

# Case 5

46M 呂X訓

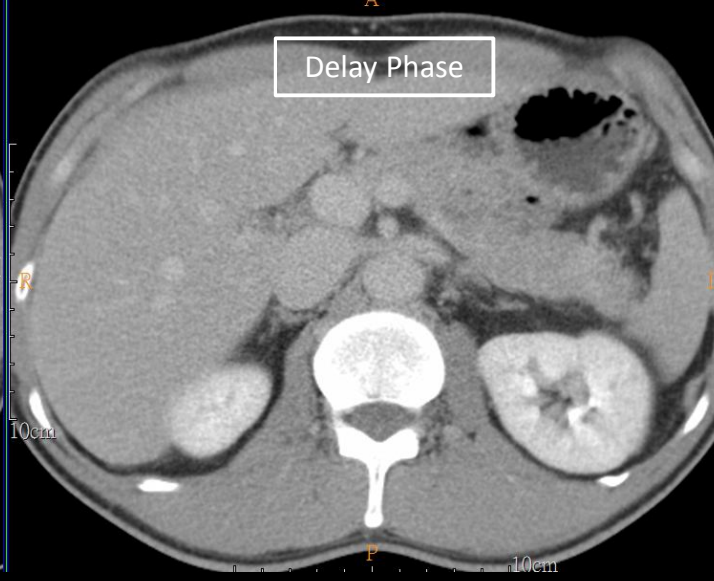
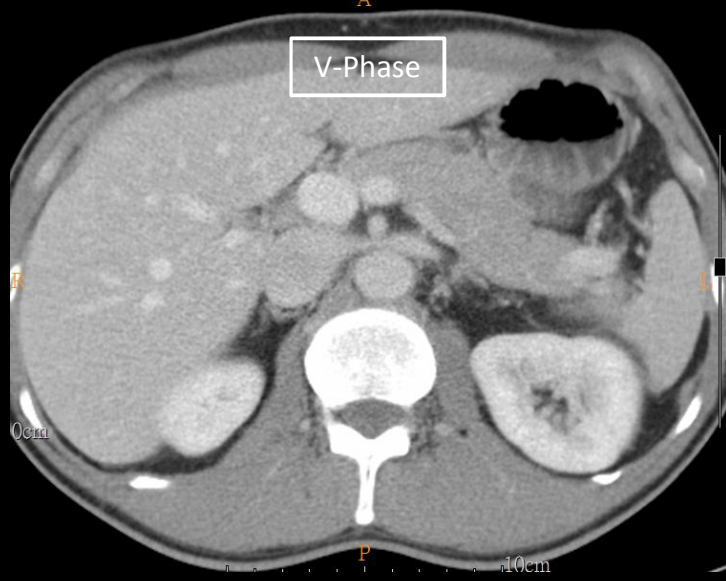
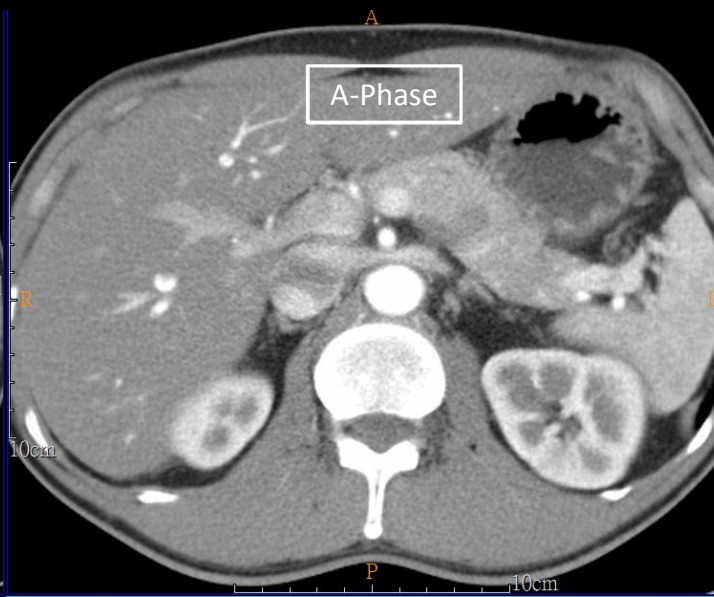
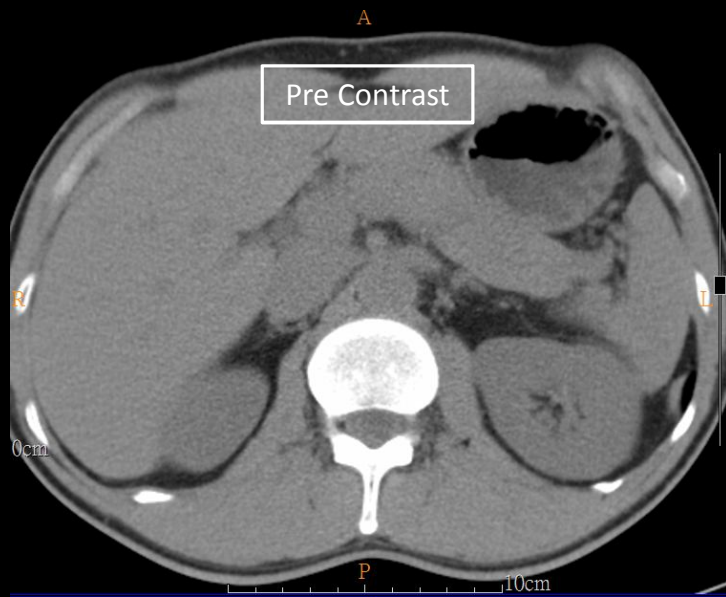
Neck lymph nodes

