



中华医学会核医学分会
技术与继续教育学组

中华医学会核医学分会第十一届委员会 技术与继续教育学组

2019年年会继续教育分会场MDT病例展示

*PET/CT*显像一例

Case Report

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病史和查体、检验

- 男，68岁，反复上腹疼痛2年余，发现肌酐升高9月
- 患者反复出现上腹疼痛，伴呕吐，无排气排便，当地医院查肌酐持续性升高，近9月肌酐波动较大（274-410 $\mu\text{mol/L}$ ）。当地医院予开同、尿毒清颗粒、百令胶囊口服，复测肌酐持续升高。可疑肠梗阻。
- 胃癌根治术后，病理提示：胃部浸润溃疡型中分化管状腺癌，侵及粘膜肌层。

病史和查体、检验

	项目	结果	参考范围
血常规	WBC	3.4 ↓	3.5-9.5 x10 ⁹ /L
	RBC	3.3 ↓	4.3-5.8 x10 ¹² /L
	Hb	101 ↓	130-175 g/L
	PLT	116 ↓	125-350 x10 ⁹ /L
生化	白蛋白	34.3 ↓	40-55 g/L
	肌酐	282.3 ↑	44-133 umol/L
	尿素	397 ↑	150-420 umol/L
	ALT、AST、尿酸 (-)		
血沉	ESR	52 ↑	0-15 mm/h
hs-CRP	Hs-CRP	4.2 ↑	0-3 mg/L
狼疮抗凝物	(-)		

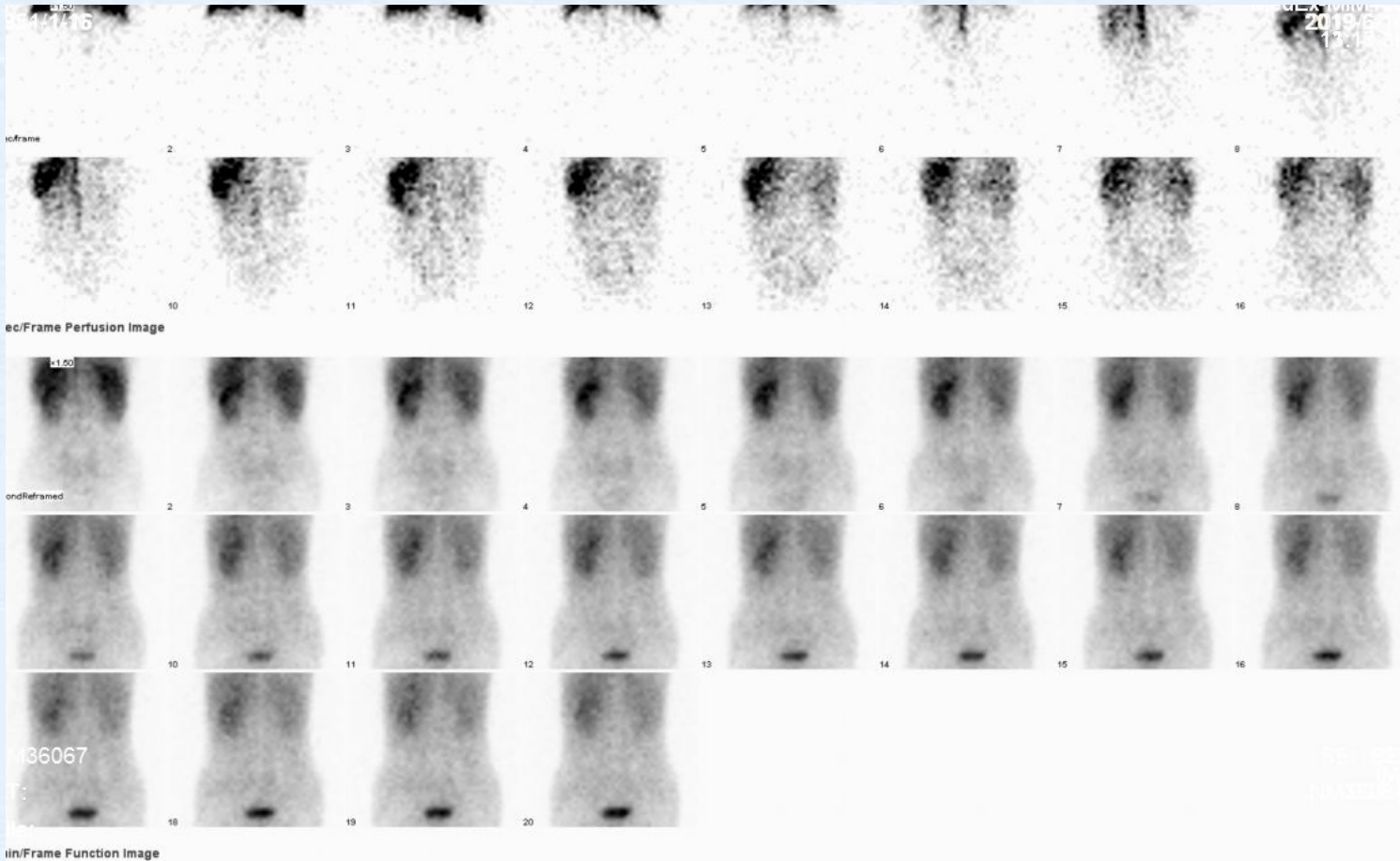
病史和查体、检验

	项目	结果	参考范围
IL-6	IL-6	7.49 ↑	<6.4 pg/ml
补体	C3、C4 (-)		
淀粉酶	淀粉酶	219 ↑	35-135 IU/L
脂肪酶	脂肪酶	20	<60 U/L
乳酸脱氢酶	LDH	152	100-240 IU/L
甲功五项	FT3	2.99 ↓	3.5-6.5 pmol/L
	余 (-)		
肿瘤标志物	细胞角蛋白19片段	3.71 ↑	<3.3 ng/ml
	胃泌素释放肽前体	80.82 ↑	<69.2 pg/ml
	余肿瘤标志物 (-)		

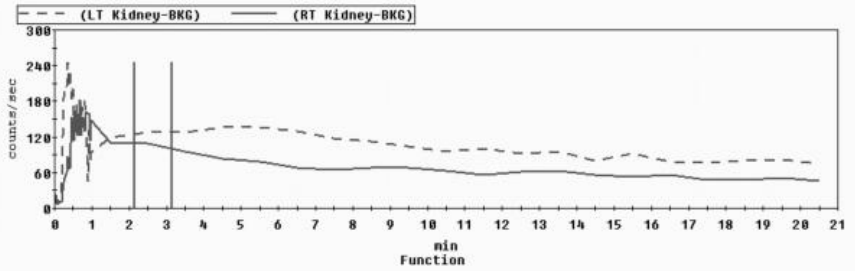
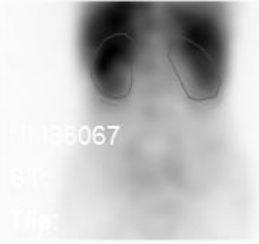
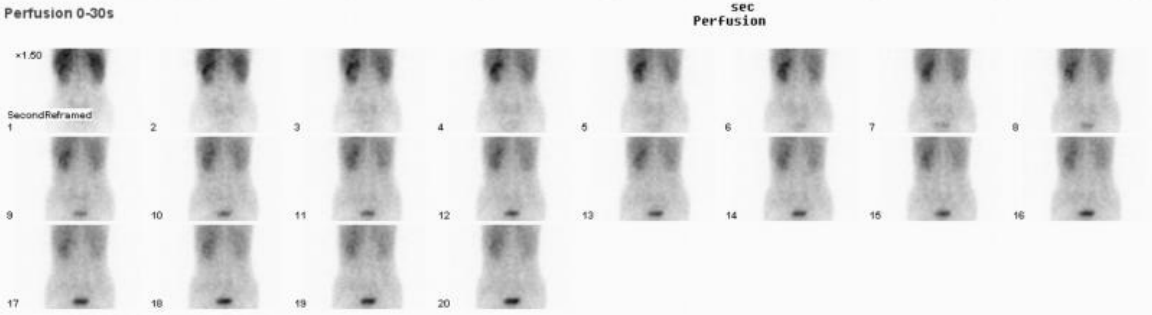
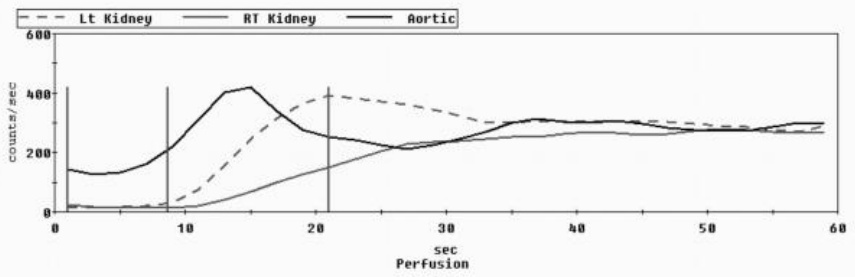
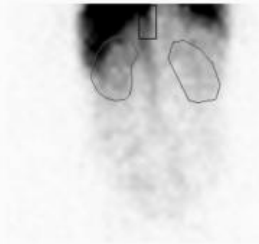
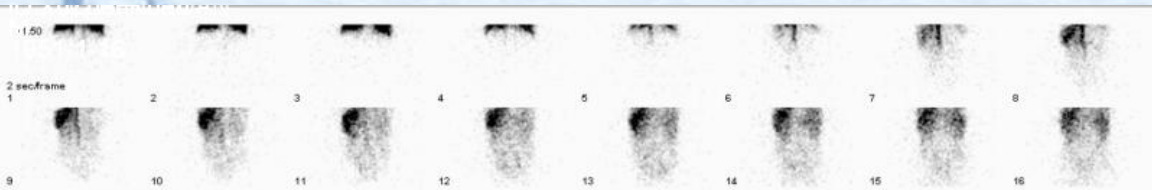
超声

