome
 Celiac disease
 Type 1 diabetes mellitus
 Autoimmune hemolytic anemia
 Immune thrombocytopenic purpura
 Lupus nephritis

Membranous nephropathy
 Rapidly progressive
 Glomerulonephritis
 Chronic sclerosing sialadenitis
 Primary biliary cirrhosis

Alloimmune disorders
 Hepatic allograft rejection
 Graft-versus-host disease

Infiltrative disorders
 Amyloidosis
 Sarcoidosis
 Systemic mastocytosis
 Hypereosinophilic syndrome
 Hodgkin's disease
 Cholangitis glandularis proliferans

Secondary etiology

Cholelithiasis/Choledocholithiasis

Infection
 Bacterial cholangitis
 Recurrent pyogenic cholangitis

Immunodeficiency-related
 Congenital immunodeficiency
 Acquired immunodeficiency
 Combined immunodeficiency
 Angioimmunoblastic lymphadenopathy

Congenital
 Caroli's disease
 Cystic fibrosis

Pancreatic disorder
 Autoimmune pancreatitis
 Chronic pancreatitis

Toxic
 Intraductal formaldehyde or hypertonic saline
 Intra-arterial chemotherapy

Ischemic
 Vascular trauma
 Hepatic allograft arterial occlusion
 Paroxysmal nocturnal hemoglobinuria
 Posttraumatic sclerosing cholangitis

Others
 Hepatic inflammatory pseudotumor
 Neoplastic/Metastatic disease
 Eosinophilic cholangitis
 Portal biliopathy
 Langerhans cell histiocytosis

11. Abdalian R, Heathcote EJ. Sclerosing cholangitis: a focus on secondary causes. *Hepatology* 2006;44:1063-74

Sclerosing cholangitis may mimic hilar or main bile duct cholangiocarcinoma

It is always difficult to get precise histologic data with biopsies or endoluminal biliary brushing and surgical resection is often decided mainly on MDCT and MR images .

Of course precise knowledge of clinico-biological evolution is fundamental to avoid unnecessary mutilating surgery

and we have to be very careful in radiology reports and to take in account clinical , biological atypies in multidisciplinary concertations.

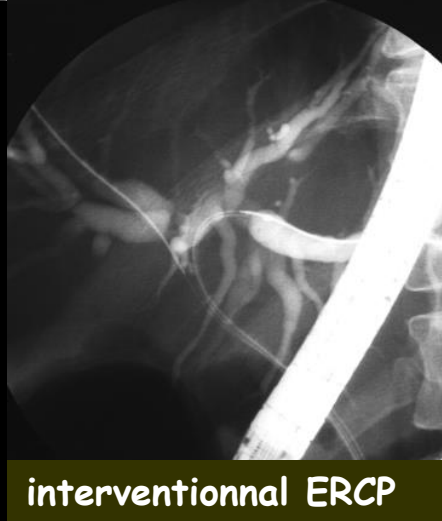
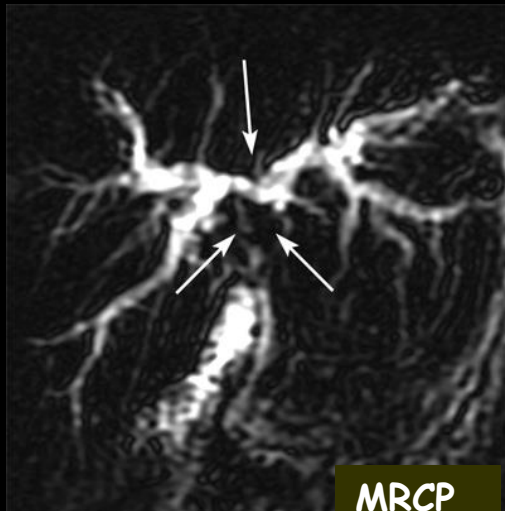
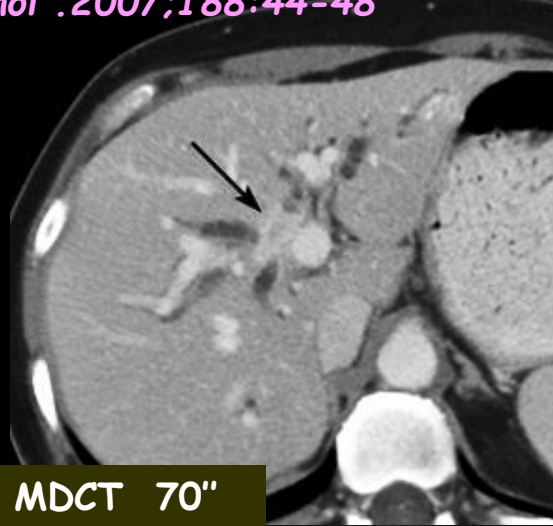
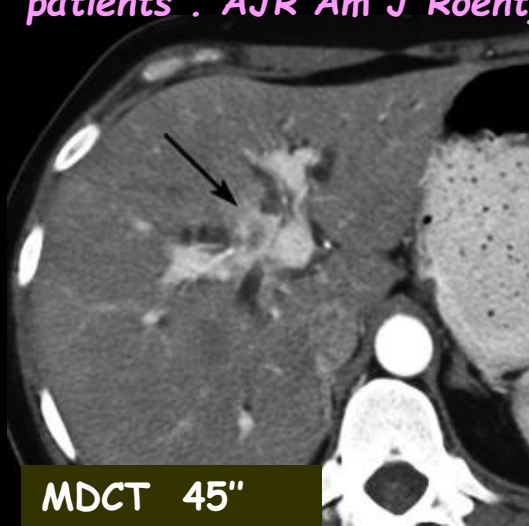
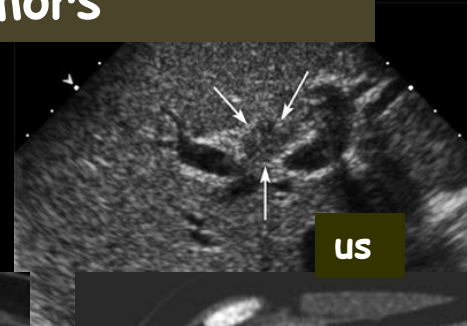
In atypical cases , one can decide

- to complete biological data ; IgG4, auto antibodies , CA 19-9 ...etc
- a supplementary follow-up of one to three months
- a trial of **corticotherapy (?)**
- repeated endobiliary brushings or better : **endoluminal biliary duct biopsy by transhepatic US guided approach**

1b. hepato biliary inflammatory pseudotumors

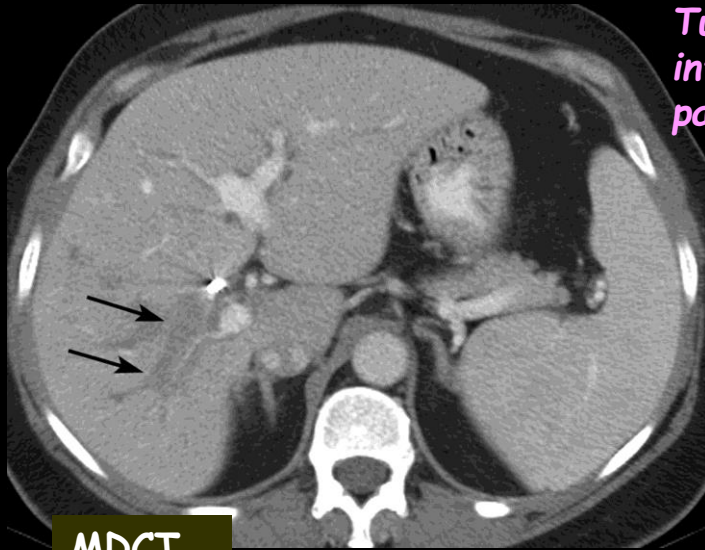
48 yo woman , jaundice ,cholestasis

Tublin ME, Moser J, Marsh JW, Gamblin TC .Biliary inflammatory pseudotumor.imaging features in seven patients . AJR Am J Roentgenol .2007;188:44-48



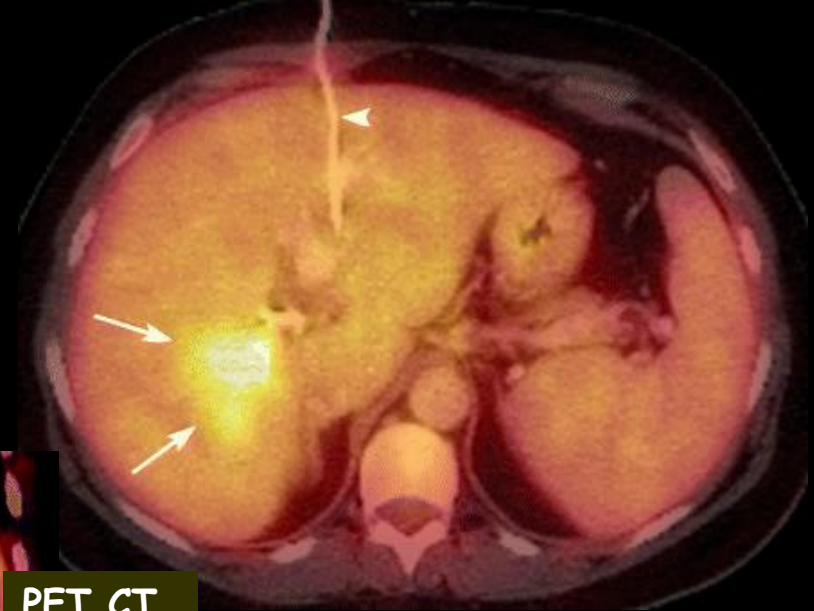
guided biopsies of the juxta hilar mass confirm the diagnosis of inflammatory pseudotumor

42 yo woman ,mild biological cholestasis without clinical icterus , probabilistic diagnosis of Klatskin's tumor



MDCT

Tublin ME, Moser J, Marsh JW, Gamblin TC .Biliary inflammatory pseudotumor.imaging features in seven patients . AJR Am J Roentgenol .2007;188:44-48

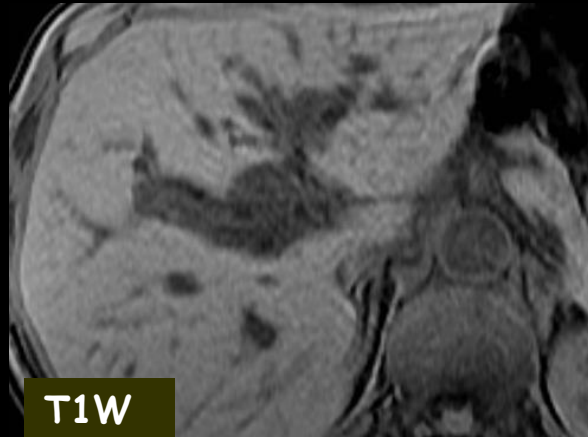
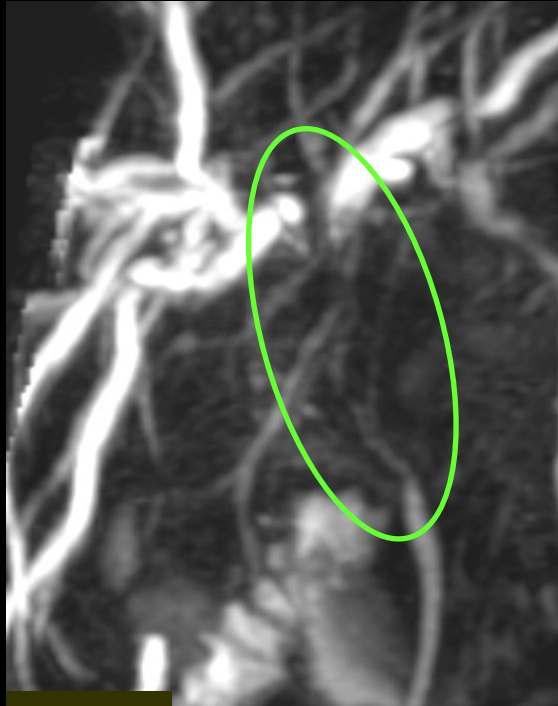


PET CT

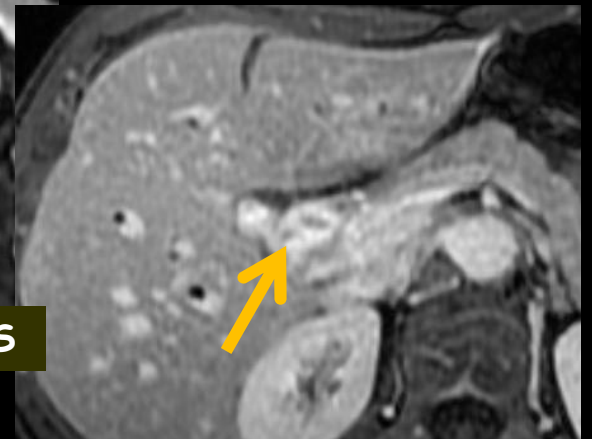
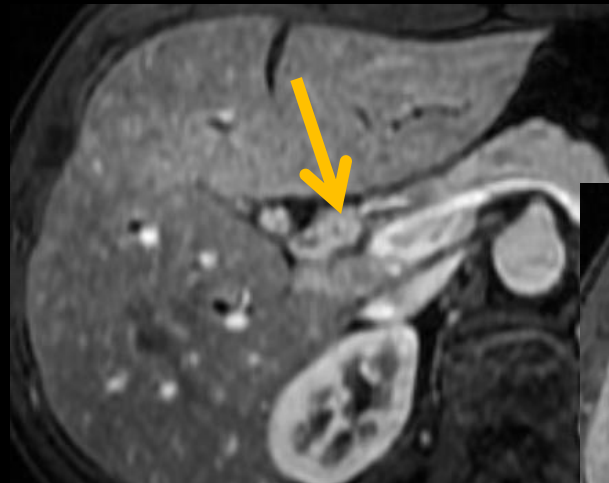


right hepatectomy (after neo-adjuvant chemotherapy !!) confirms the histological diagnosis of biliary inflammatory pseudotumor