Abdomen echo: 1.5cm tumor at body and 1.3cm at tail of pancreas ??

# Images

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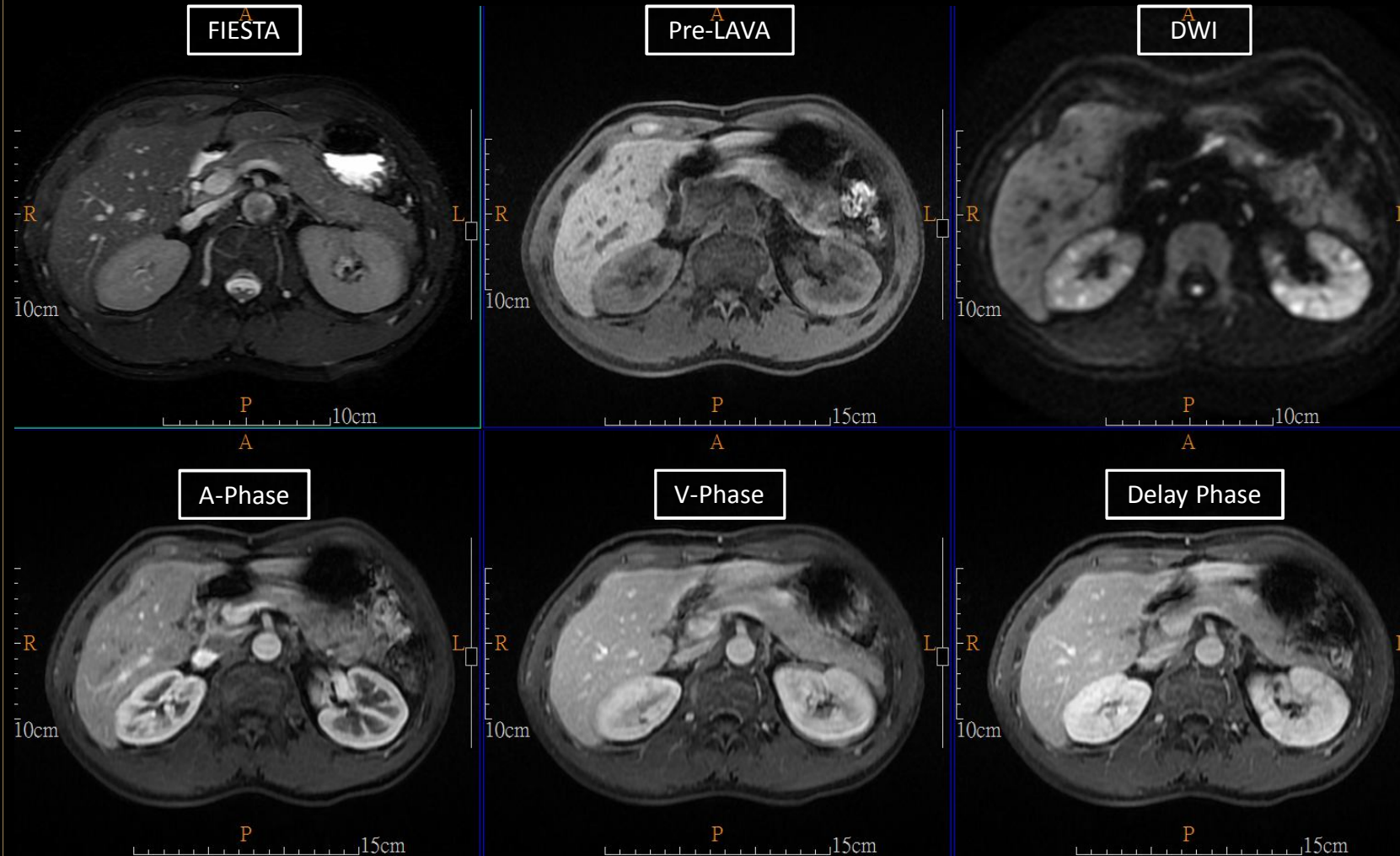
- 2019-05-31 Pancreas CT
- 2019-06-11 Pancreas MRI