- 双肾动、静脉未见狭窄，左肾副肾动脉形成
- 肠系膜上动脉：（-）
- 胆囊肿大，淤胆

肾动态显像



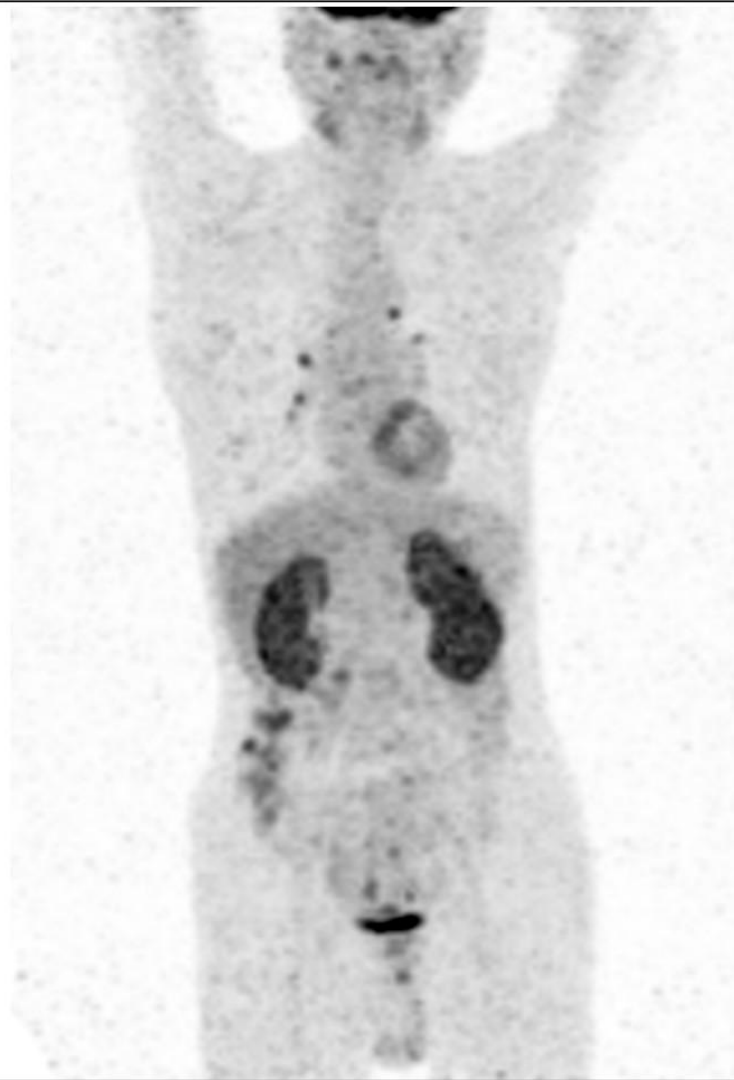
肾动态显像



GFR (ml/min) :

左肾 18 右肾 15 总肾 33

(总肾参考值≤68)