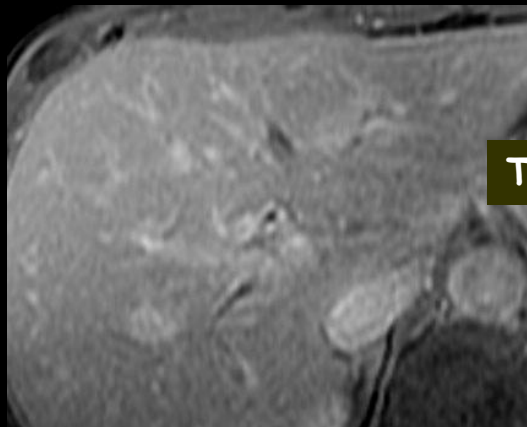
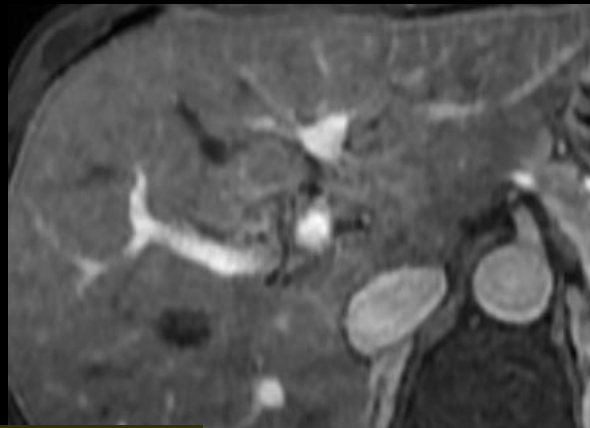
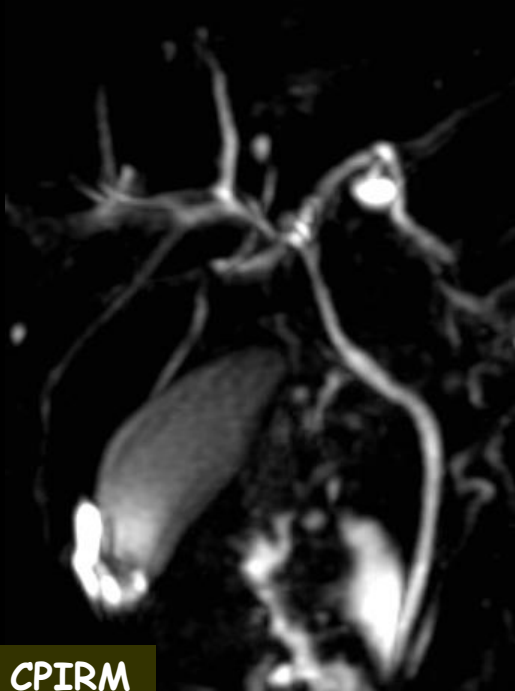
48 yo woman, right upper quadrant pain with mild fever, moderate cholestasis (bilirubin 53 mg/L), elevated AST and ALT (15/20*N) CRP 20 mg/L.



hilar mass-forming lesion with upstream biliary duct dilatation



one month later, the whole anomalies spontaneously disappeared, clinico-biologic
as radiologic ones

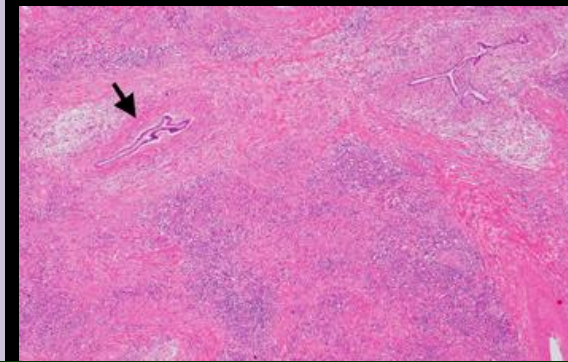


Hepatobiliary inflammatory pseudotumor is the main differential diagnosis of mass-forming hilar (Klatskin's) tumor

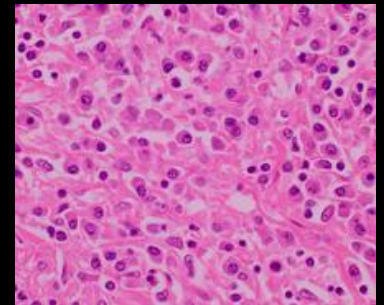
Inflammatory pseudotumors are histologically characterized by a proliferation of fibroblasts or myofibroblasts and inflammatory cell infiltration (mainly polyclonal lymphocytes and plasma cells)

Two types are described: fibrohistiocytic predominantly peripheral and lymphoplasmocytic , central (juxta hilar) and mass forming (with IgG4 elevation)

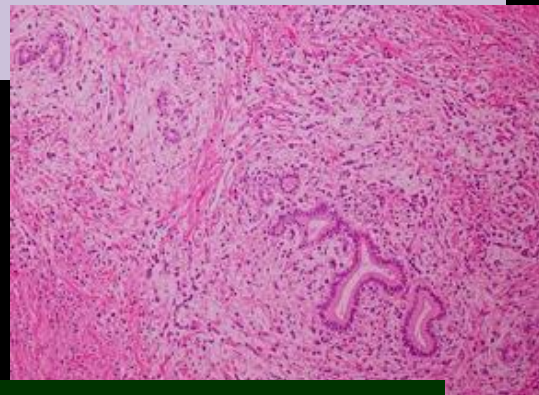
Delayed enhancement in contrasted MDCT and MRI is a strong argument for the diagnosis ; unfortunately it is also seen in cholangiocarcinoma



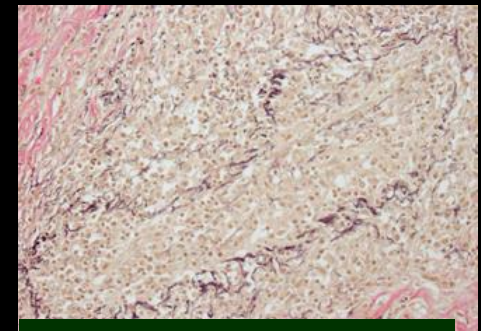
lymphoplasmocytic infiltration



oedematous "immature" fibrosis .



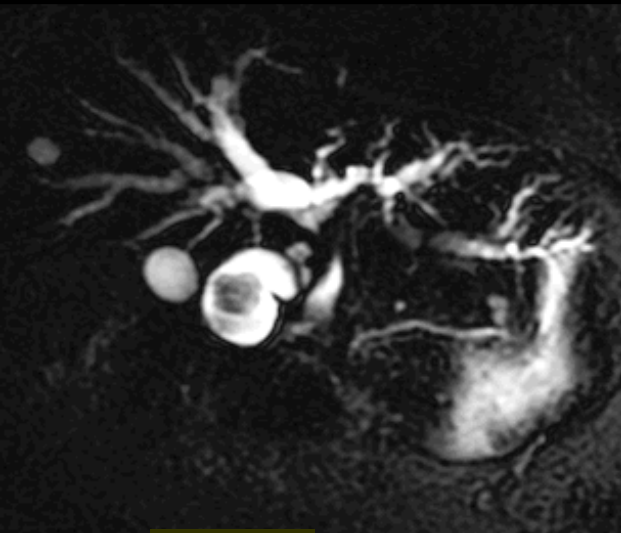
periglandular fibrosis



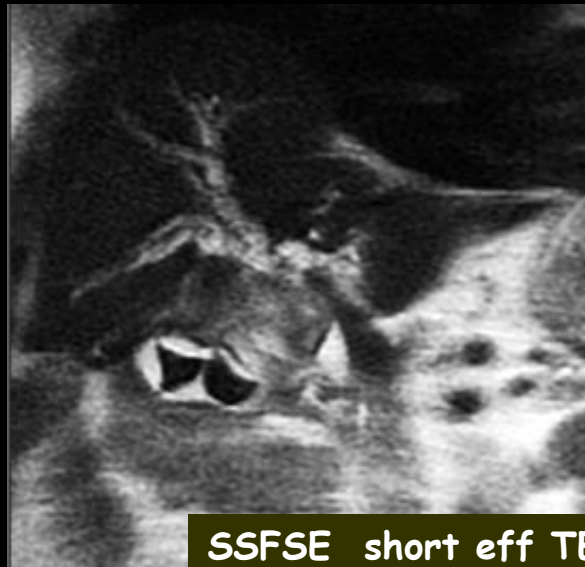
perivenous and centroluminal fibrosis

1c. differential diagnosis of main bile duct "tumor-like" lesions

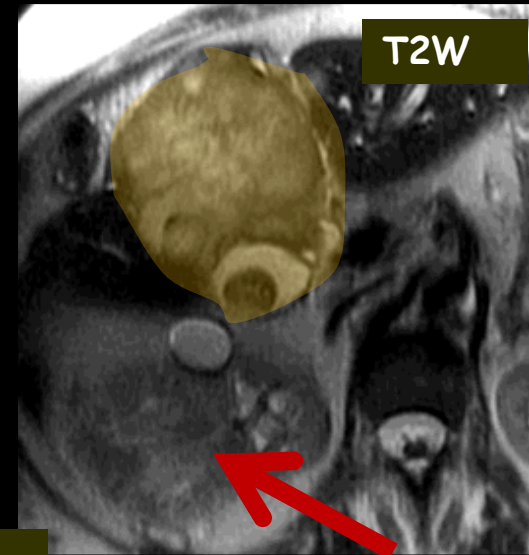
73 yo woman , inreasing jaoundice , palpable mass of the right upper quadrant



MRCP



SSFSE short eff TE



T2W



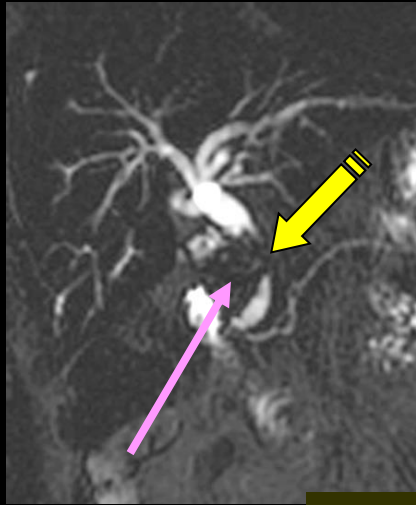
T1 gado

calculo-cancer of the gallbladder with infrahilar pedicular extension

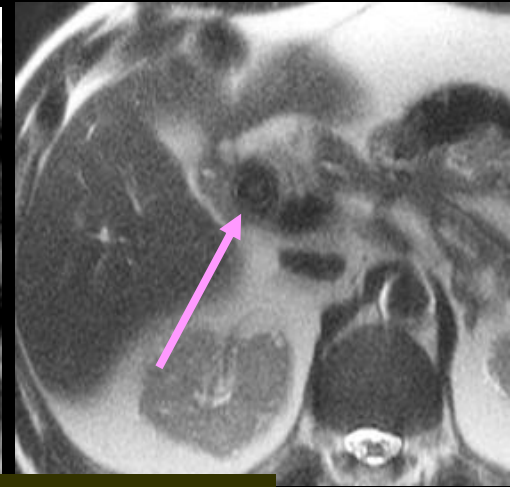
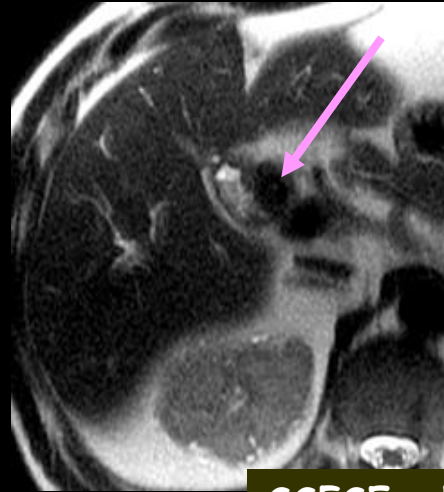
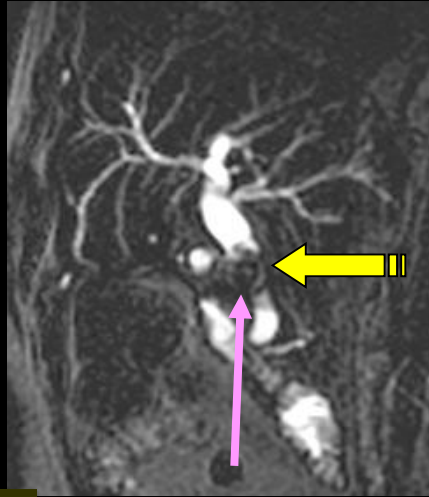
Grawitz tumor of the right kidney



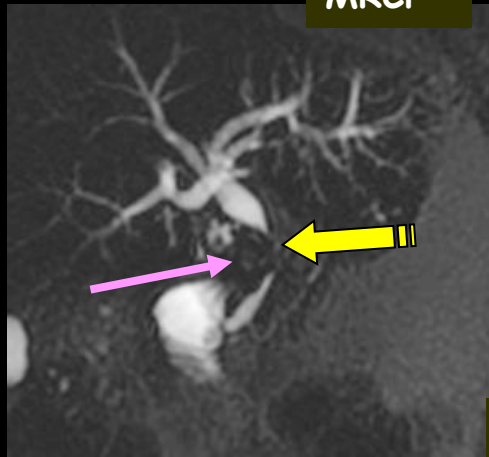
73 yo woman , biliary pain and biological cholestasis ;**cholecystectomy 3 years** , no weight loss.



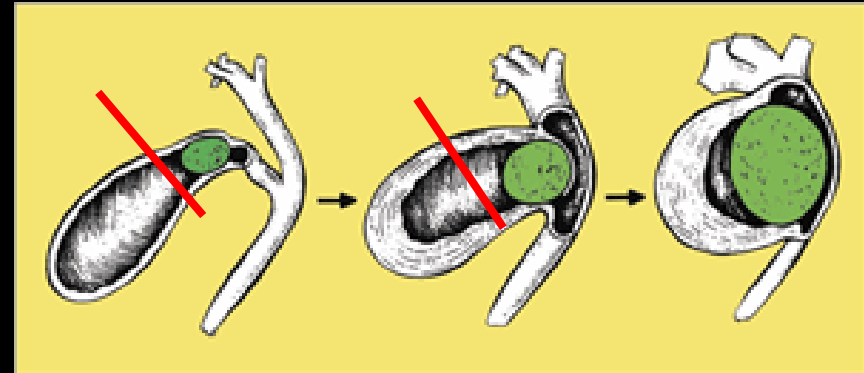
MRCP



SSFSE short eff TE

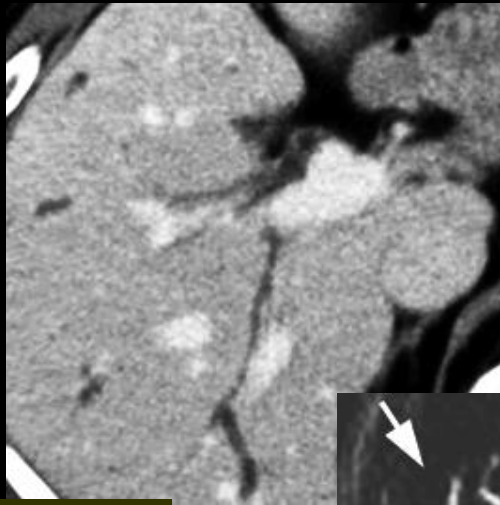


T1W gado

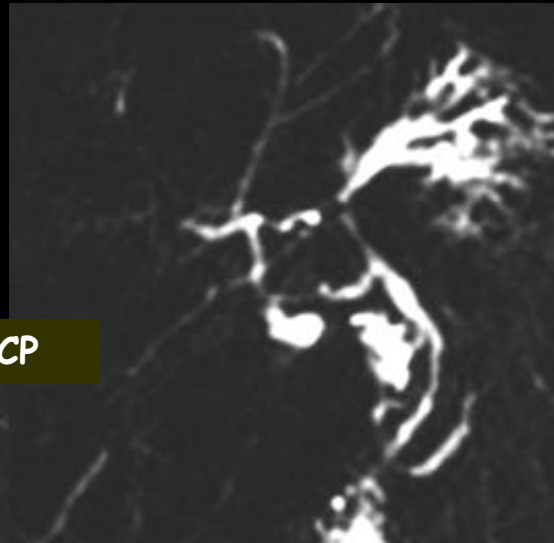


Mirizzi's syndrome (pedicular infrahilar compression due to cystic stone) can be seen even after cholecystectomy , in a long cystic duct remnant !!!