Swelling and loss of normal lobulation of pancreas

An ill-define hypo-enhanced focus at pancreatic body on arterial phase

Isodense on other phase

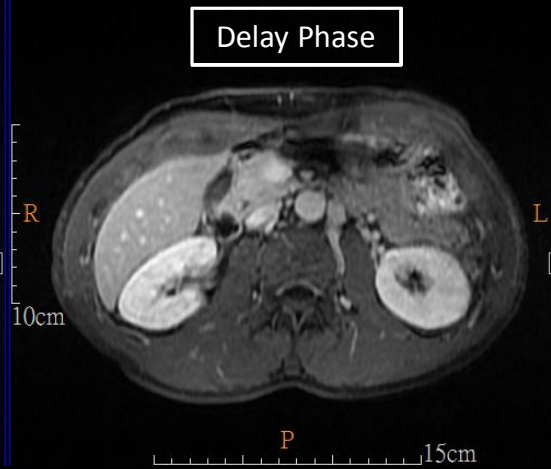
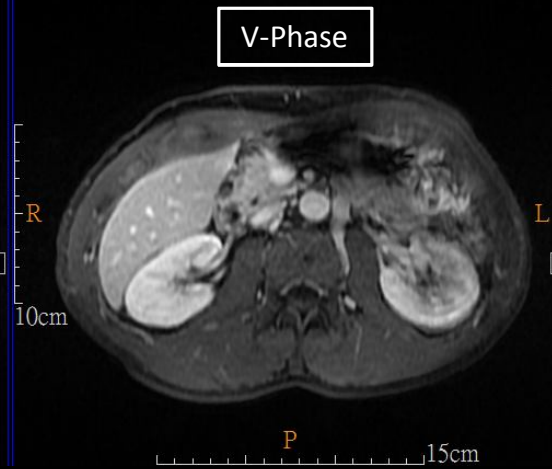
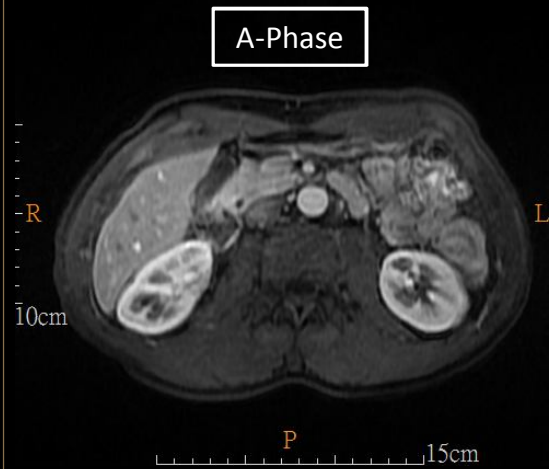
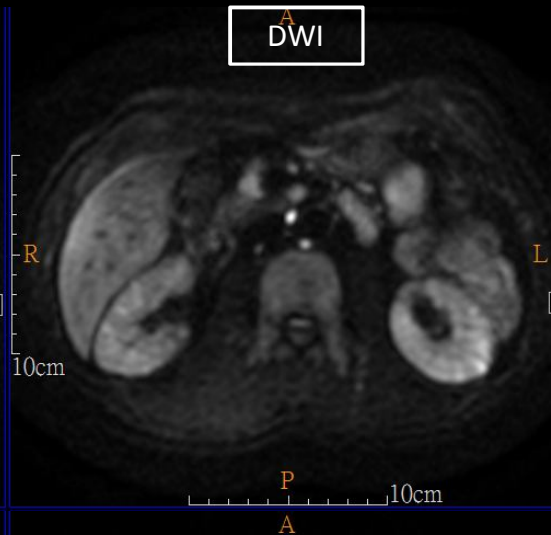
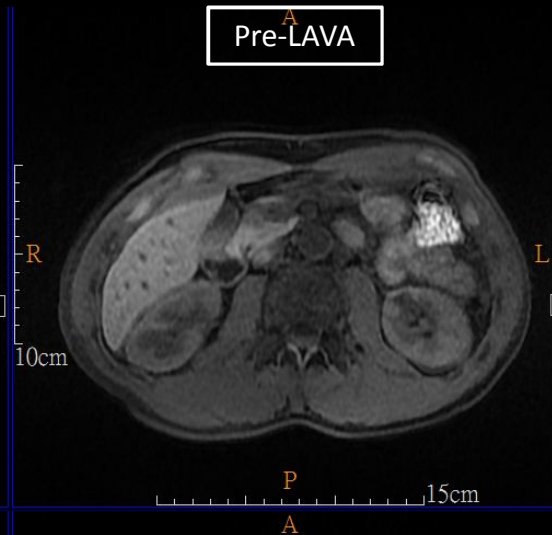
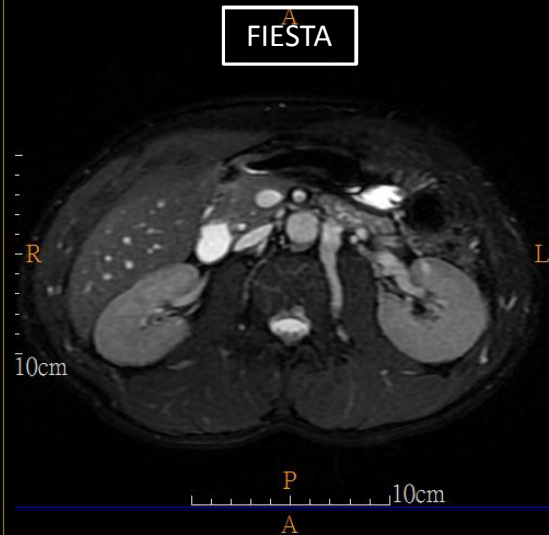