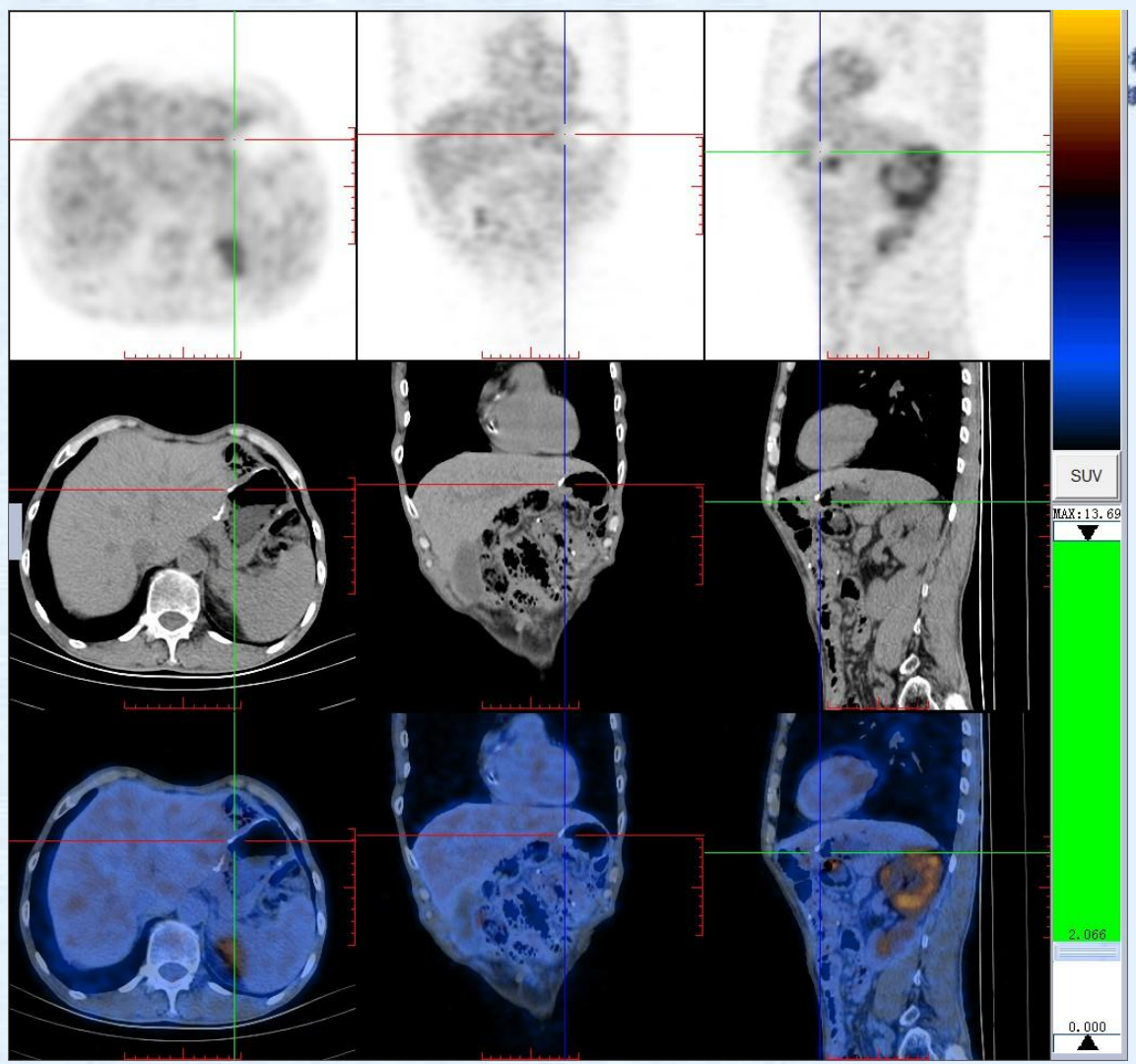
SUV

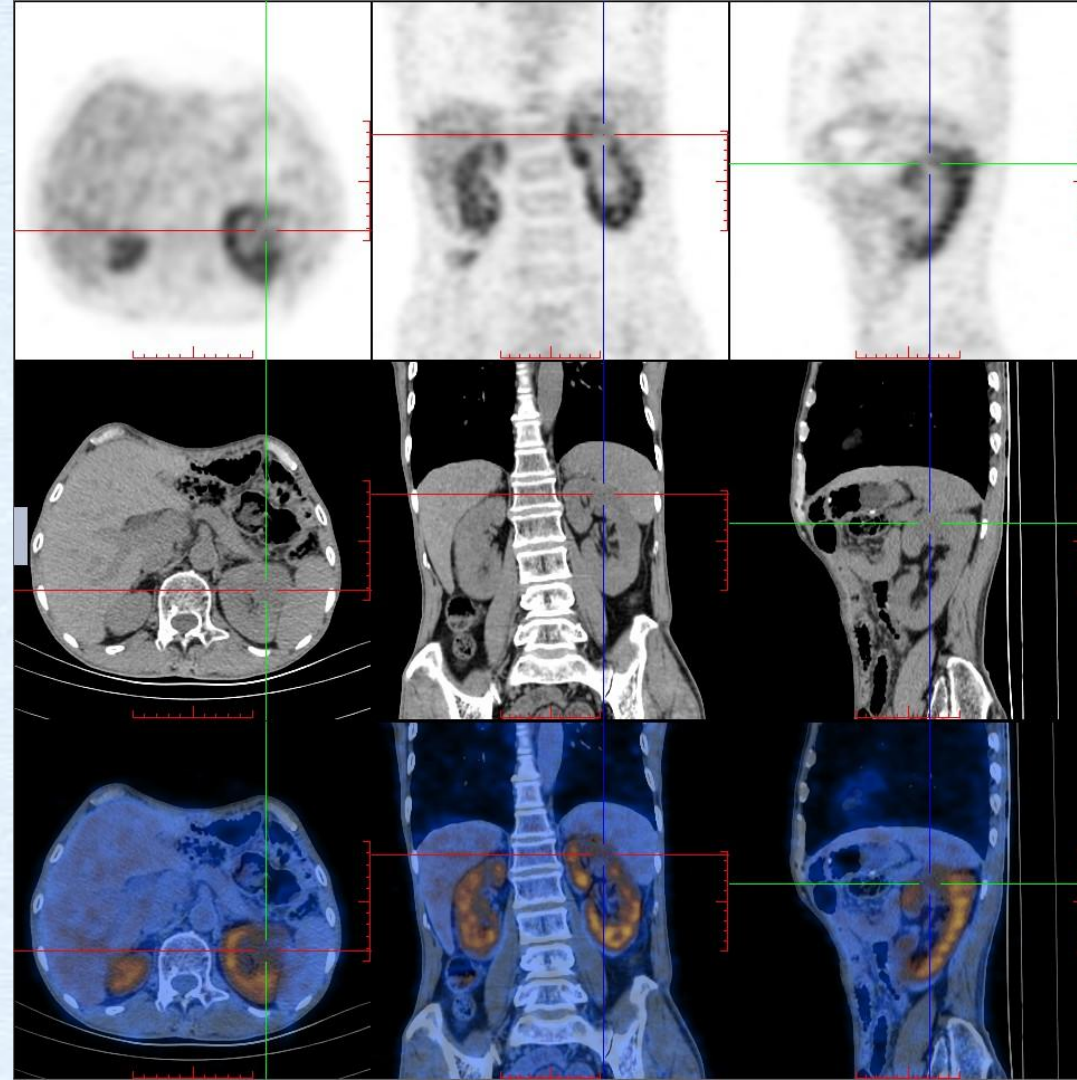
MAX: 13.69

2.066

0.000

胃癌术后

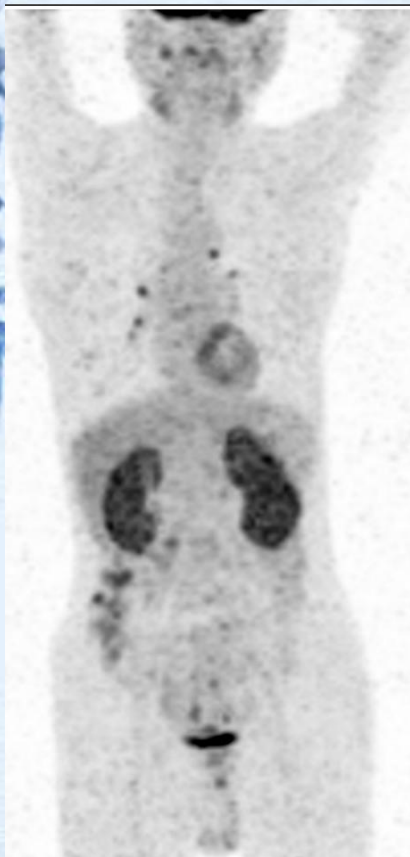




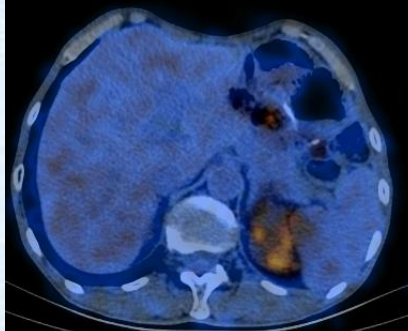
左肾低密度灶
CT值 25Hu
放射性分布↓



PET/CT

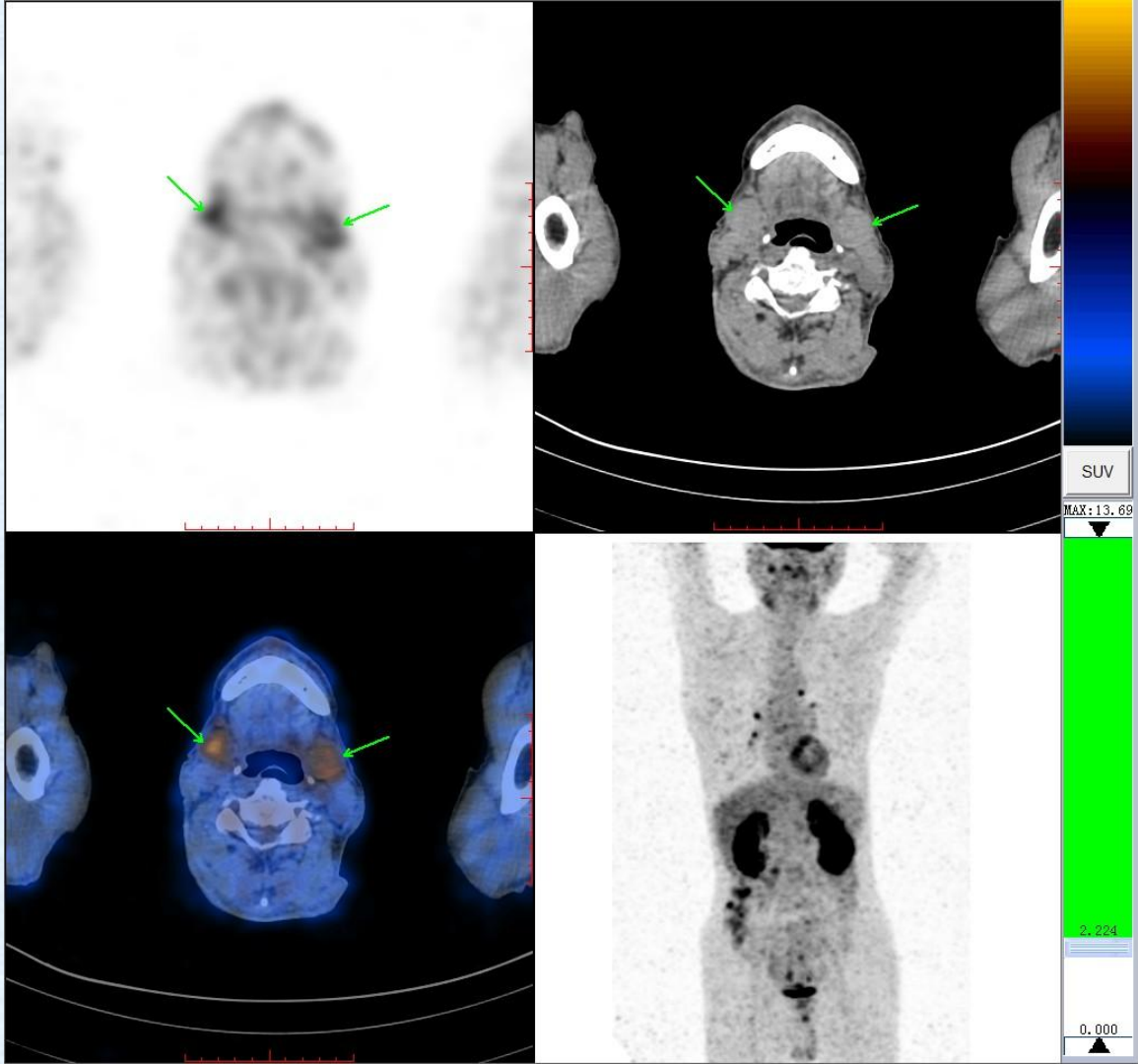


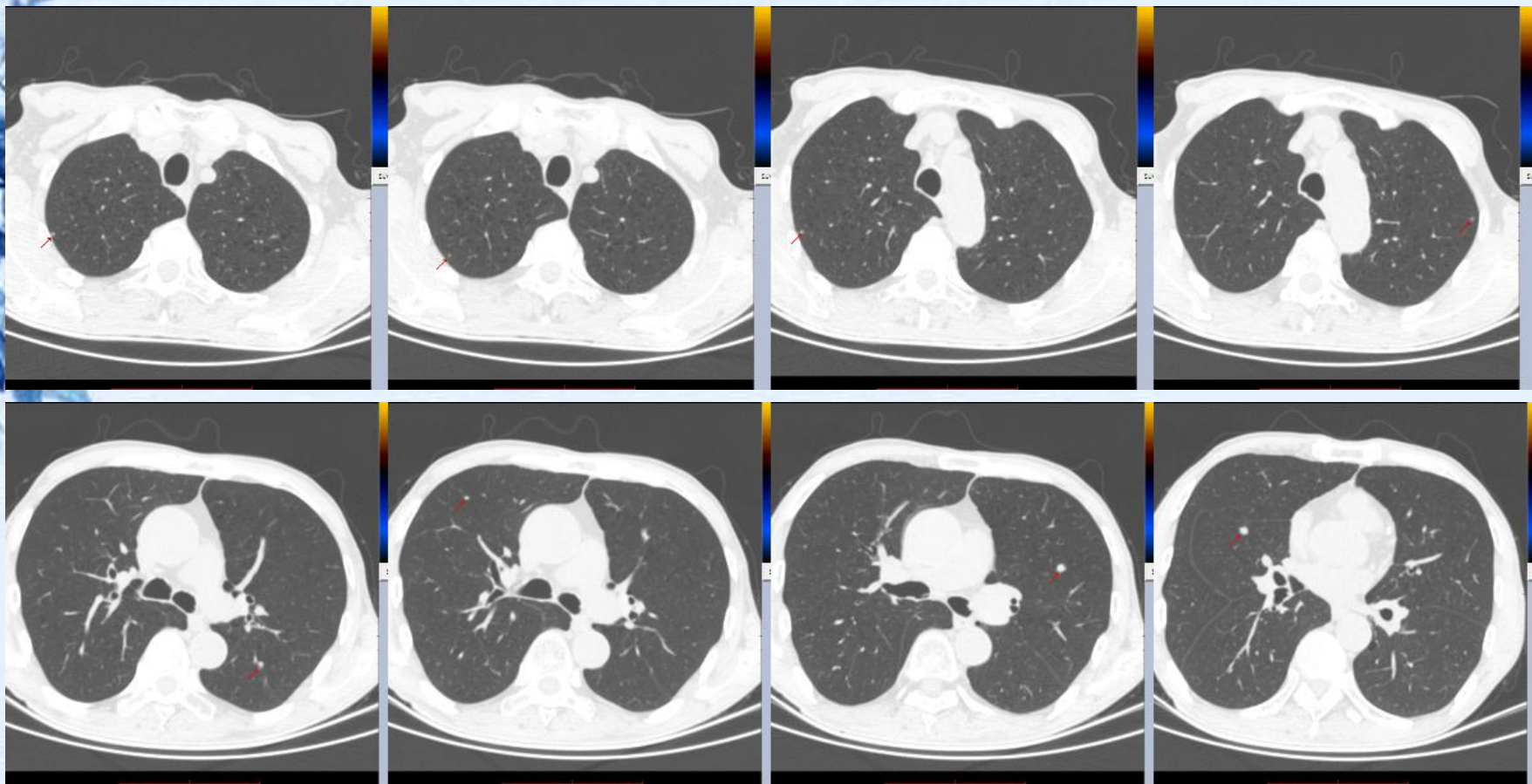
双肾SUVmax 5.0





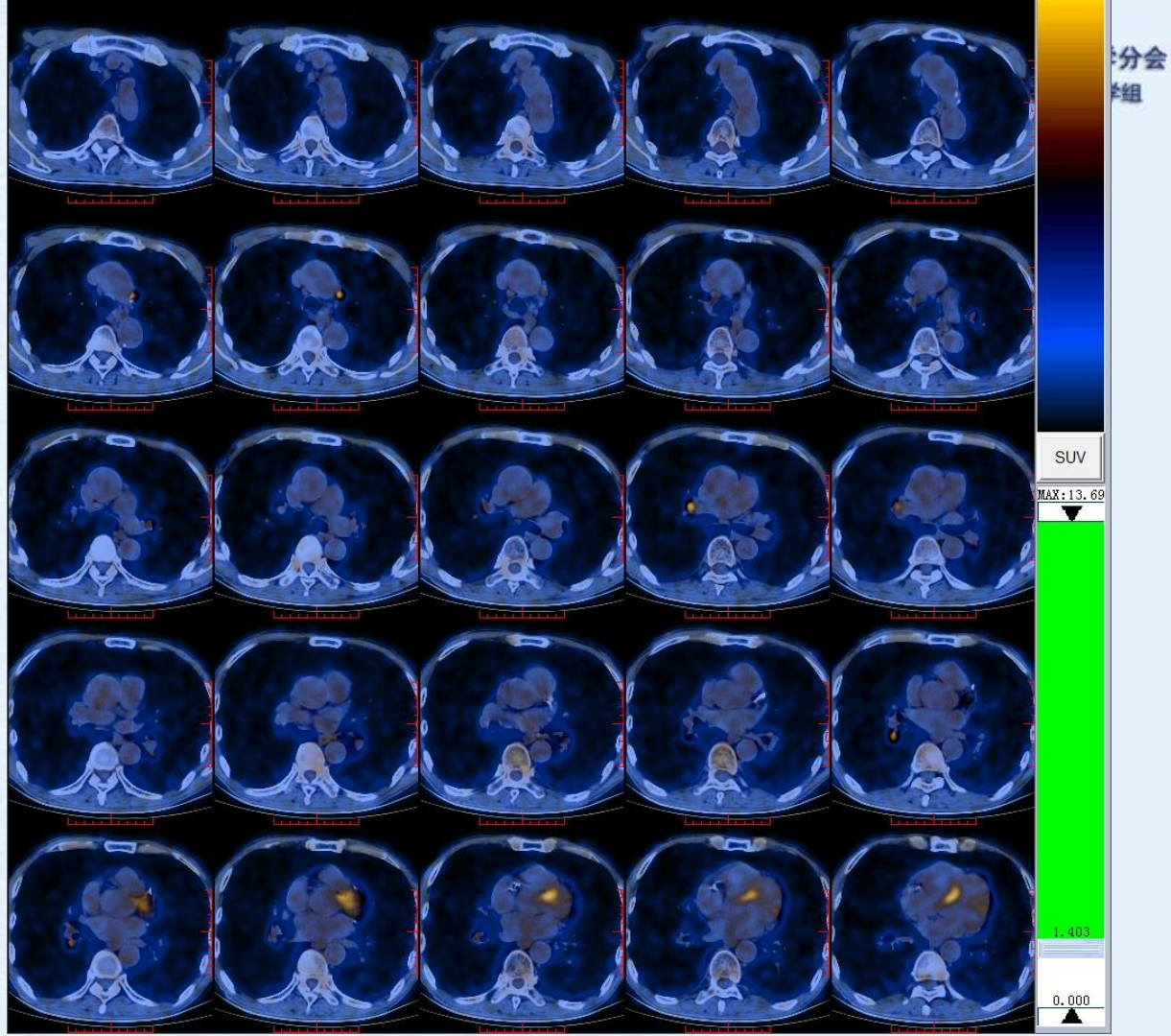
双侧颌下腺
SUVmax 3.0





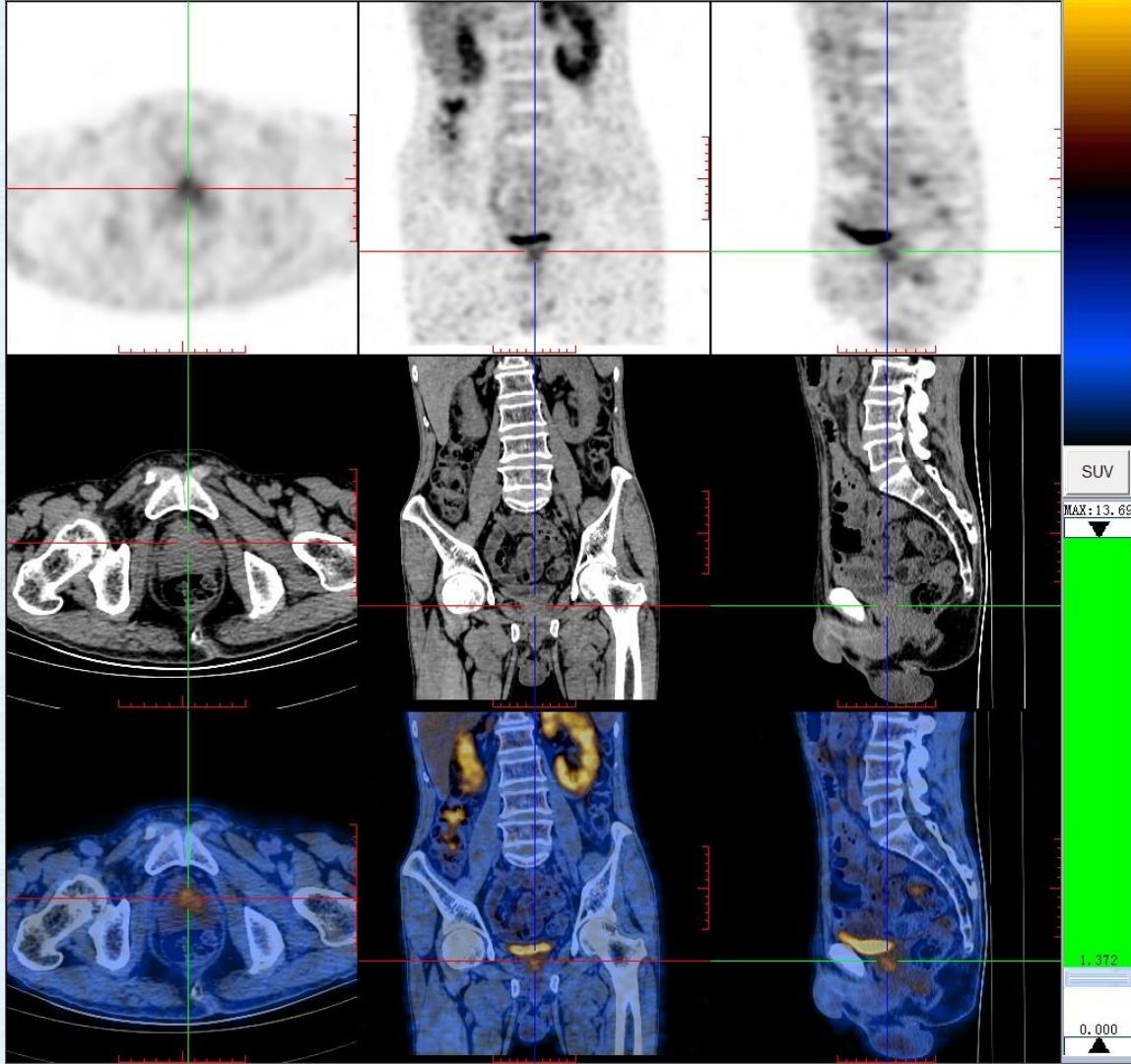