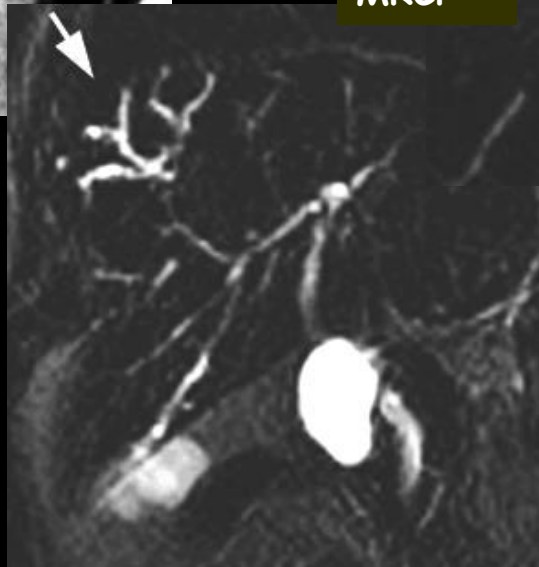
54 yo man , chronic diarrhea, weight loss, cholestasis



MDCT



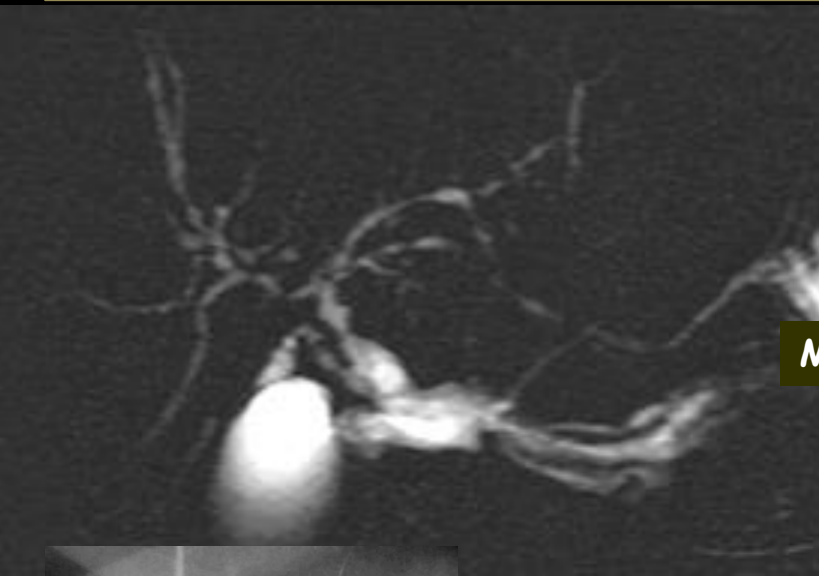
MRCP



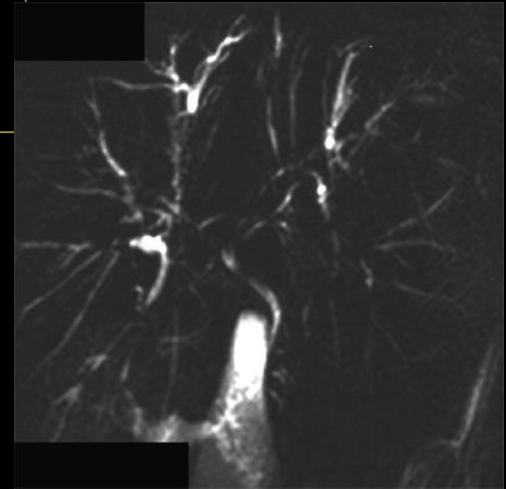
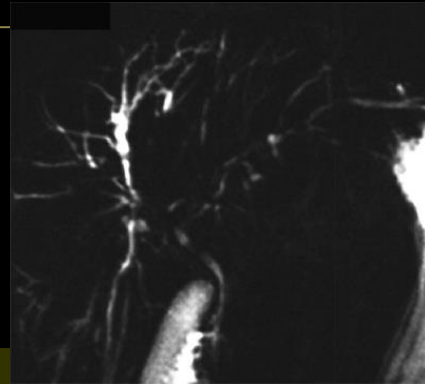
primary sclerosing cholangitis

How can we recognize a primary sclerosing cholangitis ?

Vuillierme MP et coll. Cholangites JFR 2007. Paris SFR ed 2007.



MRCP



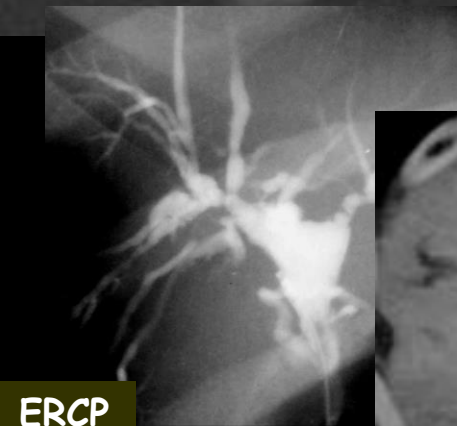
obliterating fibrous cholangitis involve large intrahepatic bile ducts (\neq primary biliary cirrhosis)

young men S/R 2/1, averag age of diagnosis : 40 yo

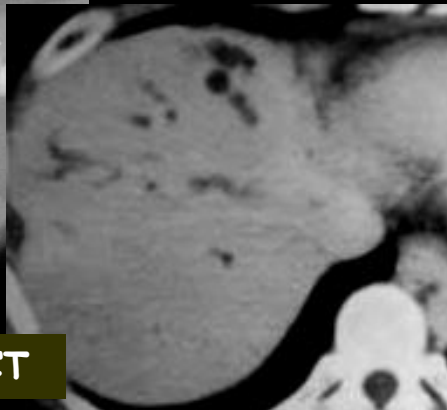
frequently (75%) associated to IBD (specially mild or infraclinic ulcerative pancolitis)

IgG4 are elevated in only 9% of the cases

annual incidence of cholangiocarcinoma : 1.5% (elevation of CA 19.9)



ERCP



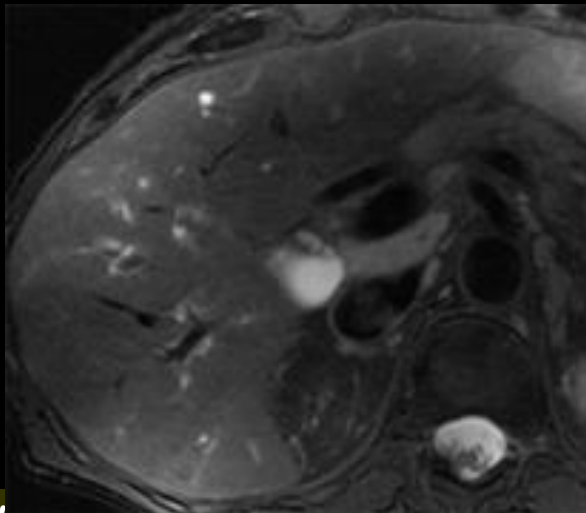
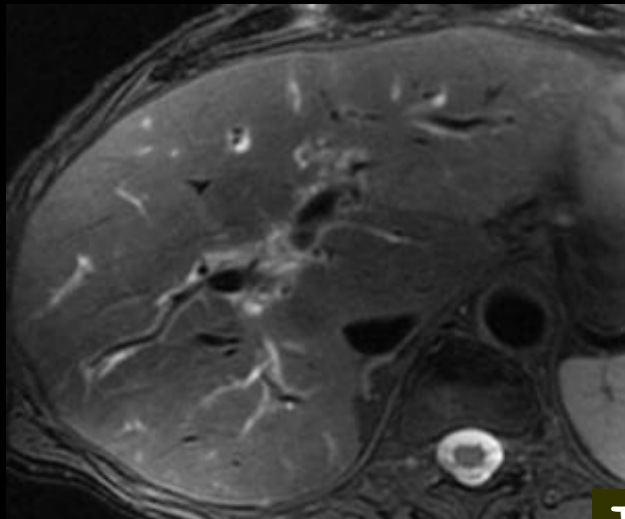
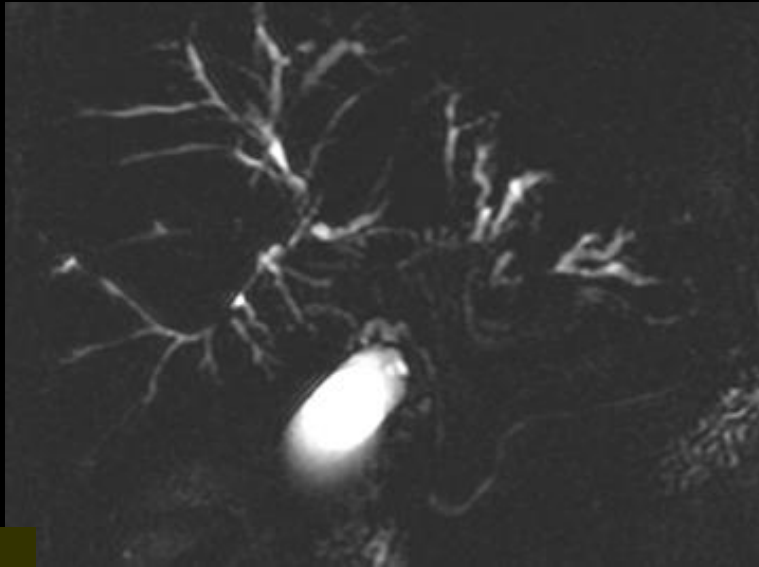
CT

short stenosis with upstream dilatations ,predominantly peripheral ; beading.

72 yo man, follow up of primary sclerosing cholangitis

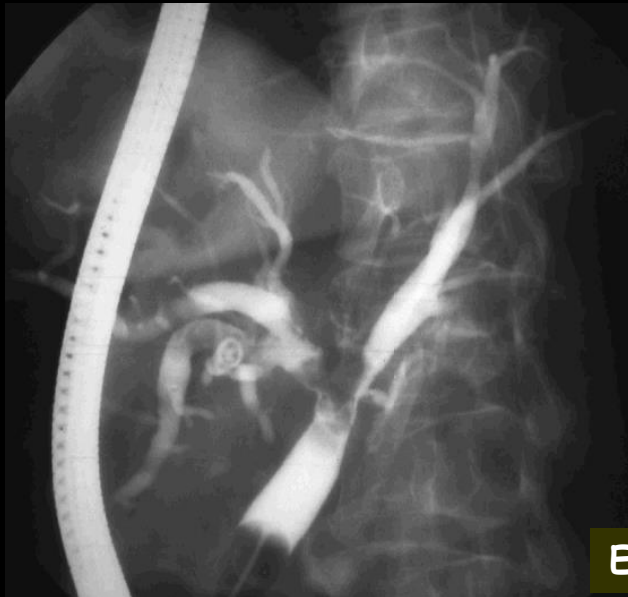
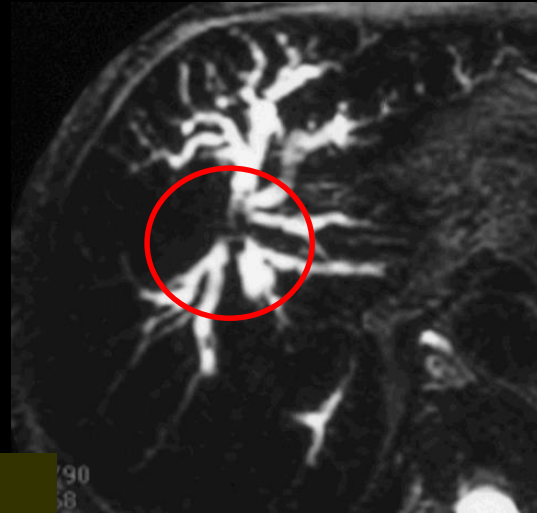
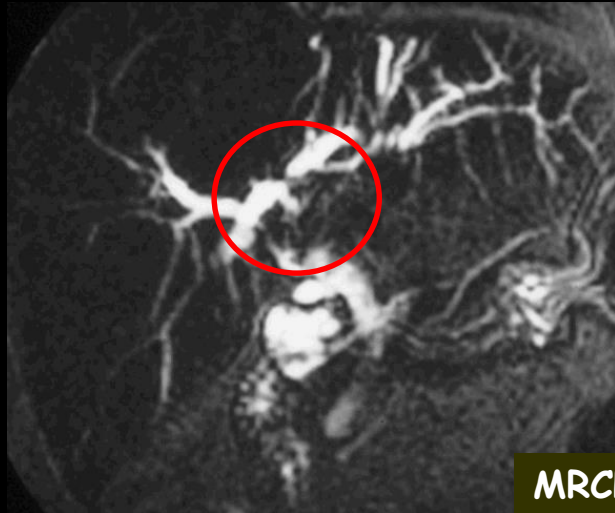