Swelling and loss of normal lobulation of pancreas

Multifoci area with high DWI signal

Hypoenhancement in early phase and moderate delayed enhancement

C/W autoimmune pancreatitis



Multifoci tiny  
hypo-enhanced lesions at  
bilateral renal parenchyma

IgG4 related disease

# Clinical Course

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- Lab data
  - RF: 18.7 IU/mL [ $<14.0$ ]
  - IgG: 1709 mg/dl [700-1600]
  - IgG4: 1640 mg/dL [3-201]
  
  - ANA: negative
  - Anti-Ro: negative
  - Anti-La: negative

# Discussion

IgG4 Related Disease



# IgG4-related Disease from Head to Toe<sup>1</sup>

*Radiographics*. 2015 Nov-Dec;35(7):2007-25.

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**Abbreviations:** CBD = common bile duct, ERCP = endoscopic retrograde cholangiopancreatography, FDG = fluorodeoxyglucose, H-E = hematoxylin-eosin, IgG4 = immunoglobulin G4

**RadioGraphics 2015;** 35:2007–2025

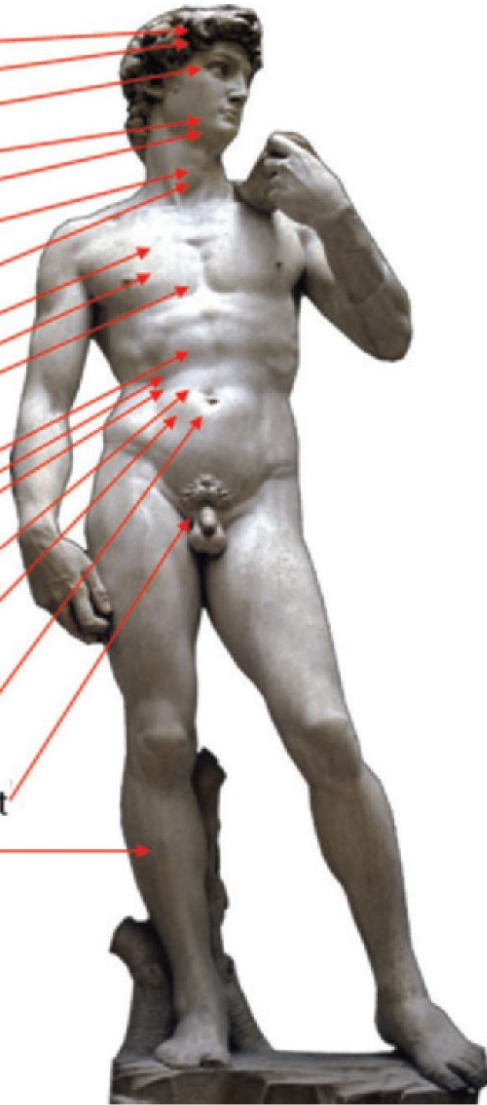
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**Content Codes:** CH GI HN

Immunoglobulin G4 (IgG4)–related disease is a relatively recently proposed clinical-pathologic entity that is characterized by fibro-inflammatory lesions rich in IgG4-positive plasma cells and, often but not always, elevated serum IgG4 concentrations. IgG4-related disease was recognized as a systemic disease in 2003, when extra-pancreatic manifestations were identified in patients with autoimmune pancreatitis. Since then, the disease has been reported as affecting virtually every organ system and has been identified in the biliary tree, salivary and lacrimal glands, periorbital tissues, lungs, lymph nodes, thyroid gland, kidneys, prostate gland, testicles, breasts, and pituitary gland. Its pathogenesis is poorly understood, but findings are consistent with both an autoimmune and an allergic disorder. Although definitive diagnosis requires histopathologic analysis, imaging plays an important role in demonstrating infiltration and enlargement of involved organs. Because of the systemic