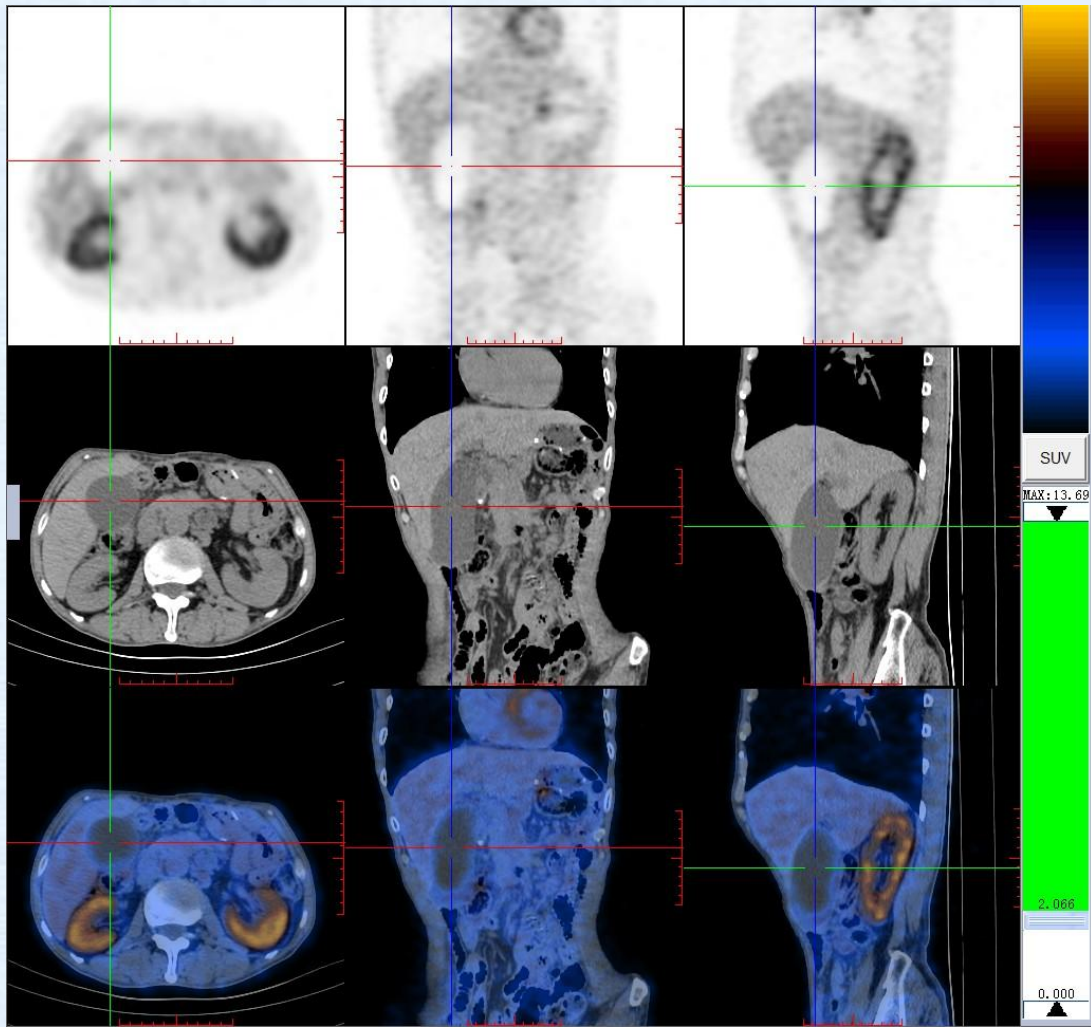
纵隔及肺门小淋巴结
SUVmax 4.5





前列腺
SUVmax 3.2





MDT目的、拟解决的问题

病史、检验

老年男性，慢性起病
反复腹痛，肌酐进行性升高

PET/CT

多器官病变

问题

最终诊断?

MDT中 临床提出的解决方法、建议，疑问

- 肾功能受损原因？最终诊断？
- 建议完善实验室检查、肾穿活检、骨穿

肾功能受损原因

肾损伤原因

肾前性

不符合

肾性

肾性

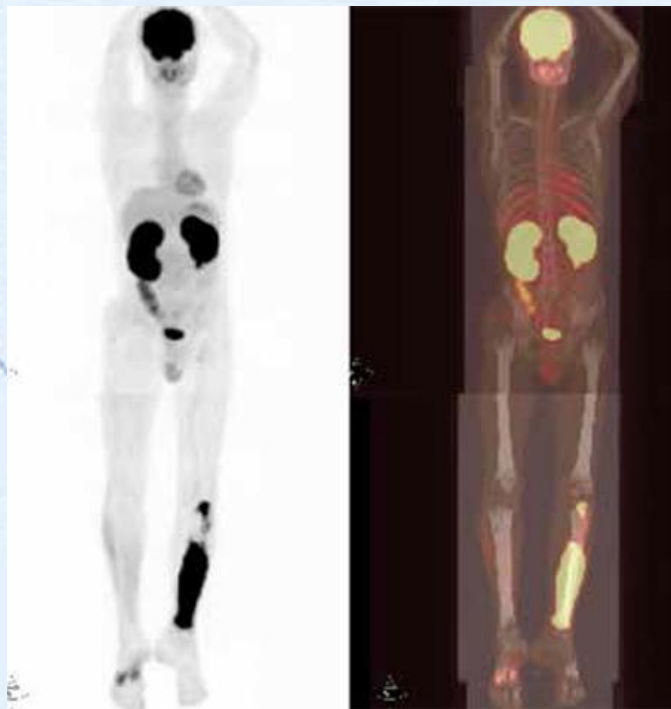
MDT中核医学能够解决那些问题？ 核医学自身有哪些疑问及其解决方案？

- 肾脏放射性分布增高可能的原因？
- 您的诊断？

双肾葡萄糖代谢增高可能的原因

- 淋巴瘤：NHL或HL患者可见肾脏受累，主要见于DLBCL，原发性肾淋巴瘤少见（肾脏不含淋巴组织）