MRCP



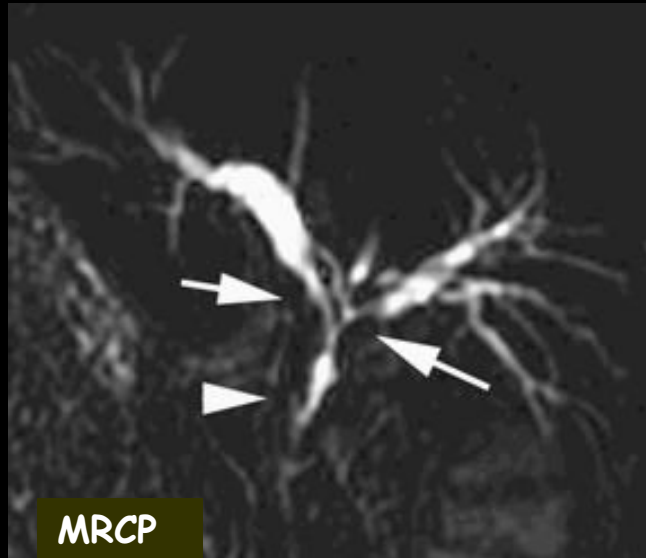
T2W

69 yo woman ; **cholecystectomy 12 years ago** ; biliary pain , fever, cholestasis

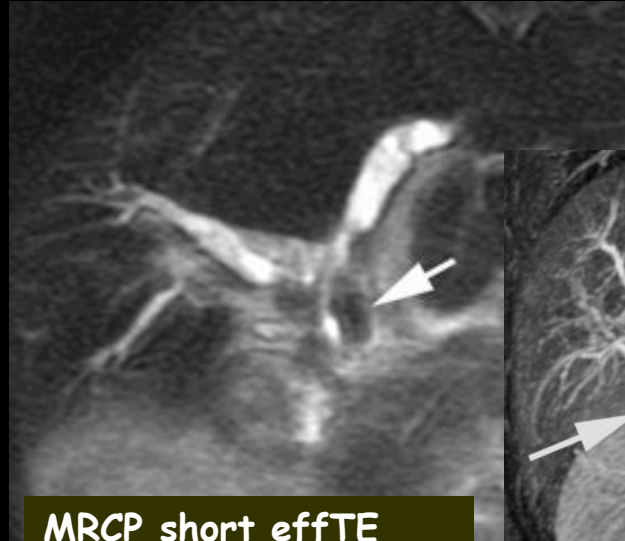


Cholesterol stones
incrusted in the wall
of proximal hilar bile
ducts (confirmed at
surgery)

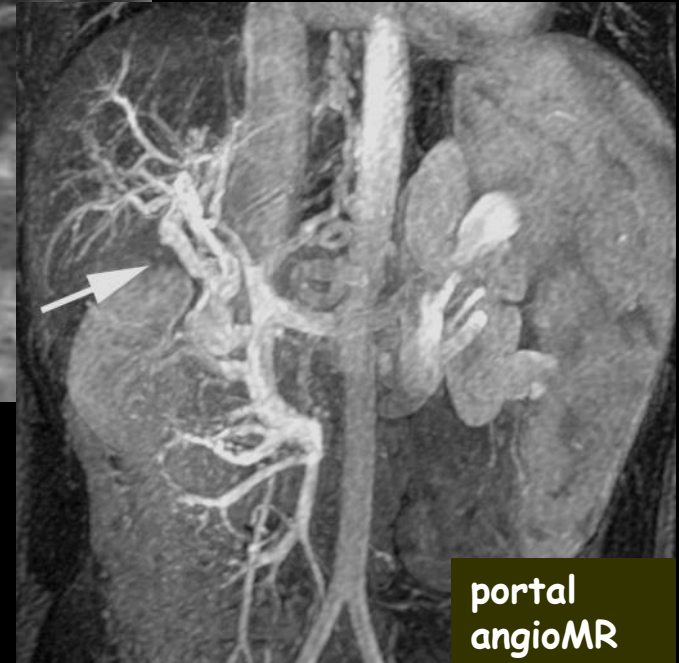
18yo teenager; **cystic fibrosis** with liver insufficiency



MRCP



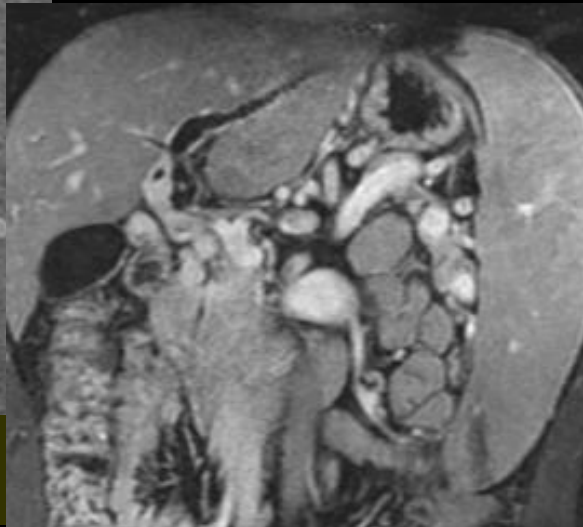
MRCP short effTE



portal
angioMR



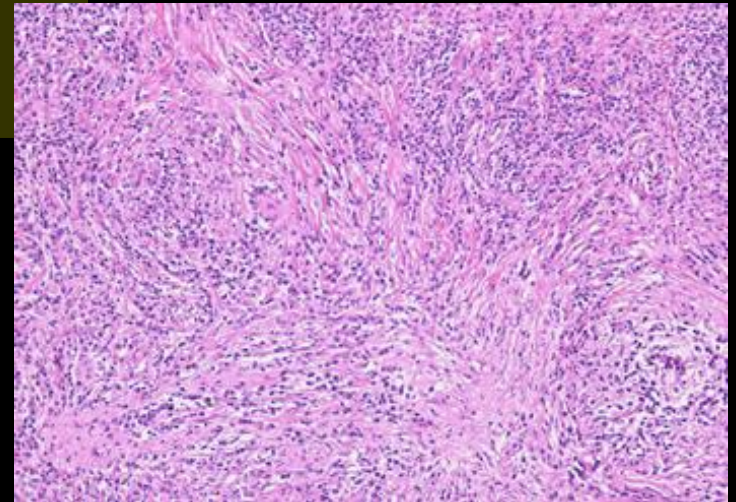
T1W
gado



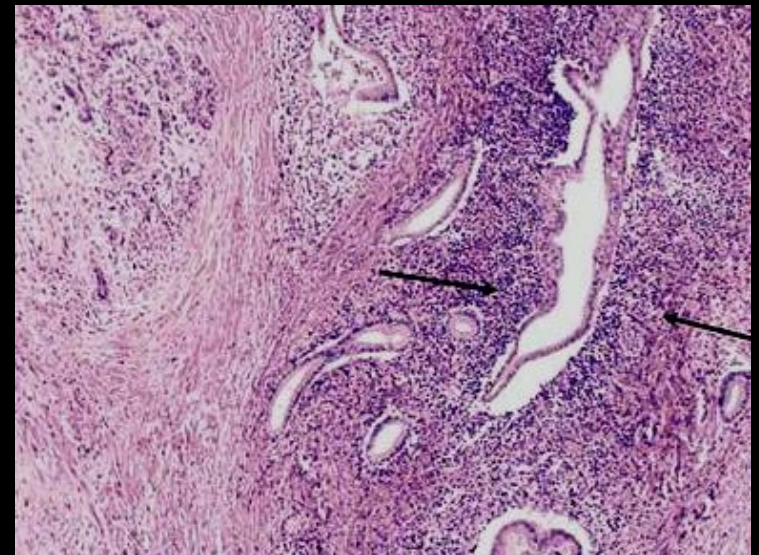
Pedicular and porta hepatis portal varices due to portal hypertension (secondary biliary cirrhosis and portal thrombosis .often misnamed "portal cavernoma")

2a. autoimmune pancreatitis

- autoimmune pancreatitis (AIP) is a **chronic inflammation** of the pancreas due to a **postulated autoimmune mechanism** .
- occasionally, AIP is **associated with other autoimmune disorders** such as chronic IBD , autoimmune cholangitis, Sjögren's disease, retroperitoneal fibrosis, diabetes mellitus , SLE, vasculitis , thyroiditis
- histologic hallmark is **inflammatory lymphoplasmacytic periductal inflammation** with **massive fibrosis**
- AIP is the main differential diagnosis of pancreatic adenocarcinoma and is important to avoid unnecessary surgery
- remarkable response to steroid therapy** because of its autoimmune pathogenesis **must be used to confirm the diagnosis**



lymphoplasmacytic sclerosing pancreatitis



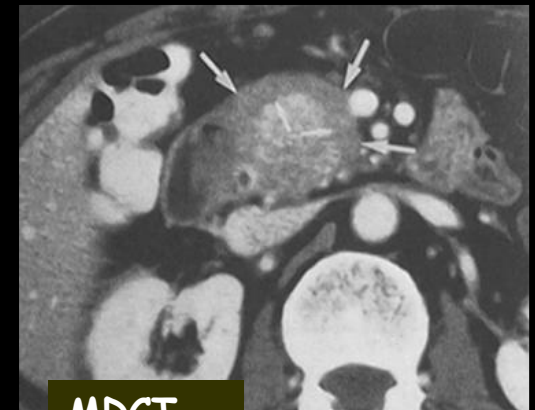
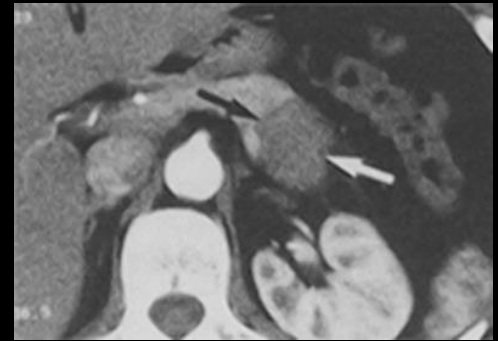
periductal collar of
lymphoplasmacytic infiltration

MDCT imaging of autoimmune pancreatitis

- focal (head) or diffuse hypertrophy (sausage shaped) of the pancreas
- loss of lobularity
- no peripancreatic fat stranding
- peripheral rim of a hypoattenuation "halo"
- associated distant lesions (autoimmune cholangitis , retroperitoneal fibrosis ..)