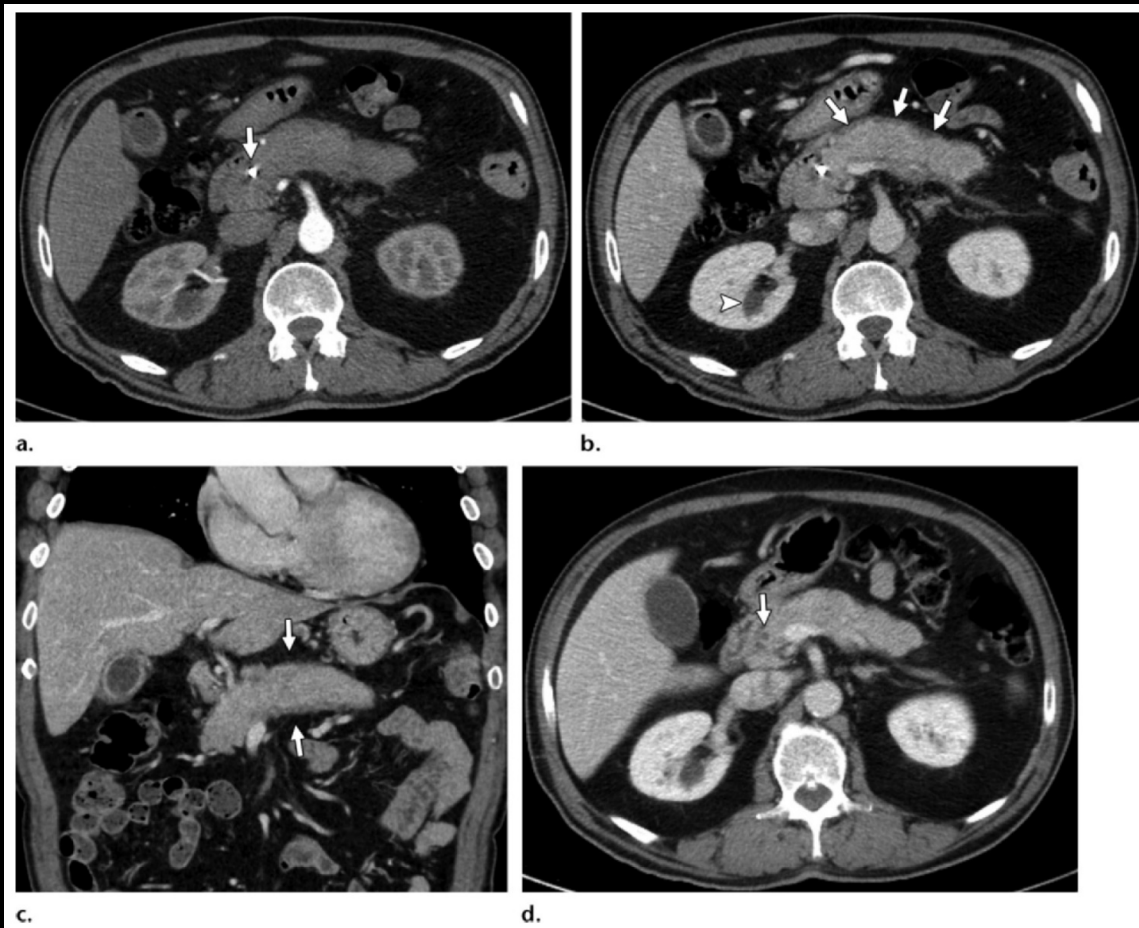


- Hypertrophic pachymeningitis
- Autoimmune hypophysitis
- Orbital pseudotumor
- Mikulicz's disease
- Kuttner's tumor
- Riedel's thyroiditis
- Hashimoto's thyroiditis
- Pulmonary pseudotumor
- Interstitial pneumonia
- Lymphadenopathy
- Autoimmune pancreatitis
- Sclerosing cholangitis
- Liver pseudotumor
- Retroperitoneal fibrosis
- Tubulointerstitial nephritis
- Inflammatory aortic aneurysm
- Prostatitis, testicular involvement
- Cutaneous pseudolymphoma