- 白血病：少见，主要发生于儿童、成人急性髓系白血病，主要表现为肾脏肿大、弥漫性肾脏浸润，或多灶性肿块
- 转移：尸检发现20%的癌症患者有肾转移，常见的有肺癌、乳腺癌、结肠癌、恶性黑色素瘤。CT上通常为双侧、多发、小病灶
- 其它：肾炎、结节病、IgG-RD

淋巴瘤

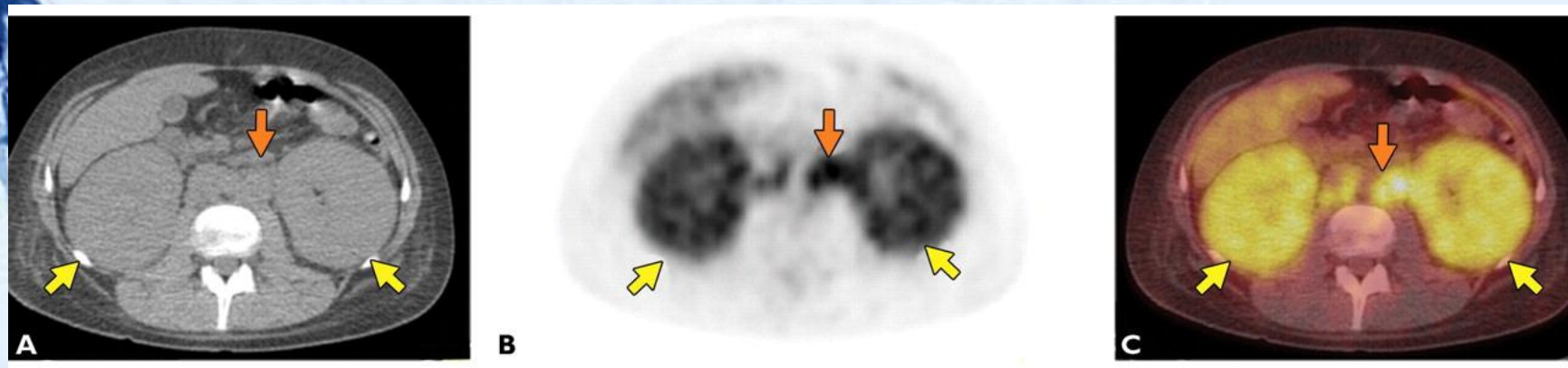


化疗前



化疗后

白血病



女，22岁，白血病，急性肾衰。病理：白血病累及双肾

转移



FIGURE 1. PET images of a 64-year-old patient with intraglomerular metastases of malignant melanoma, which is a rare entity with very few cases reported in the literature.¹⁻⁸ Coronal PET images show diffuse bilateral glomerular FDG uptake.