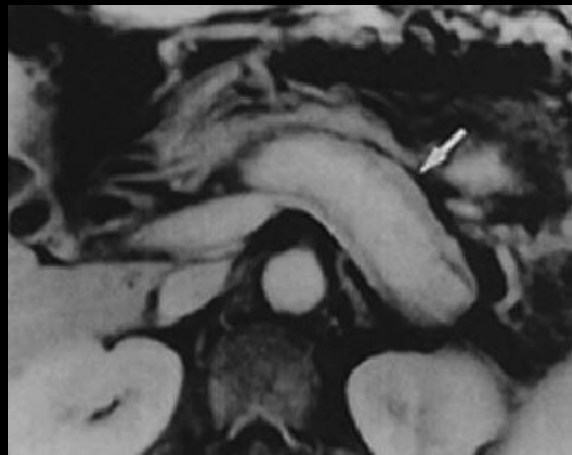
MDCT



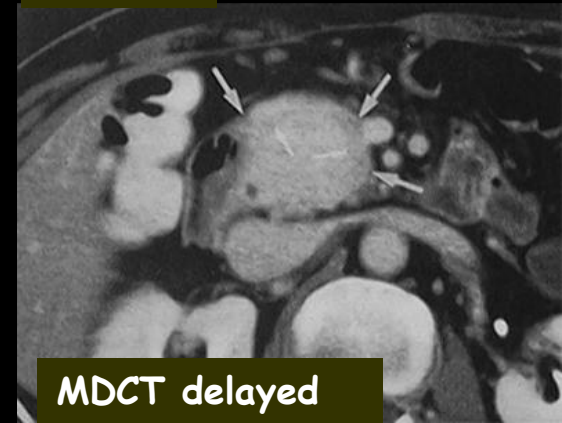
MDCT



T2W

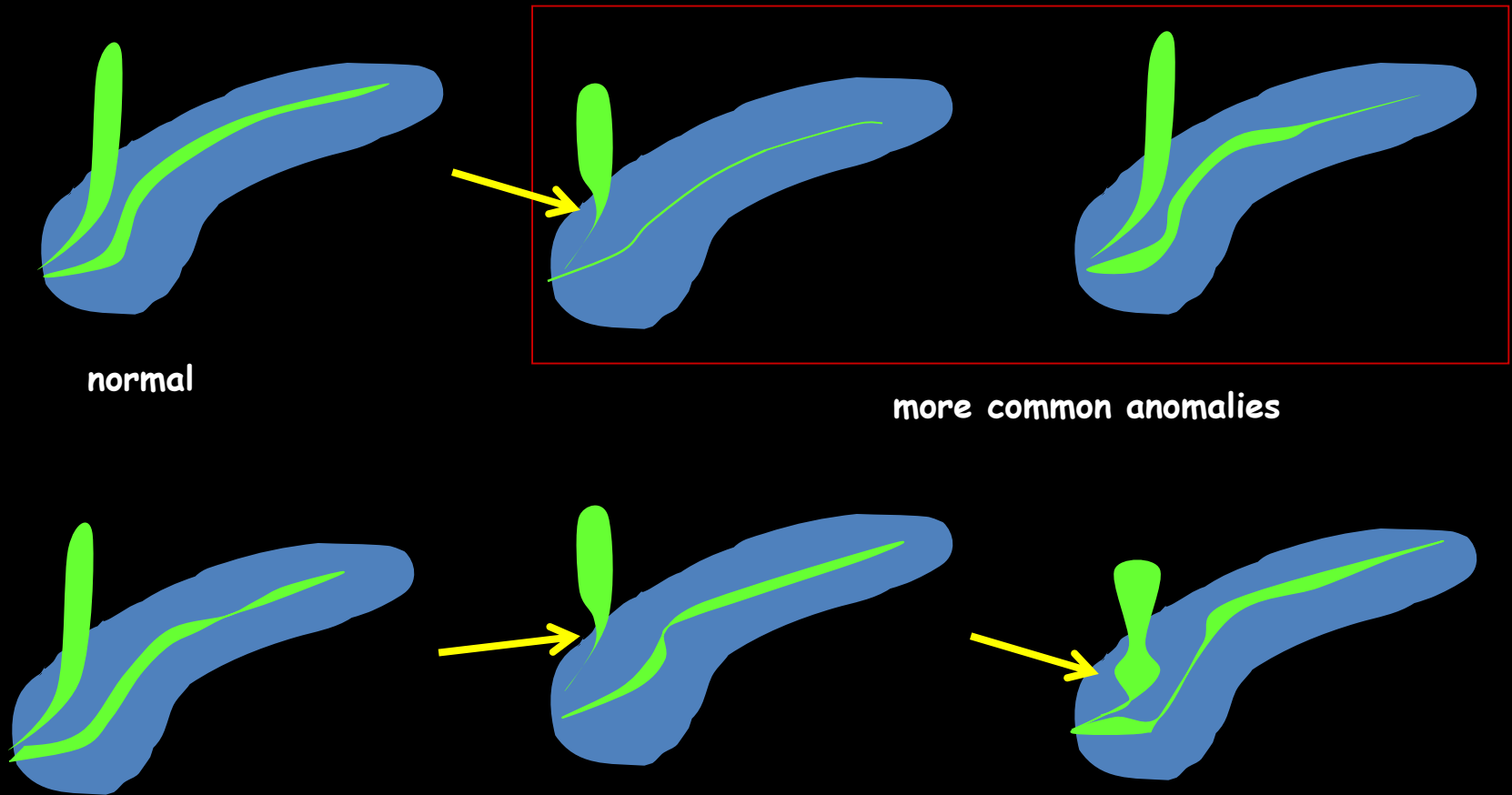


T1W gado ,delayed

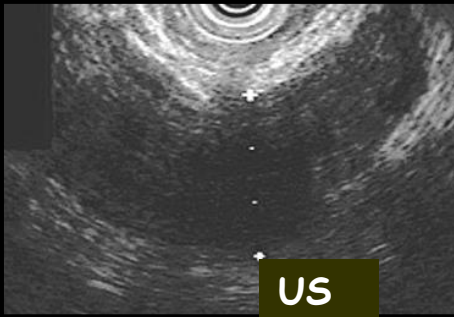


MDCT delayed

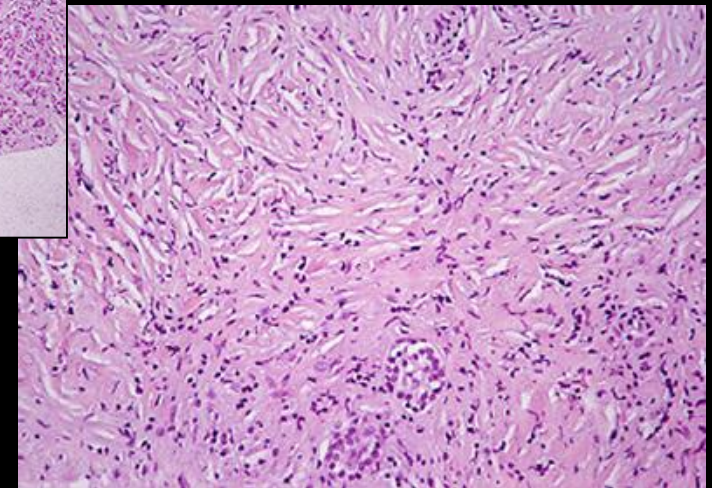
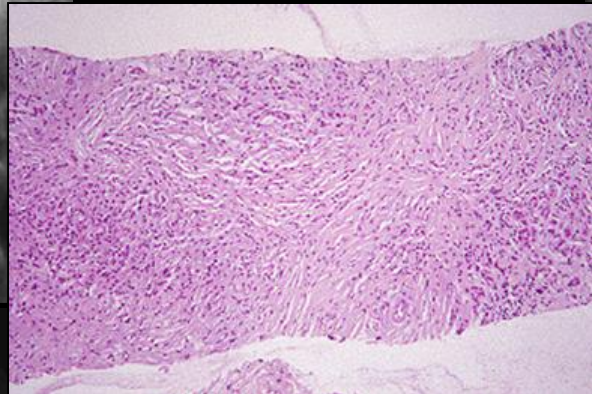
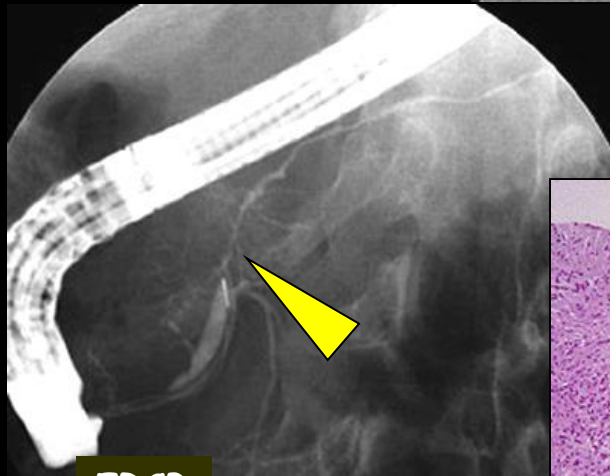
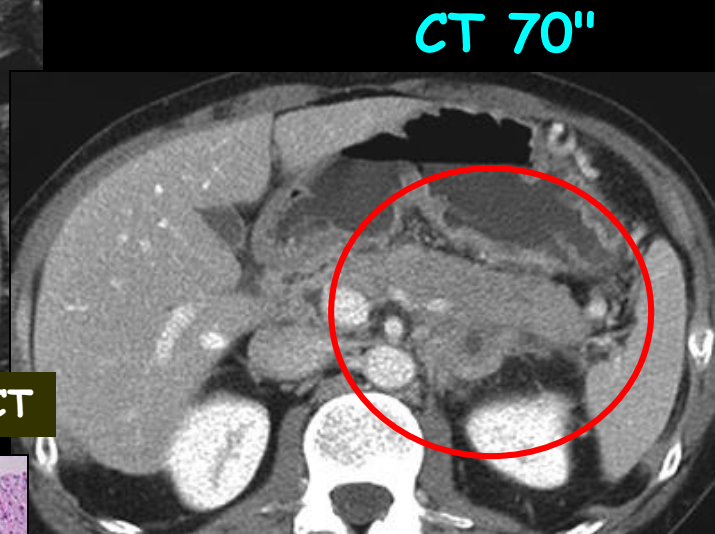
ductal pancreatobiliary lesions in autoimmune pancreatitis (MRCP or retrograde endoscopic wirsungography)



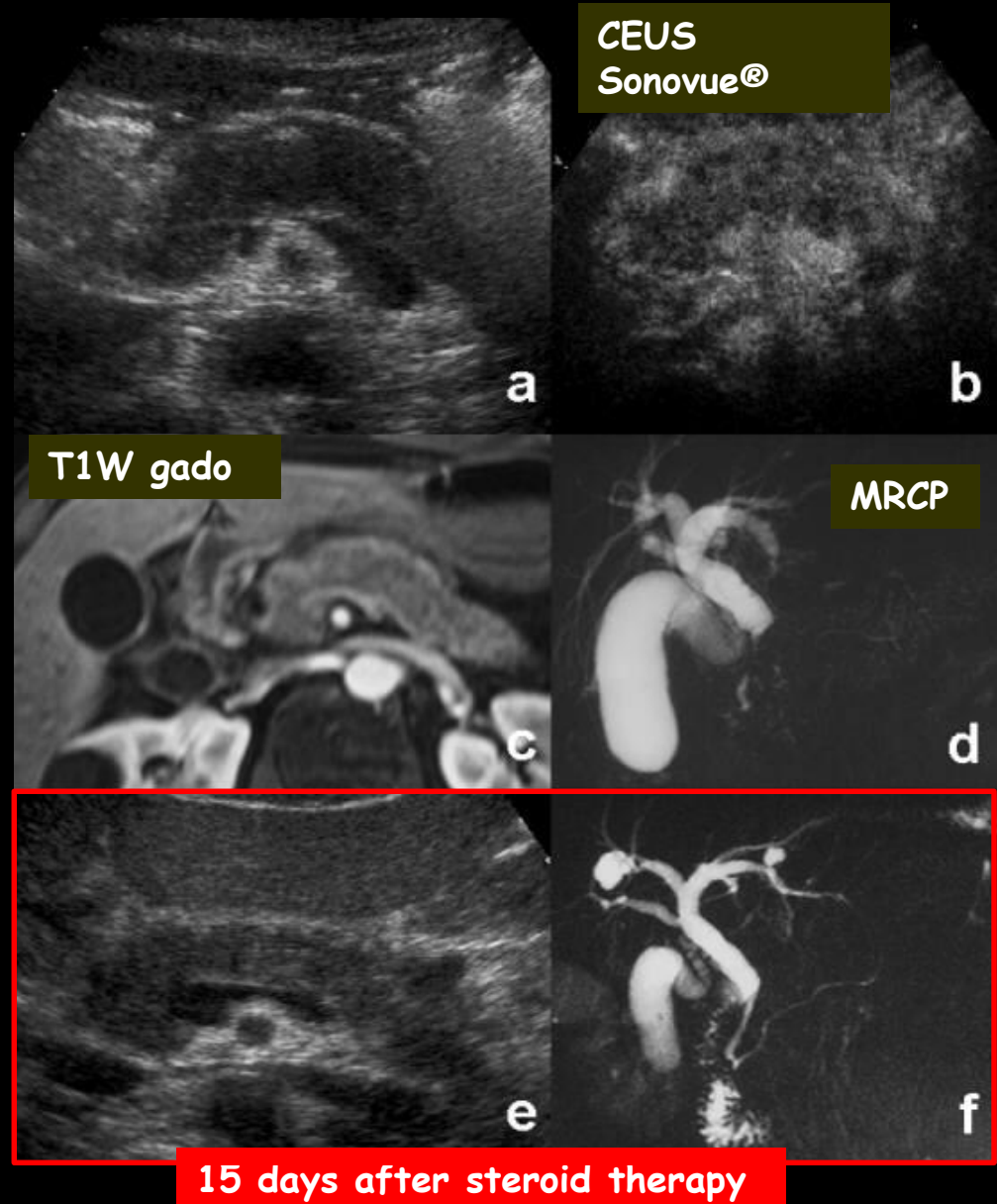
Diffuse pancreatic ductal narrowing without or with minimal upstream dilatation ; stenosis of intrapancreatic main bile duct is frequently observed



CT 35"

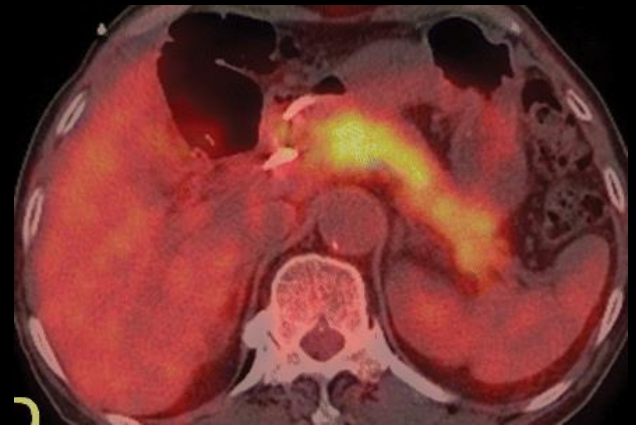


-quick response to corticosteroid (40 mg /day for 1 week followed by a taper of the daily dose by 5 mg per week) is essential to know and to use for the diagnosis



.Morana G et al. Autoimmune pancreatitis: Instrumental diagnosis JOP. J Pancreas (Online) 2005; 6(1 Suppl.):102-107

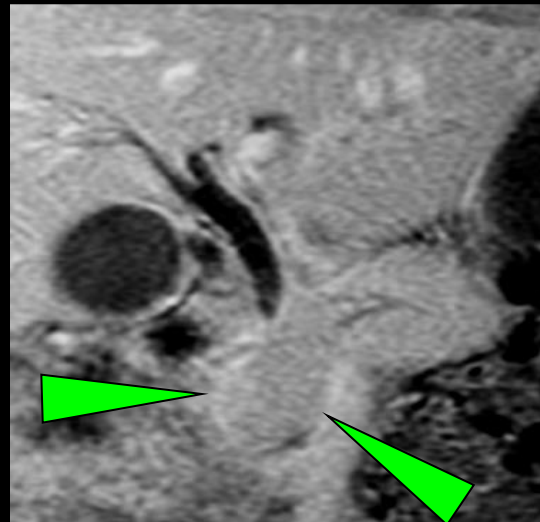
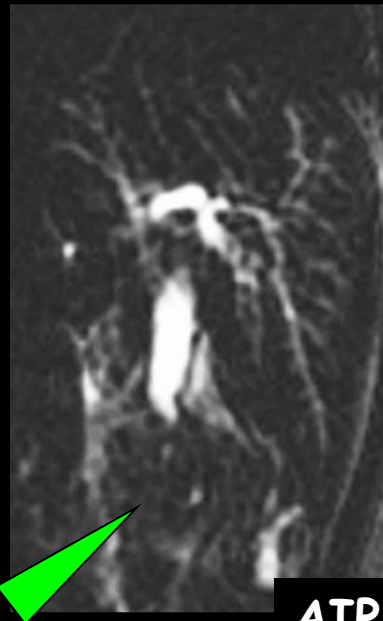
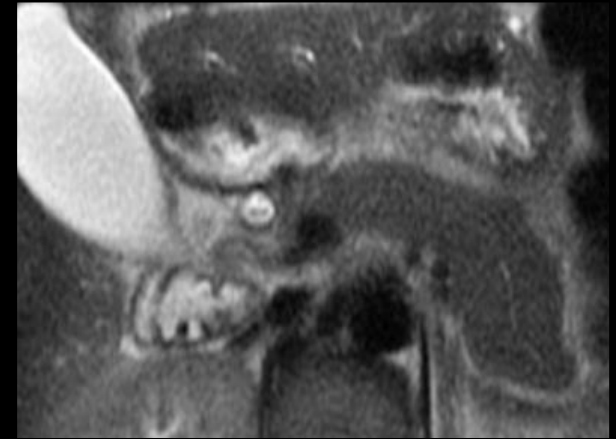
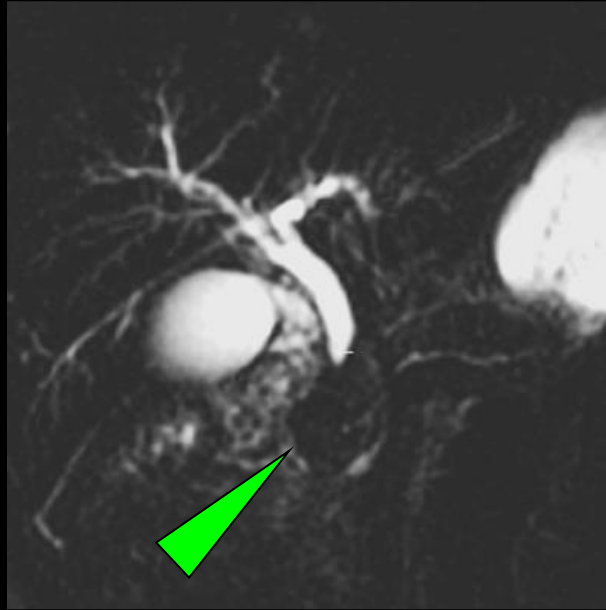
55yo man ,AIP ,diffuse homogeneous hypermetabolism with 18 FDG PET/CT can help for differential diagnosis with pancreatic adenocarcinoma



Utility of ^{18}F -FDG PET/CT for differentiation of autoimmune pancreatitis with atypical pancreatic imaging findings from pancreatic cancer

Lee TY¹, Kim MH², Park DH et al.² AJR Am J Roentgenol 2009;193:343-348

36 yo woman ,pain of the right upper quadrant with cholestasis and anorexia.