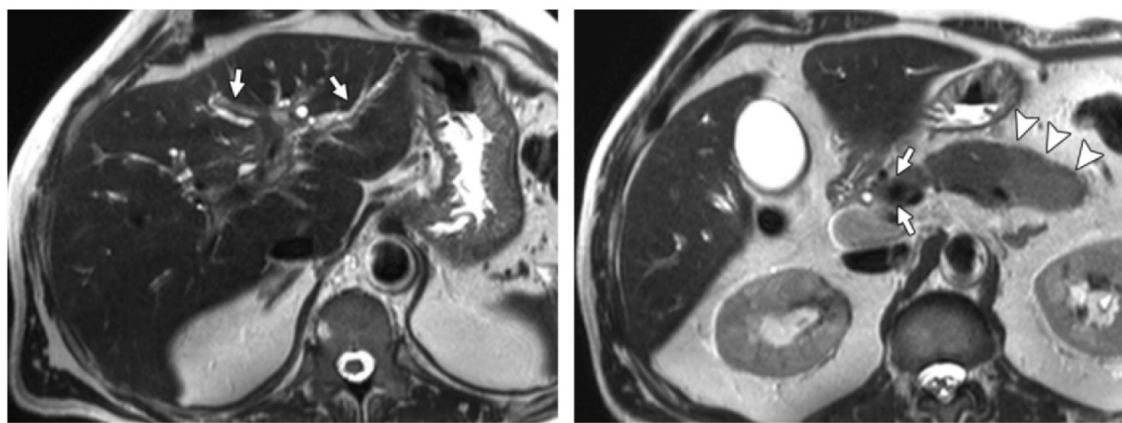
- Autoimmune Pancreatitis
- Sclerosing Cholangitis
- Salivary and Lacrimal Gland Involvement
- Thyroiditis
- Renal Disease
- Retroperitoneal Fibrosis
- Sclerosing Mesenteritis

# Autoimmune Pancreatitis



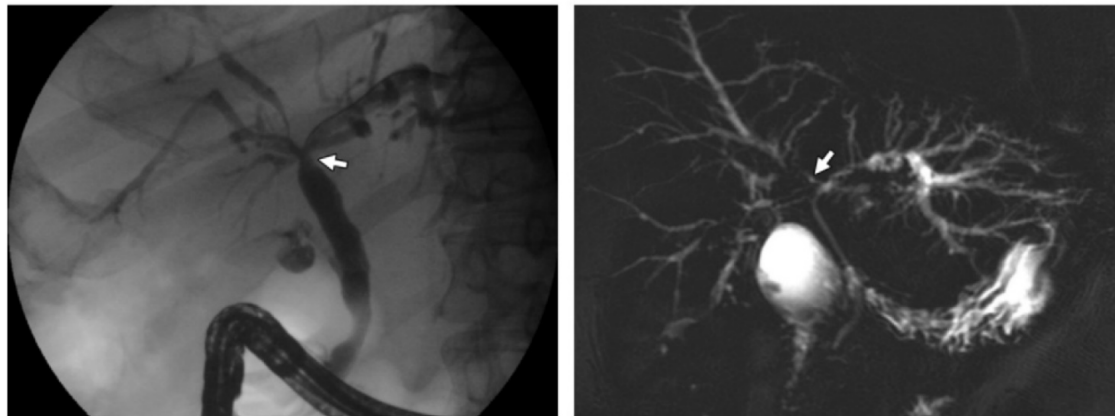
- Loss of lobular contours (sausage like appearance)
- Narrowing of the CBD, with endoscopic stent placement
- Hypoattenuating halo around the pancreatic body and tail
- Absence of peripancreatic stranding
- (D) Follow-up CT image obtained 4 months later after steroid administration shows a marked response to treatment, with resolution of the hypoattenuating peripancreatic halo

# Sclerosing Cholangitis



a.

b.