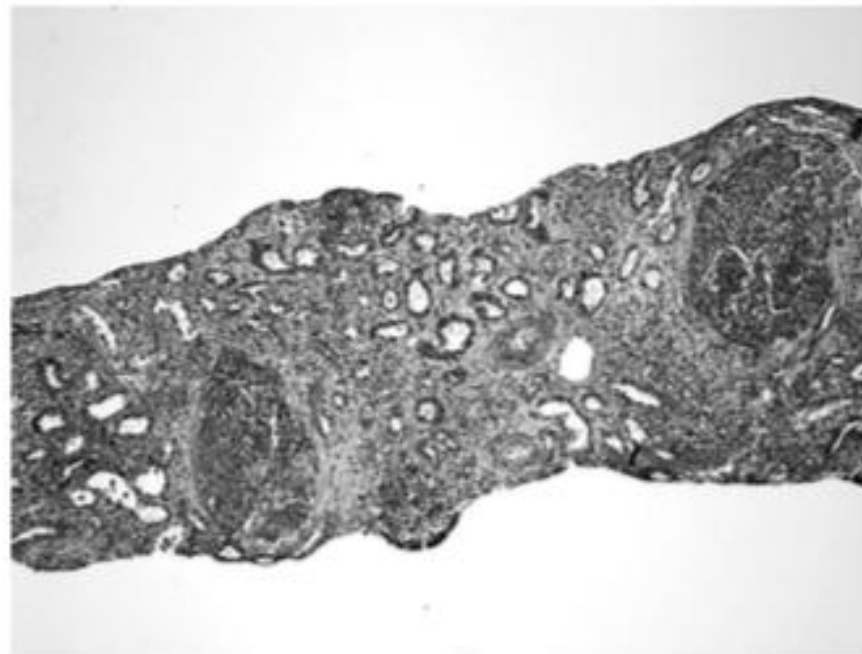
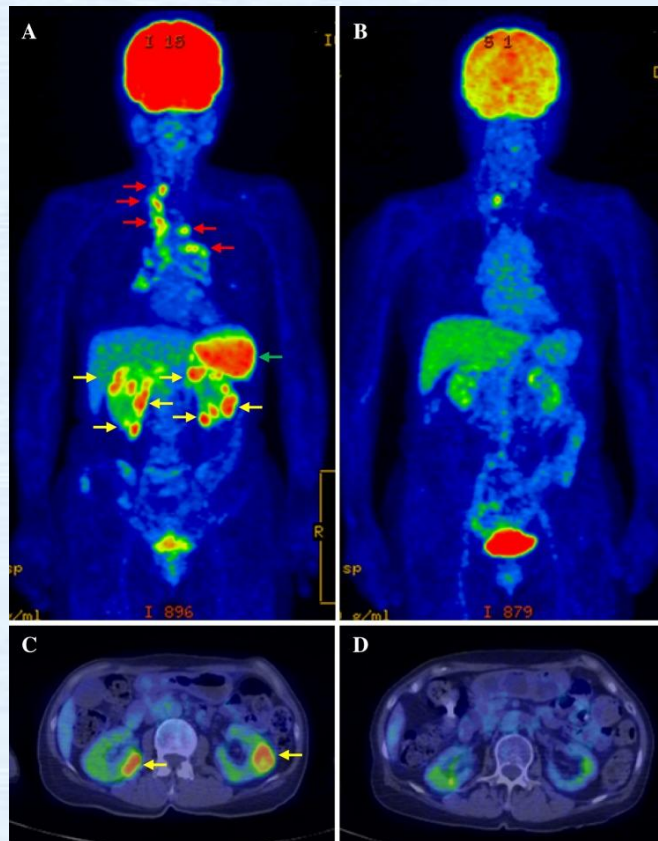
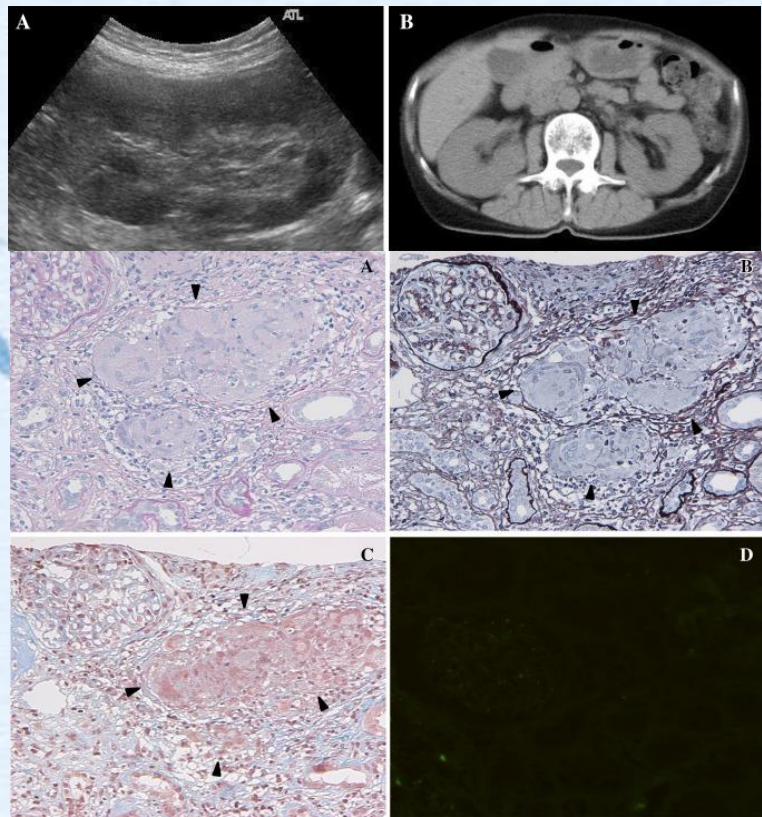
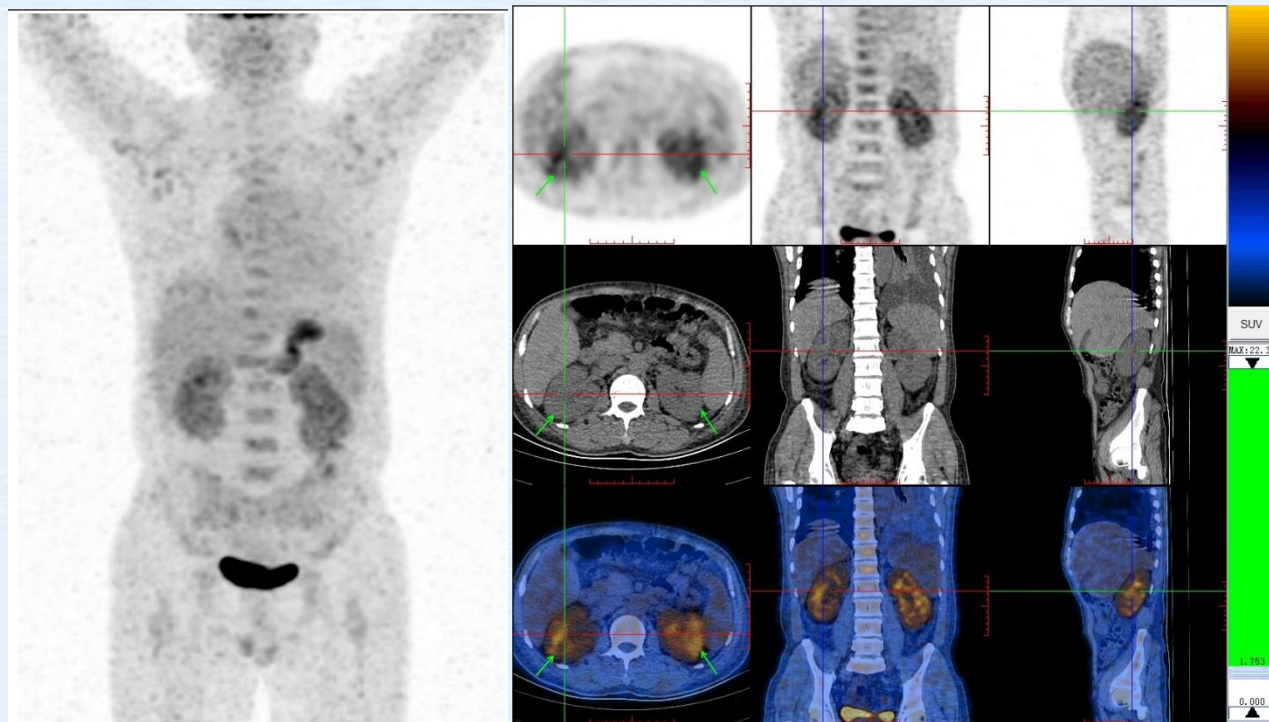


FIGURE 2. Renal biopsy revealed intracapillary tumor thrombi and diffuse crescent-like extracapillary metastatic melanoma cells in the glomeruli (hematoxylin-eosin, original magnification $\times 100$).

结节病



狼疮性肾炎



男, 50岁, SUVmax 5.4。肾穿病理: 毛细血管内增生性狼疮性肾炎

实验室检查

	项目	结果	参考范围
免疫球蛋白	IgA	2.33	0.69-3.82 g/L
	IgM	0.5 ↓	0.63-2.77 g/L
	IgG	17.3 ↑	7.23-16.85 g/L
	IgG1	8.35	4.9-11.4 g/L
	IgG2	8.01 ↑	1.69-7.86 g/L
	IgG3	1.5 ↑	0.11-0.85 g/L
	IgG4	11.7 ↑	0.03-2.01 g/L

肾穿病理

- 结合临床，符合轻度系膜增生性狼疮性肾炎（II型）伴IgG4相关性肾小管间质肾病可能性大及缺血性肾损伤。
- 完善免疫组化提示肾组织间质内纤维组织增生，伴有灶性胶原硬化，并弥漫性小淋巴及浆细胞浸润，细胞形态无异型；IHC：CD3+++/弥漫，CD20++/灶性，CD138+++，IgG 230-240/HPF，IgG4 160-170/HPF，AE1/AE3 肾小管+，IgG4比IgG为65%。
- 综上，符合**肾IgG4相关性肾间质病**，伴肾小管萎缩。考虑**lgG4相关肾病**诊断明确

骨穿病理

- (骨髓) 穿刺皮质骨及骨髓组织, 骨髓增生稍活跃, 三系可见, 各阶段粒细胞均可见; 散在造红岛; 巨核细胞2-6个/HPF。
- 各系细胞未见明显形态异常。间质内少许浆细胞浸润 (CD138+, kappa+, lambda+, IgG 40/HPF, IgG4 20/HPF, IgG4: IgG为50%, 约占5%), 未见异常上皮 (AE1/AE3 -)。请结合临床其他检查综合分析。