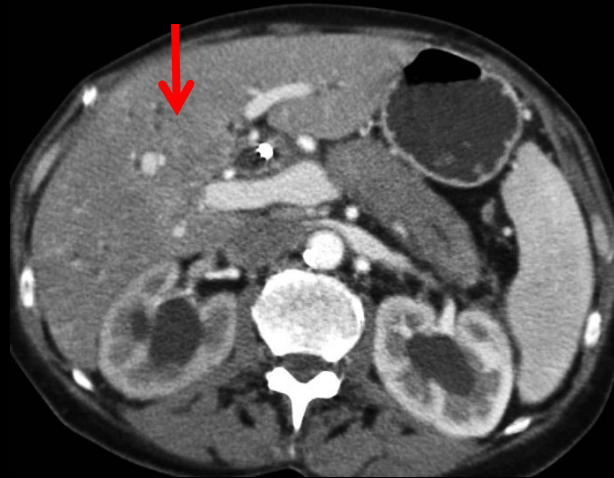


- homogeneous mass of the pancreatic head ;
- dilatation of supra pancreatic main bile duct
- no dilatation of the upstream main pancreatuc duct

AIP localised to pancreatic head

-are AIP one of the 'IgG4 related diseases" ?

63 yo woman ,abdominal pain ,cholestasis, renal insufficiency



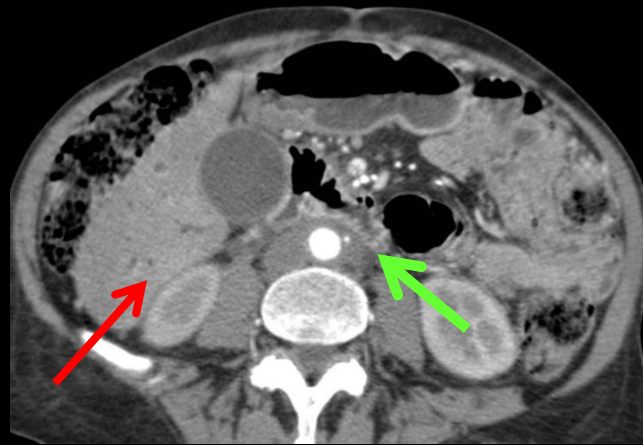
diffuse hypertrophy of the pancreas with loss of lobularity

no dilatation of the main pancreatic duct

intrahepatic bile duct dilatation (autoimmune cholangitis)



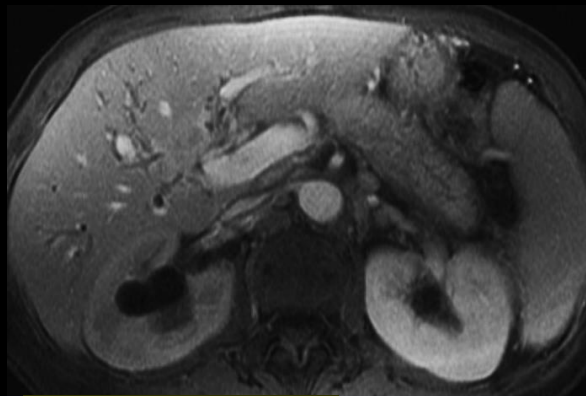
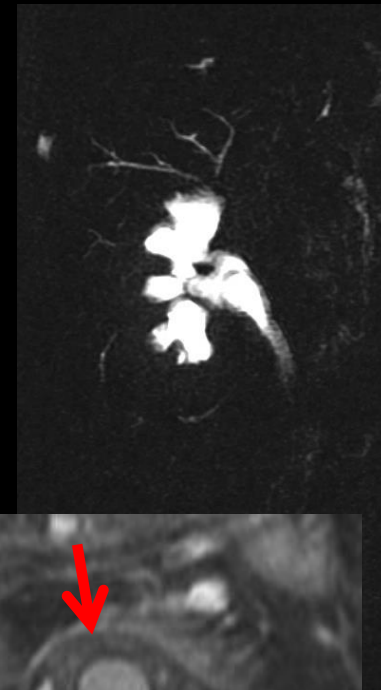
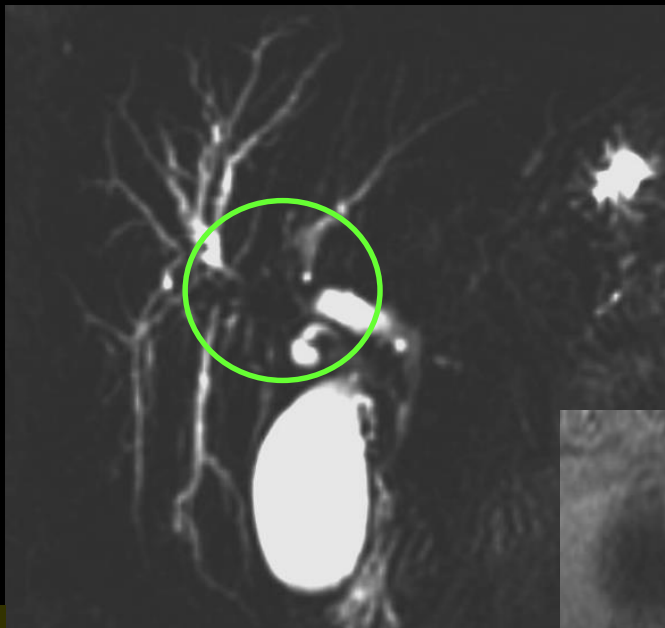
CT



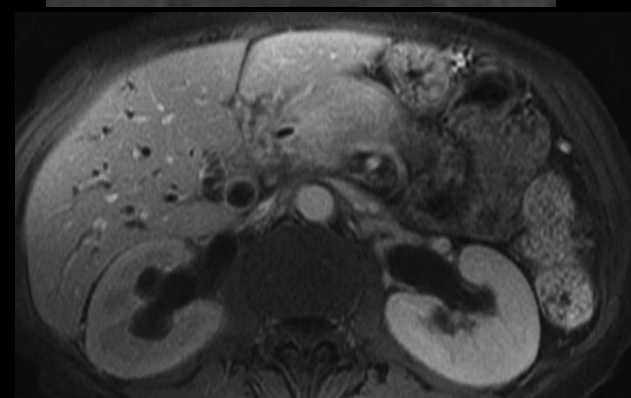
retroperitoneal fibrosis with ureterohydronephrosis

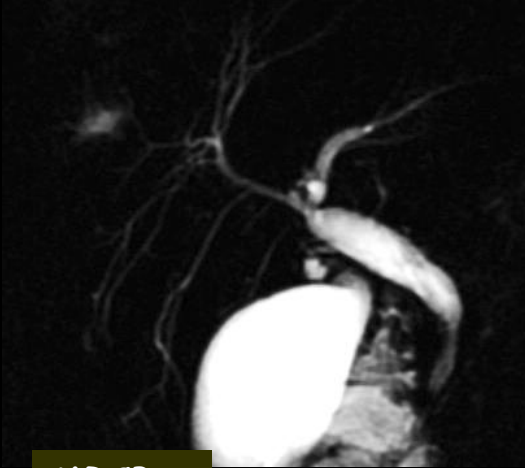


MRCP

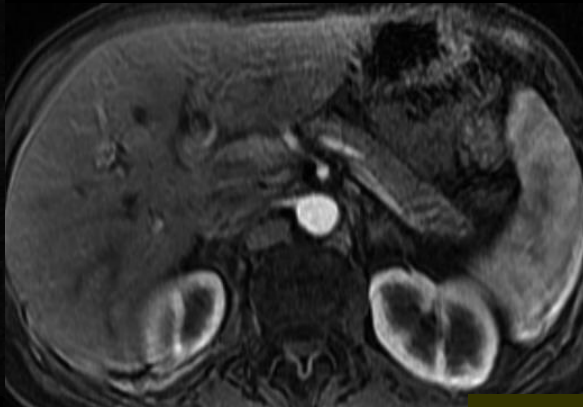


T1W gado FS





MRCP



T1W gado FS



after one month of corticosteroids

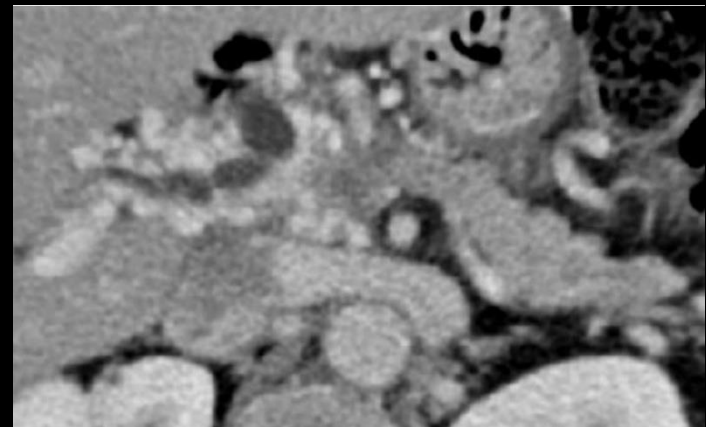
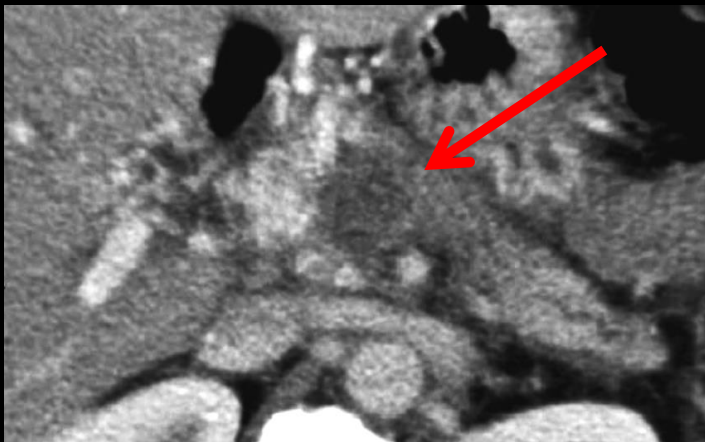


CT

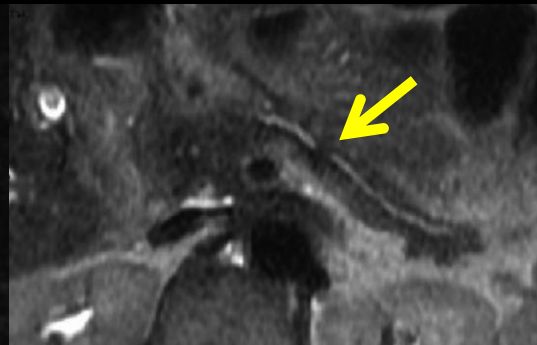


three months later

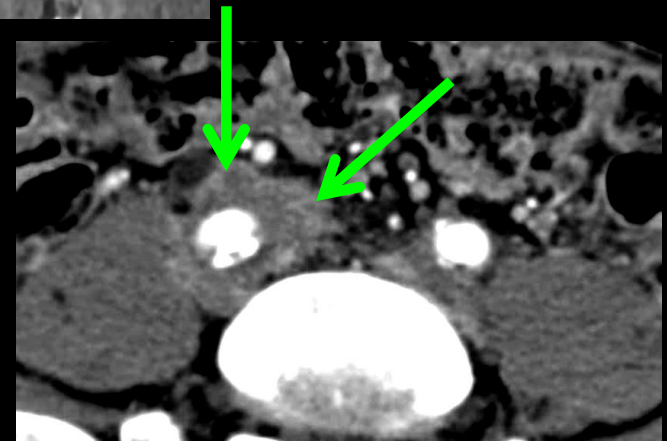
40 yo man ,intense epigastric pain with weight loss and increasing cholestasis ; low back pain.



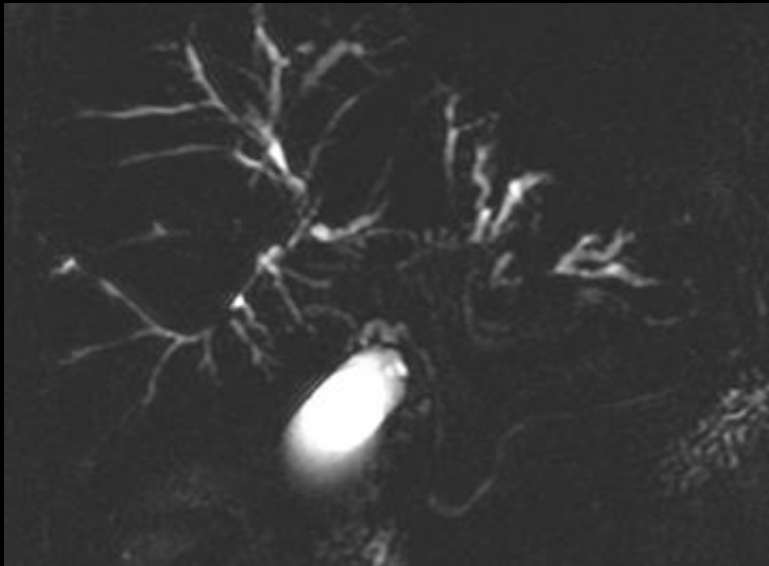
évolution after corticosteroid



AIP , iliac retroperitoneal fibrosis portal vein thrombosis with cavernoma .



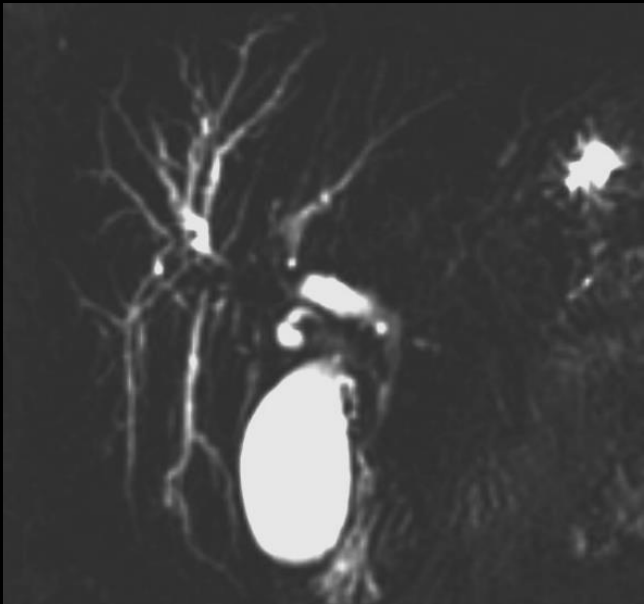
How can we recognize identify autoimmune corticosenensitive cholangitis ?