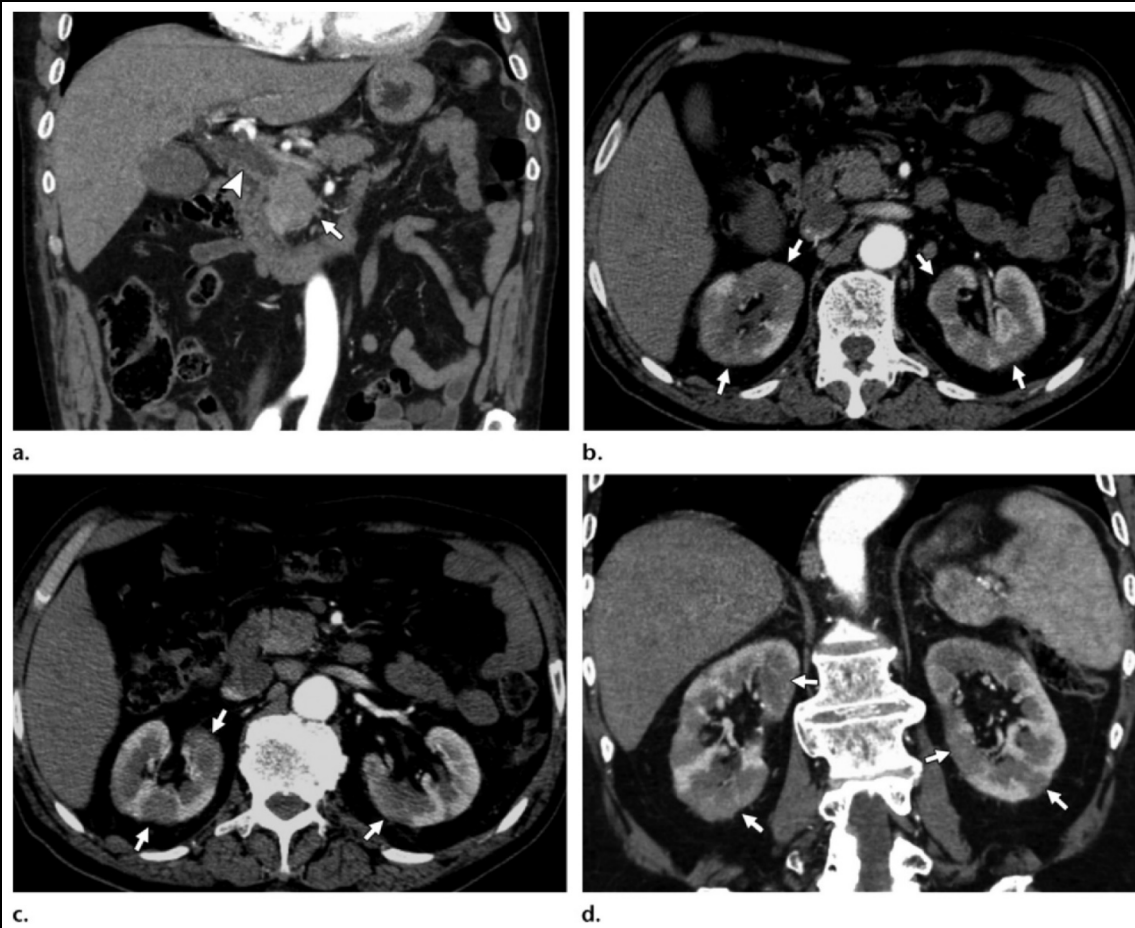


c.

d.

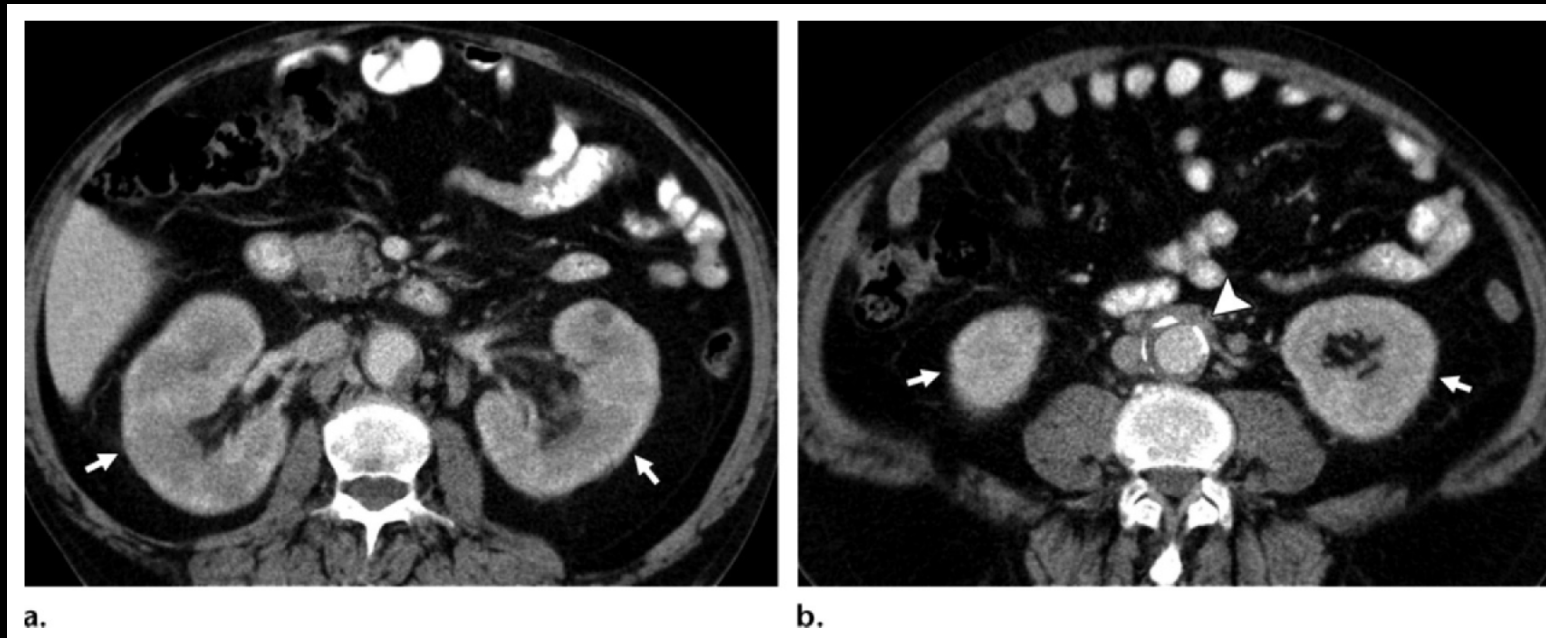
- (A) Diffuse dilatation of the intrahepatic biliary ducts
- (B) Mild wall thickening of the CBD. Autoimmune pancreatitis
- (C) Multiple strictures in the intrahepatic bile ducts with prestenotic dilatation. CHD stricture.
- (D) 1 year later after steroid treatment shows persistent narrowing of the intrahepatic bile ducts but resolution of the CBD dilatation