MDT结果

- PET/CT
- 血清学IgG4↑
- 肾穿病理



IgG4相关性疾病

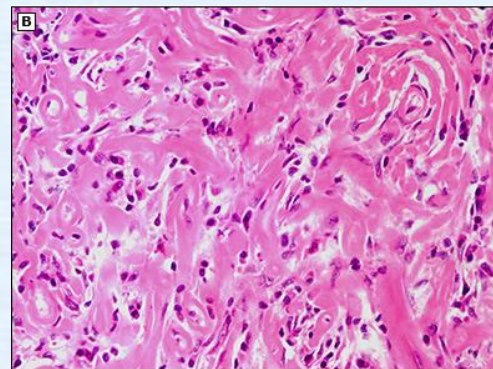
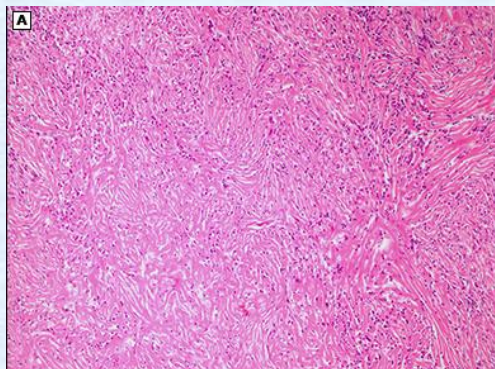
IgG4-RD

- IgG4相关性疾病（immunoglobulin G4-related disease,IgG4-RD）：免疫介导的纤维炎性疾病，2011年被统一命名为IgG4-RD
- 发病机制仍不完全清楚。有证据表明：为一种自身免疫性疾病，T细胞有重要作用；IgG4抗体本身不致病

Previous name	Target organ(s)
Mikulicz's disease	Salivary and lacrimal glands
Küttner's tumor	Submandibular glands
Riedel's thyroiditis	Thyroid
Chronic sclerosing aortitis	Aorta
Inflammatory abdominal aortitis	Abdominal aorta
Retroperitoneal fibrosis	Retroperitoneum
Autoimmune pancreatitis	Pancreas
Sclerosing cholangitis	Biliary tree
Orbital pseudotumor	Orbital adnexa
Eosinophilic angiocentric fibrosis	Sinuses and nasal cavities
Multifocal fibrosclerosis	Various organs

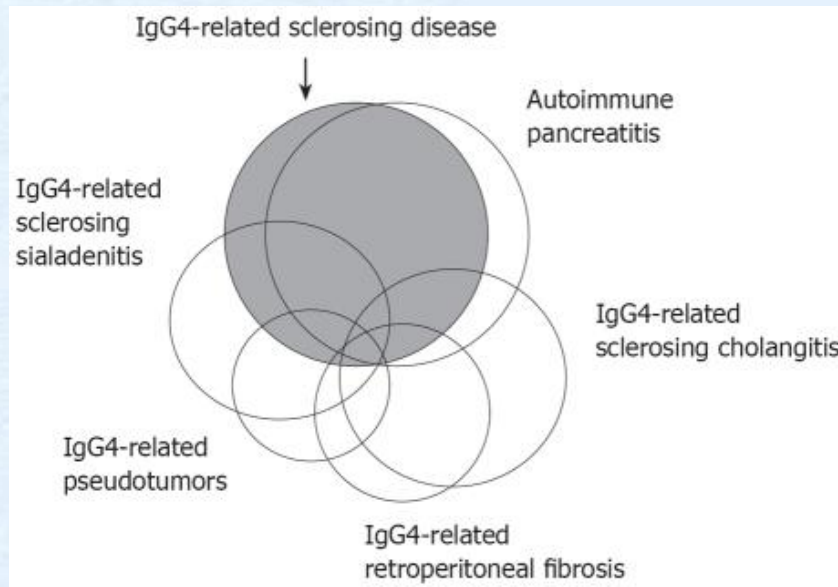
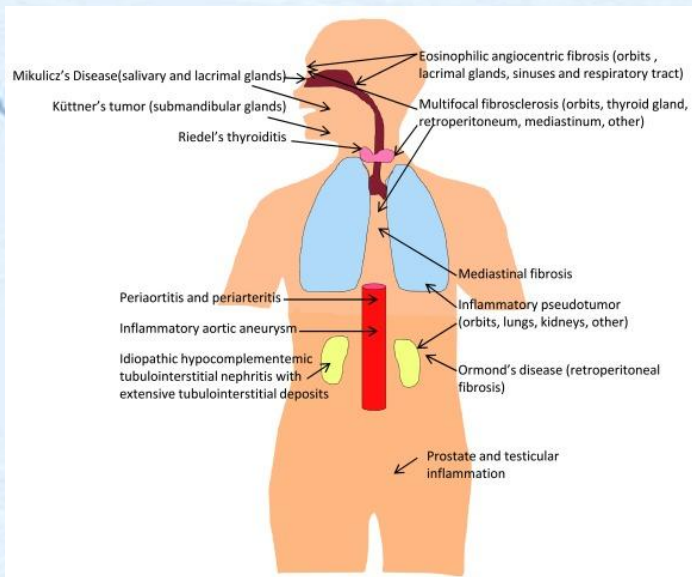
IgG4-RD

- 病理特征：病变组织中以IgG4+浆细胞为主的淋巴浆细胞浸润，常伴有“席纹”状纤维化、阻塞性静脉炎、嗜酸粒细胞浸润
- 约2/3的患者血清IgG4水平升高，一部分患者存在典型组织病理学改变，但血清IgG4水平正常。血清和组织IgG4升高不具特异性
- 糖皮质激素初始治疗有效



IgG4-RD

- 通常略好发于中老年男性，也可见于女性、儿童。一或多个器官受累，可有无症状性淋巴结肿大（通常无压痛），多伴有血清IgG4水平升高





诊断标准

Comprehensive diagnostic criteria for IgG4-RD, 2011 (27)

1. Clinical examination showing characteristic diffuse/localized swelling or masses in single or multiple organs.
2. Hematological examination shows increased serum IgG4 concentrations ($>135\text{mg dl}^{-1}$).
3. Histopathologic examination shows
 - (i) Marked lymphocyte and plasmacyte infiltration and fibrosis.
 - (ii) Infiltration of IgG4+ plasma cells: ratio of IgG4+/IgG+ cells $>40\%$ and >10 IgG4+ plasma cells per HPF.

Definite: 1+2 + 3

Probable: 1+3

Possible: 1+2

However, it is important to differentiate IgG4-RD from malignant tumors of each organ (e.g. cancer, lymphoma) and similar diseases (e.g. SS, primary sclerosing cholangitis, Castleman's disease, secondary RPF, Wegener's granulomatosis, sarcoidosis, Churg-Strauss syndrome) by additional histopathological examination.

Even when patients cannot be diagnosed using the CCD criteria, they may be diagnosed using organ-specific diagnostic criteria for IgG4-RD.

临床表型

- 有研究根据其最常见表现和受累器官分为4种主要临床表型

主要临床表型

胰肝胆疾病

腹膜后纤维化和/或主动脉炎

局限于头颈部的疾病

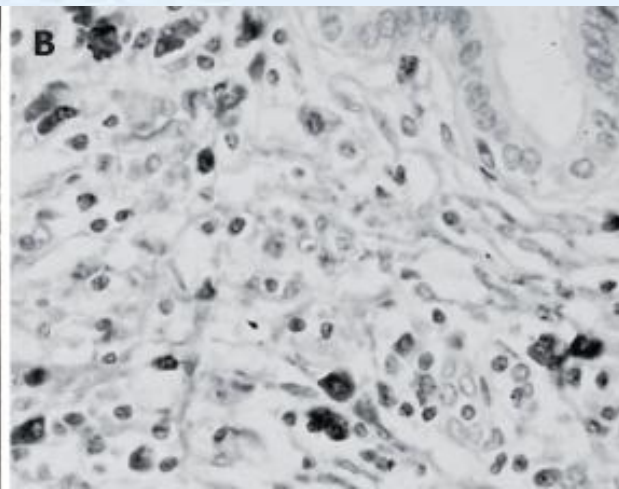
全身受累的典型Mikulicz综合征

自身免疫性胰腺炎

- 自身免疫性胰腺炎（AIP）：最常见的IgG4-RD亚型之一。分**1型（最常见，与IgG4-RD 有关）**、2型（特发性导管中心性胰腺炎）
- 常表现为胰腺肿块或无痛性阻塞性黄疸，可被误诊为**胰腺癌**（血清IgG4有助鉴别，但部分胰腺癌患者血清IgG4也可↑）。可有DM、体重下降等。
- 大多数1型AIP同时存在其他IgG4-RD，如硬化性胆管炎、腹膜后纤维化等