MRCP



primary sclerosing cholangitis



autoimmune cholangitis

Are serologic data useful for the diagnosis of PAI ? ?

Finkelberg DL et al. *N Engl J Med* 2006 ; 336 : 2670-6

Table 1. Diagnostic Criteria for Autoimmune Pancreatitis.*

| Findings on Imaging Radiography (One Required) | | Serologic and Histologic Findings (One Required) | | |
|---|--|--|---|--|
| Cross-Sectional Imaging | ERCP or MRCP | Serologic Analysis | Pancreatic–Biliary Histologic Analysis | Nongastrointestinal Histologic Analysis |
| Diffusely enlarged pancreas | Segmental pancreatic ductal narrowing | Elevated serum IgG4 level | Periductal lympho- plasmacytic infil- tration or fibrosis | Tubulointerstitial nephri- tis with immune de- posits within tubular basement membranes |
| Enhanced peripheral rim of hypoatten- uation “halo” | Focal pancreatic duc- tal narrowing | Elevated serum IgG or gamma globulin level | Obliterative phlebitis | Pulmonary interstitial lymphoplasmacytic infiltration with IgG4- positive plasma cells† |
| Low-attenuation mass in head of pancreas | Diffuse pancreatic ductal narrowing | Presence of ALA, ACA II, ASMA, or ANA | IgG4-positive plasma cells in tissue† | Chronic sialadenitis with IgG4-positive plasma cells† |

* Criteria were modified from those of the Japan Pancreas Society.³⁰ ERCP denotes endoscopic retrograde cholangiopancreatography, MRCP magnetic resonance cholangiopancreatography, ALA antilactoferrin antibody, ACA II anti-carbonic anhydrase II antibody, ASMA anti-smooth-muscle antibody, and ANA antinuclear antibody.

† The presence of tissue IgG4-positive cells is not necessarily abnormal, but an increased number of infiltrating IgG4-positive plasma cells is abnormal.

Is there an interest of ^{18}F FDG PET/CT for the diagnosis of PAI ??



MDCT

66 yo woman AIP ,metabolic hyperactivity in caudal pancreas, thyroid and salivary glands . These extrapancreatic sites of ^{18}F FDG are not seen in pancreatic adenocarcinoma

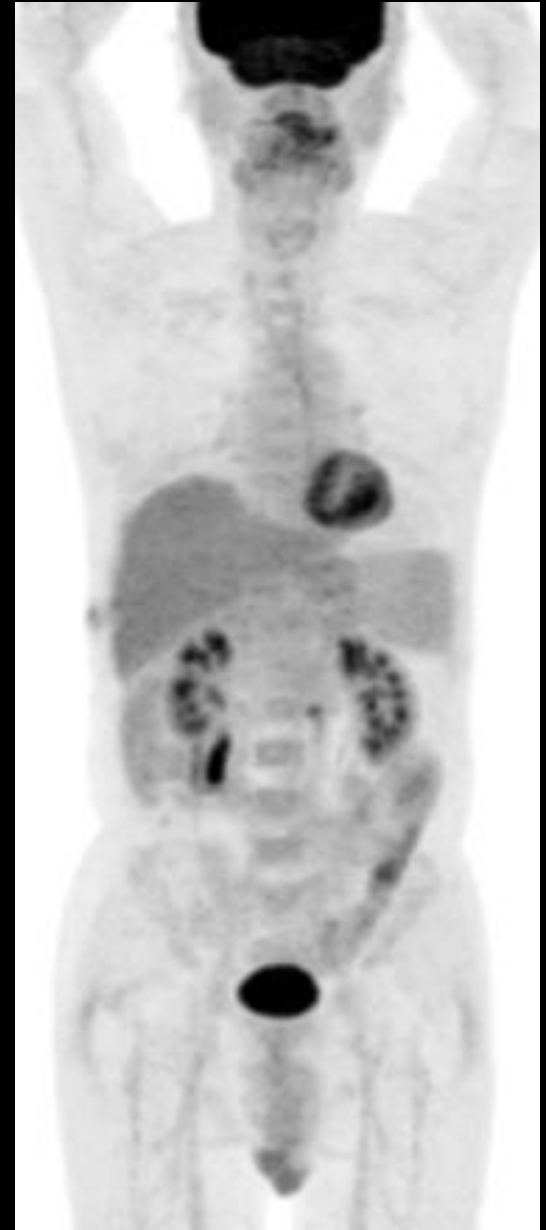
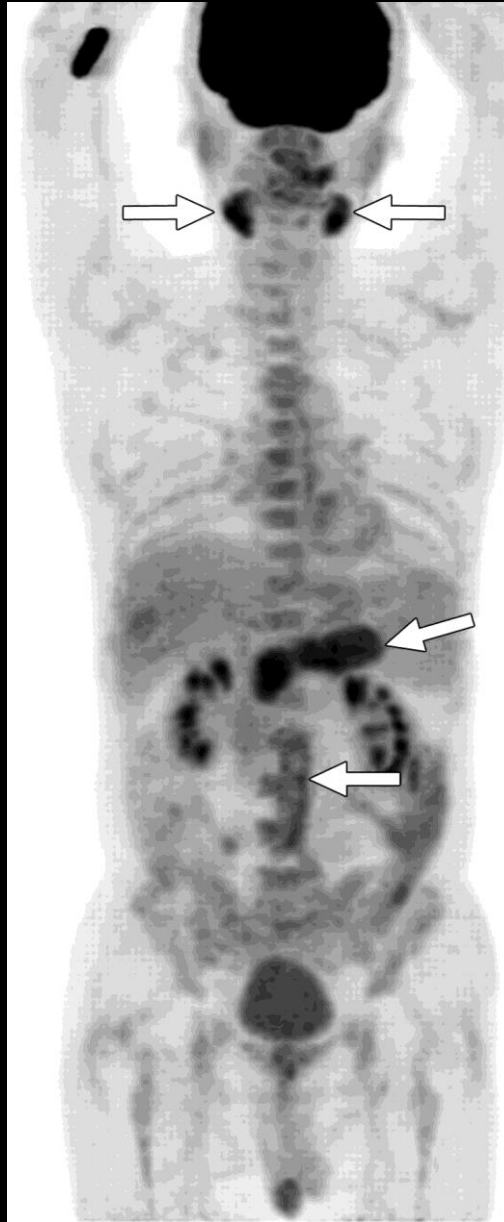


PET ^{18}F FDG

Utility of ^{18}F -FDG PET/CT for differentiation of autoimmune pancreatitis with atypical pancreatic imaging findings from pancreatic cancer

Lee TY¹, Kim MH², Park DH et al.² AJR Am J Roentgenol 2009;193:343-348

71 yo man ,PAI ,sialadénitis and retroperitoneal fibrosis with uretrohydronephrosis

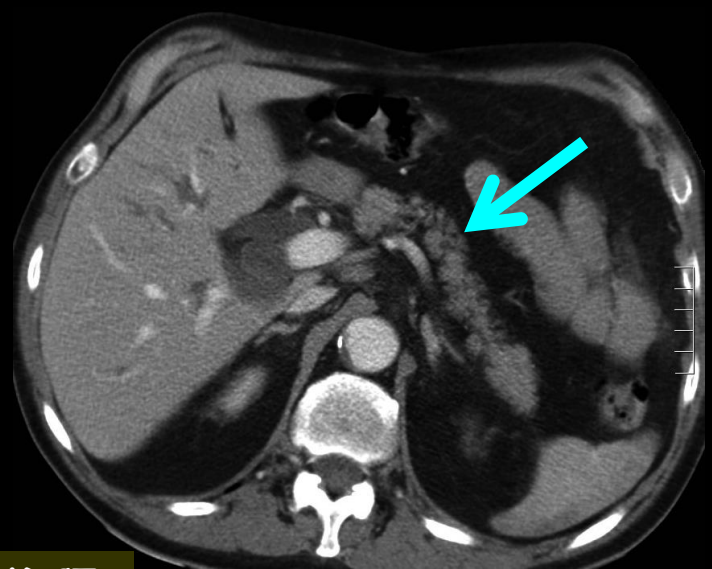


Utility of ^{18}F -FDG PET/CT for differentiation of autoimmune pancreatitis with atypical pancreatic imaging findings from pancreatic cancer

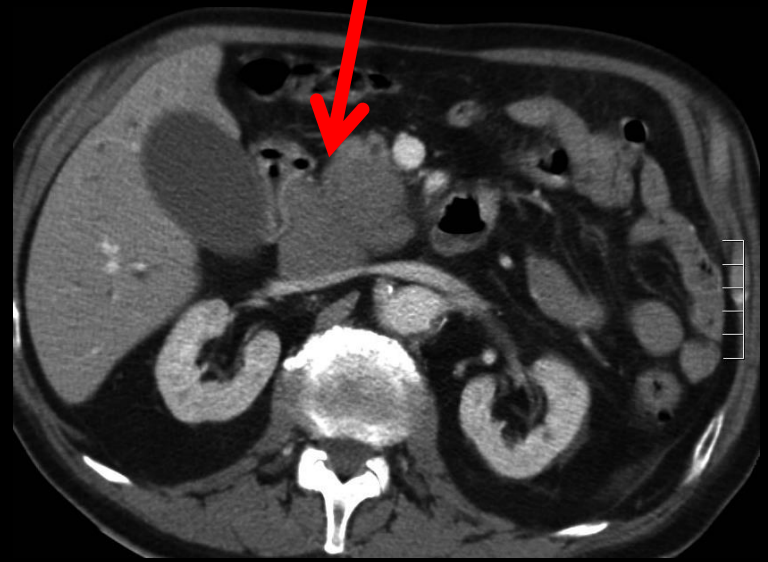
Lee TY¹, Kim MH², Park DH et al.² AJR Am J Roentgenol 2009;193:343-348

One exemple of differential diagnosis of AIP

80 yo man ,weight loss,cholestasis ; it's not an AIP nor an adenocarcinoma . What is it ? ? ?



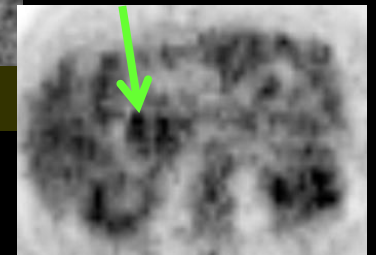
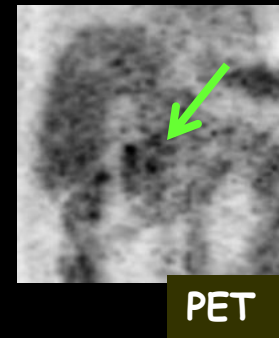
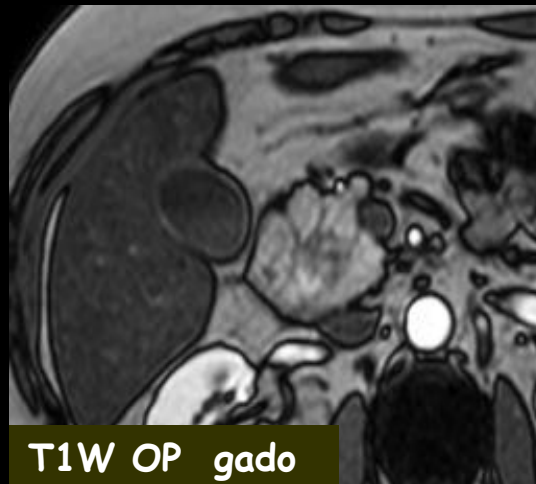
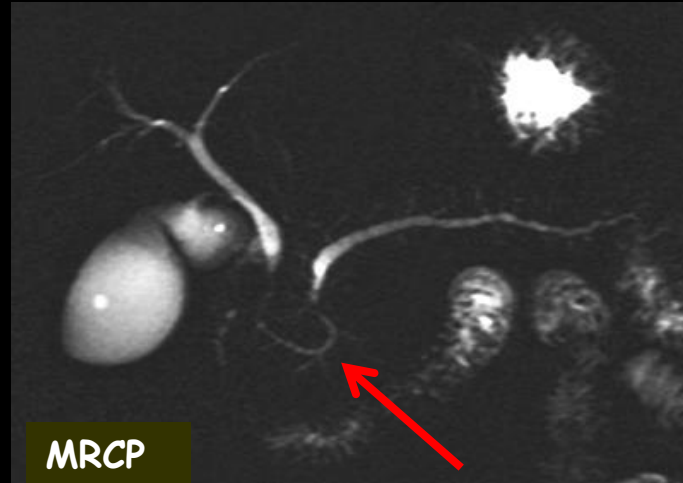
MDCT



High grade NHL with pancreatic localisation

2b. Inflammatory pseudotumor of the pancreas

56 yo man, alcoholic and smoker, clinical presentation of mild pancreatitis



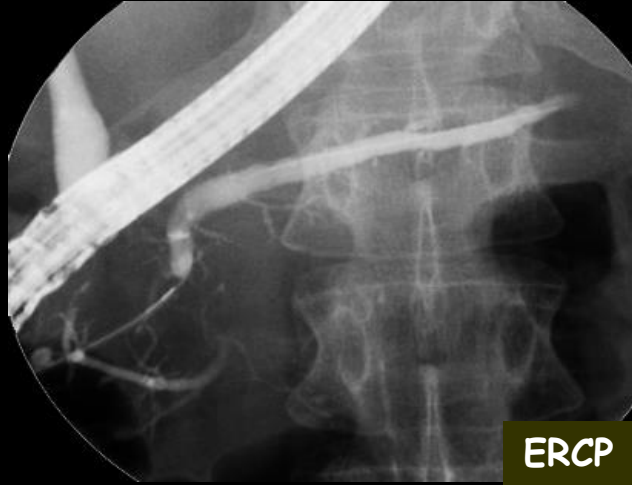
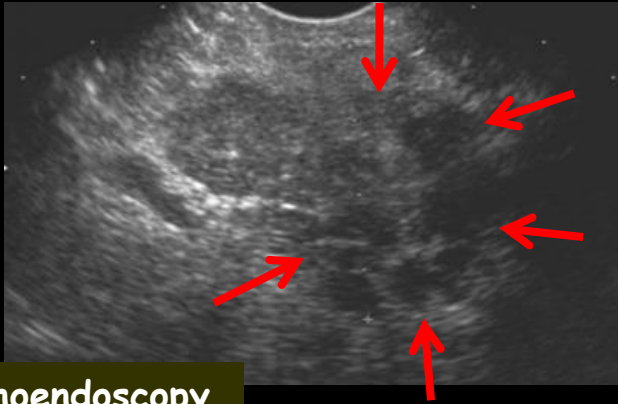
Obs Pr .B Dupas Nantes