# Renal Disease (Multifocal Type)



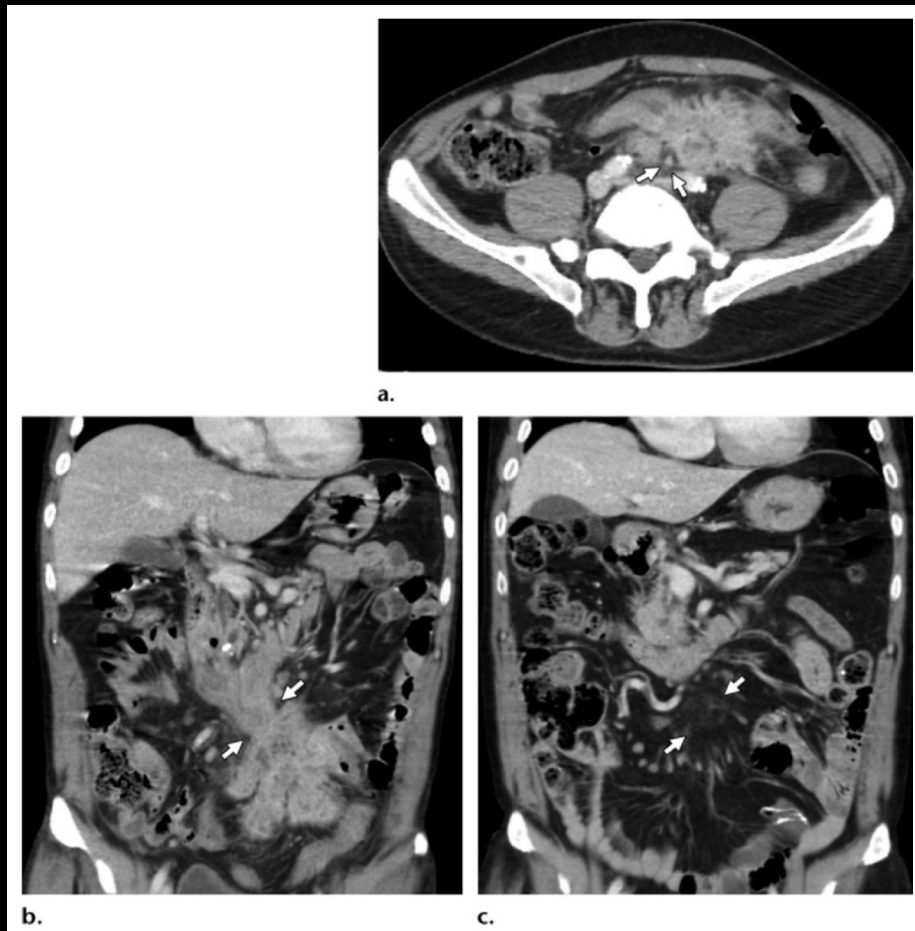
- (A) Autoimmune pancreatitis with extrahepatic bile duct dilatation
- (B-D) Multiple **well-defined, wedge-shaped, low-attenuation lesions** in both kidneys. Histopathologic analysis demonstrated **tubulointerstitial nephritis**

# Renal Disease (Diffuse Type)



- (A) Enlargement of both kidneys produced by diffuse low-attenuation areas
- (B) IgG4-related retroperitoneal fibrosis

# Sclerosing Mesenteritis



- (A) Ill-defined soft-tissue mass in the mesentery, with a preserved halo of fat around the superior mesenteric artery (fat ring sign)
- (B) With infiltration and retraction of the duodenum
- (C) 9 months later after steroid therapy shows complete resolution of the mass

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# Thanks for your attention

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