2 échoendoscopique guided biopsies with
cytologic analysis : no suspect cell;

new imaging investigations after 3 months

CA 19-9 136,8 U/ml

échoendoscopy
e

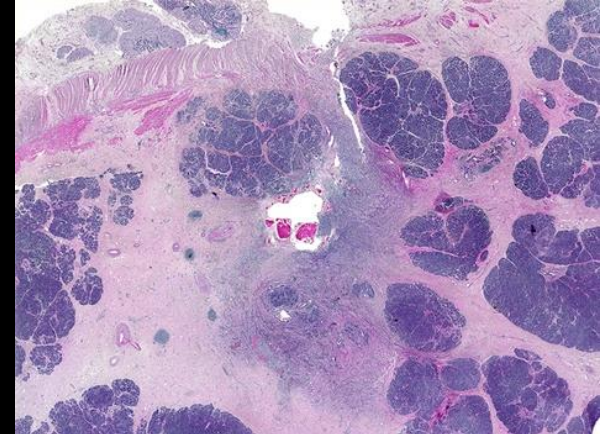
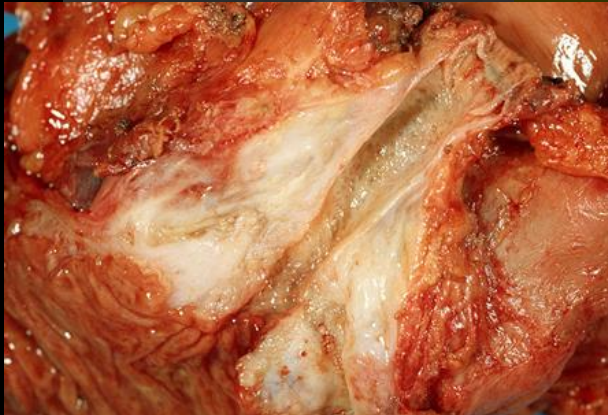


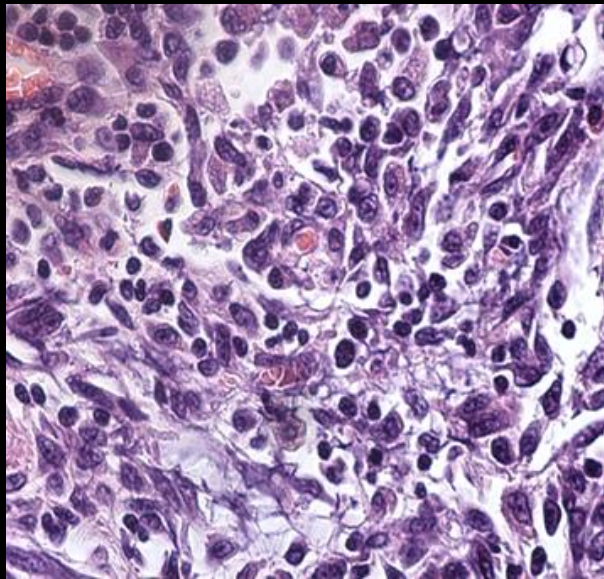
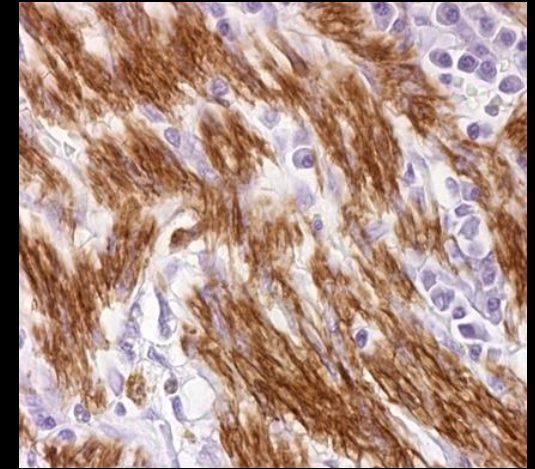
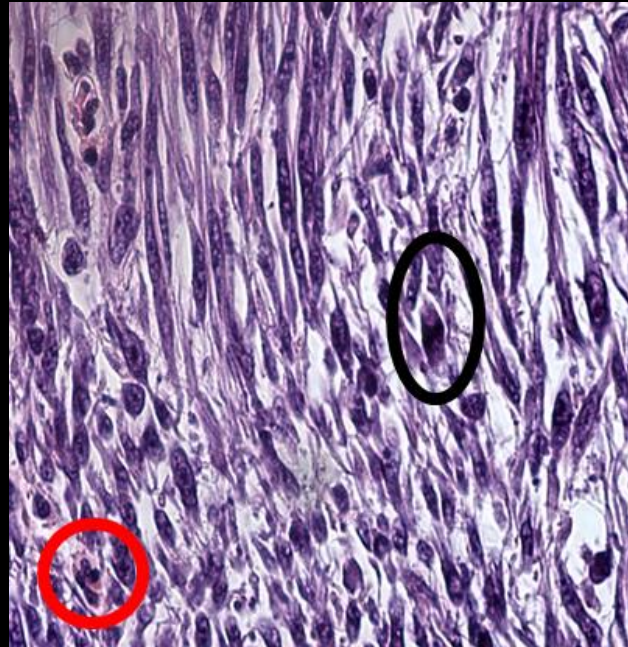
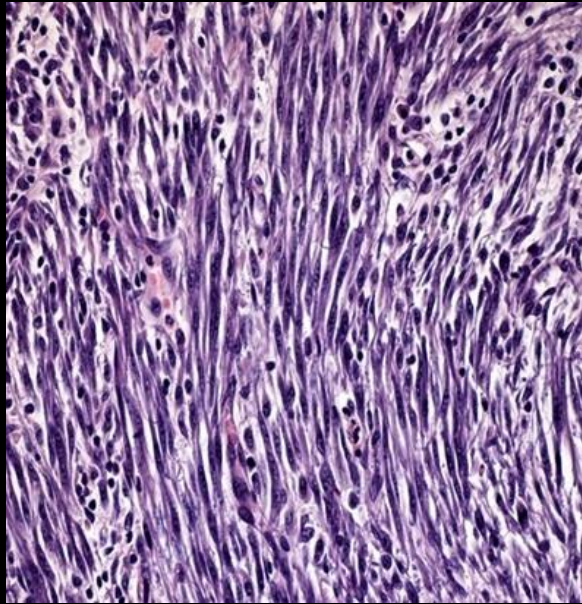
ERCP

Obs Pr .B Dupas Nantes

MRCP

Surgery is decided : Whipple intervention



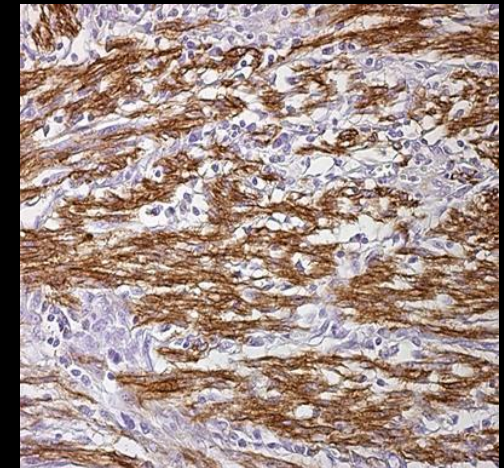


inflammatory pseudotumor or
inflammatory myofibroblastic
tumor

cellular fusiform proliferation
with variable degrees of
atypia

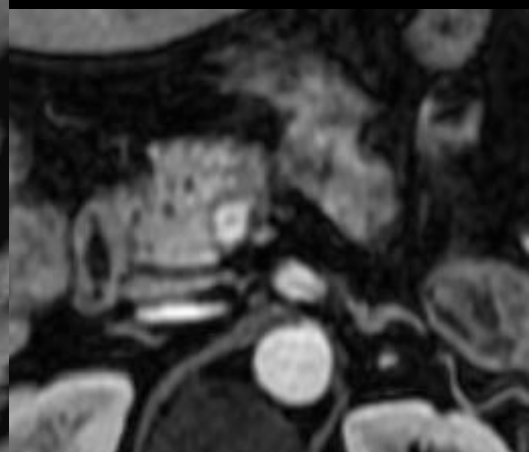
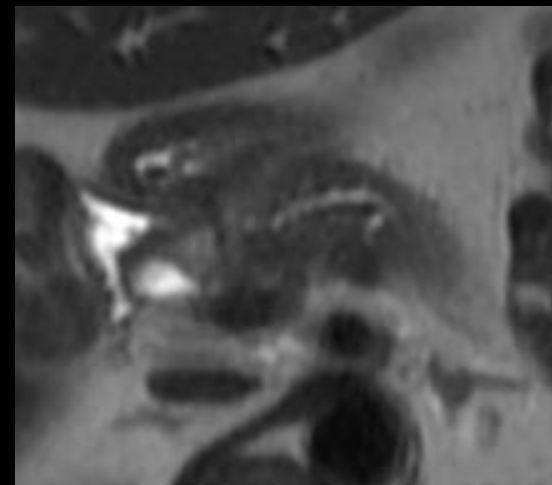
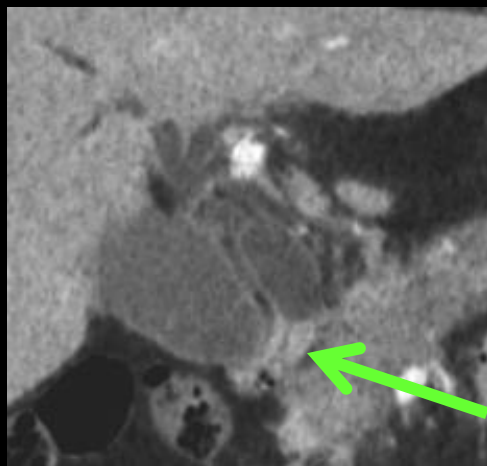
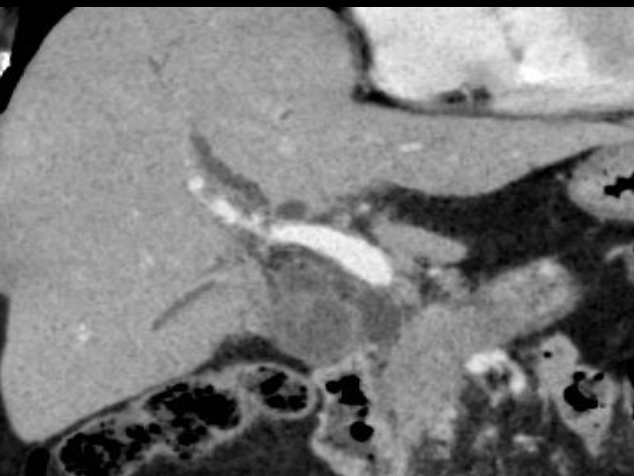
lymphoplasmocytic infiltration

myxoid stroma



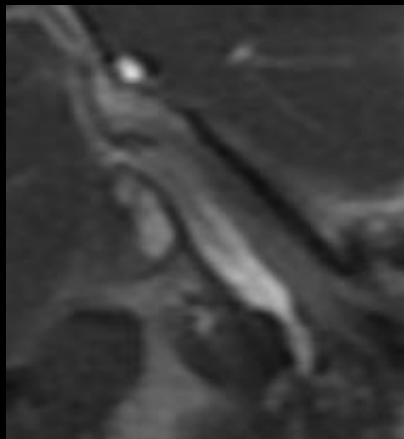
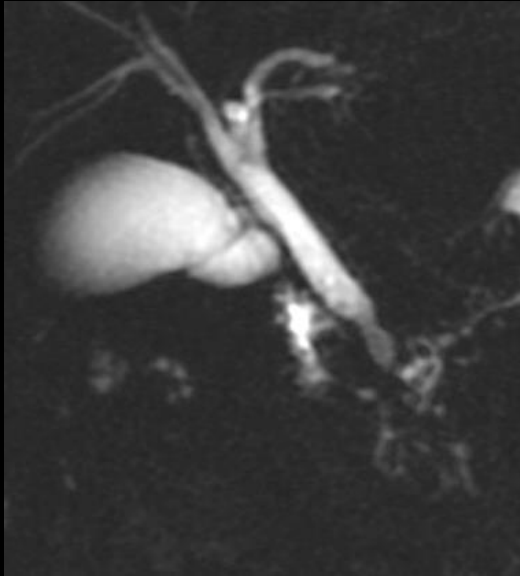
immunological staining :
smooth muscle actin
specific +++

71 yo woman , ictèrus , seric bilirubin :180 mg/L.

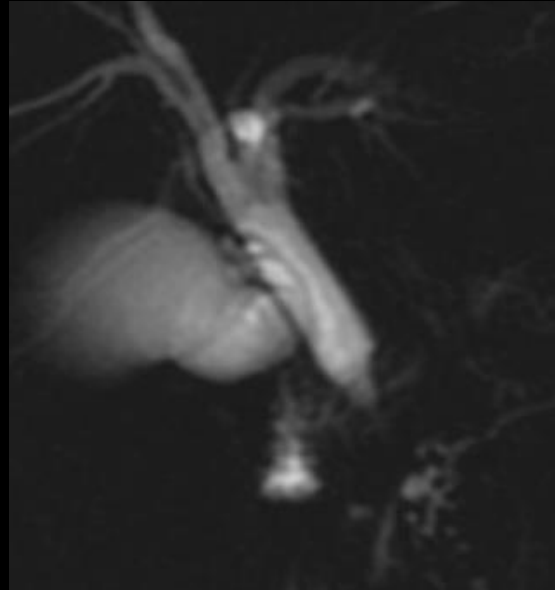


Fluctuating evolution of cholestasis and clinical jaundice

Follow up 3 months



Follow up 6 months



intervention (Whipple)

inflammatory pancreato-
cholangitis (fibro-hyaline
transformation and
lymphoplasmocytic
infiltration)

Au total

Inflammatory pancreatobiliary "tumor-like" lesions are a **challenge for radiologist and surgeons**

It is often impossible to get histological data despite progress of endoscopic and radiologic guided biopsies and decisions must be based on MDCT and MR images

Biology can contribute to diagnosis when Ig G4 are elevated but this is quite rare in west europa

Clinicians have to be very attentive to **atypies** in clinical or biological evolution

It is often wise **to wait some weeks before surgery (3 months for example)** in equivocal cases and a corticosteroid test can be useful to avoid unnecessary mutilating surgery .