自身免疫性胰腺炎

- 典型影像：胰腺弥漫性肿大，“腊肠状”；有时增强扫描边缘胶囊状低密度环
- 其它：胰腺肿物或节段性肿大；胰管狭窄；胰腺萎缩；胰腺钙化；胰腺炎



自身免疫性胰腺炎

A



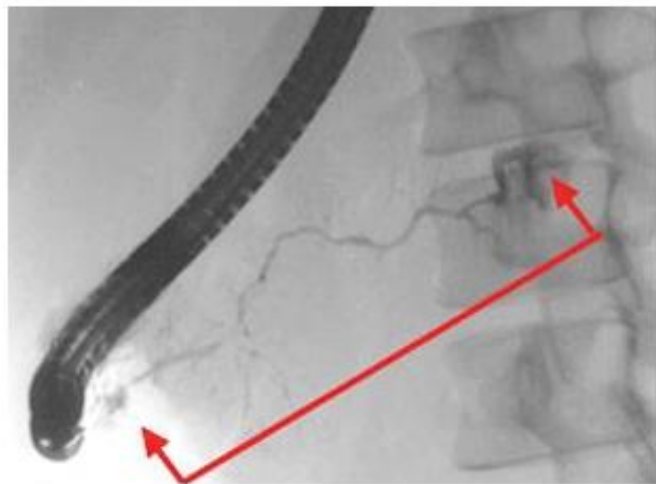
CT: 胰腺弥漫肿大, 胰周可见水肿晕轮

B



MRCP: 胆总管末端狭窄, 主胰管狭窄, 不伴扩张

自身免疫性胰腺炎



CT：胰腺弥漫肿大，胰周低密度晕

Pancreatogram：主胰管弥漫不规则狭窄

自身免疫性胰腺炎

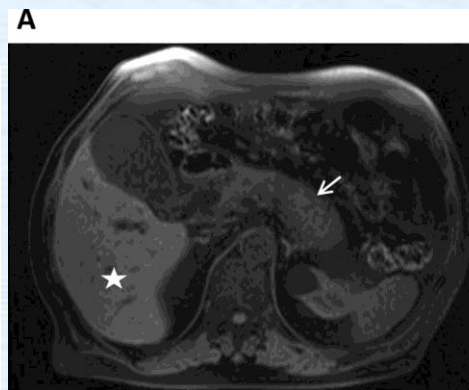
- 激素治疗有效。部分患者治疗中或停止治疗后复发。



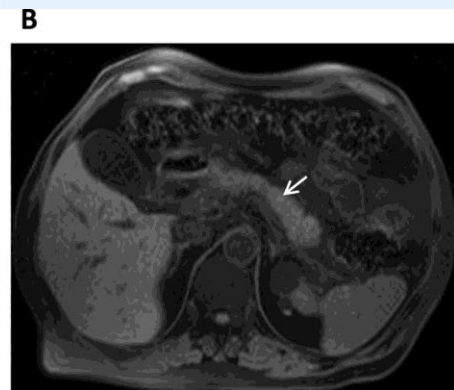
激素治疗前



激素治疗后



激素治疗前

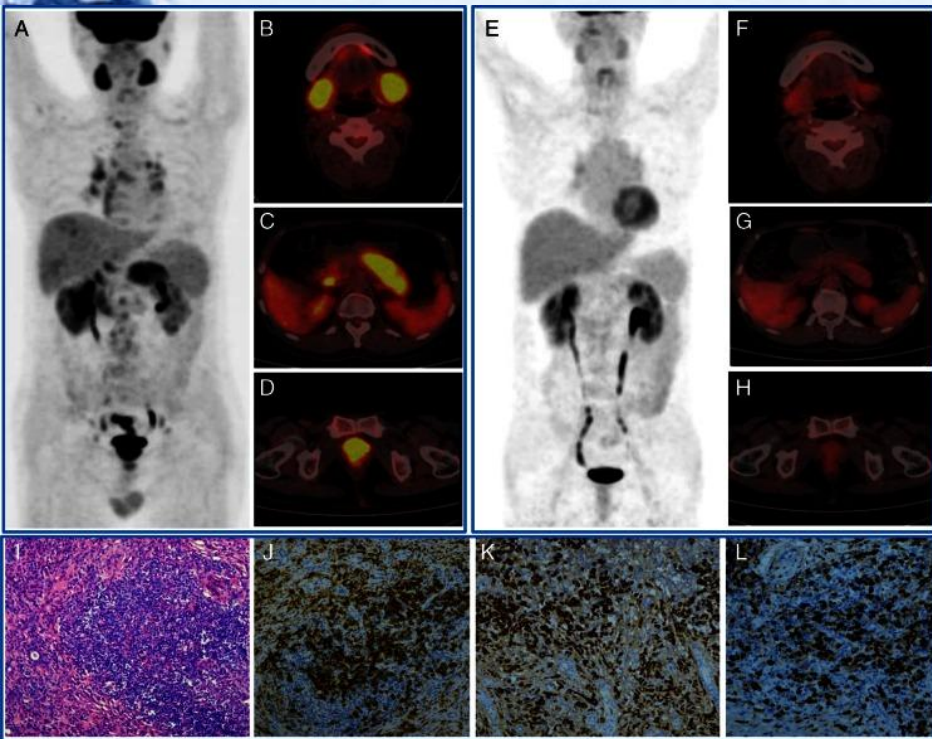


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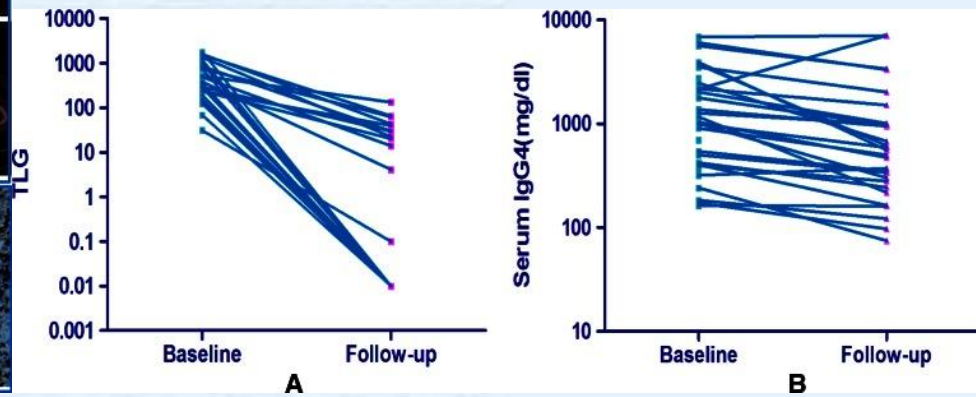
自身免疫性胰腺炎

激素治疗前

激素治疗后



男, 54岁, 多器官受累
血清IgG4升高

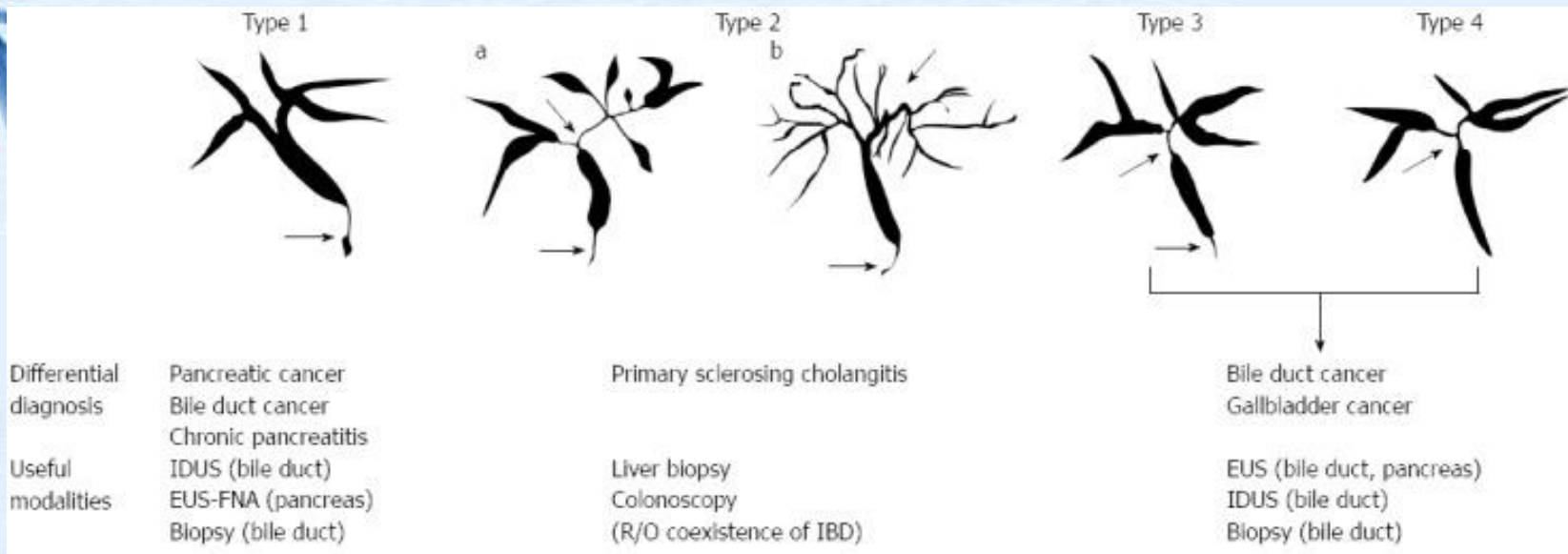


IgG4相关性硬化性胆管炎

- 1型AIP最常见的胰腺外表现；IgG4-RD第二常见表现，仅次于AIP
- 组织活检：胆管壁内IgG4浆细胞浸润和严重间质纤维化
- 临床和影像学特征类似原发性硬化型胆管炎（PSC），需与原发性硬化性胆管炎、胆管细胞癌鉴别
- 血清IgG4升高、激素治疗有效，可与PSC鉴别（PSC阻塞性黄疸少见）

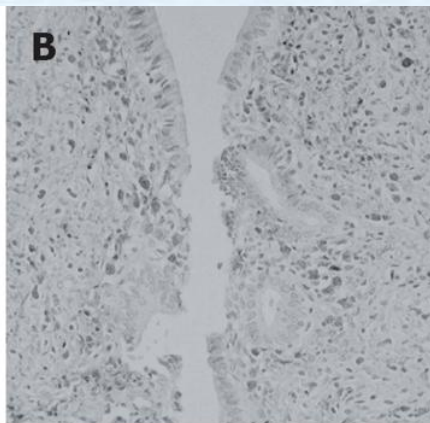
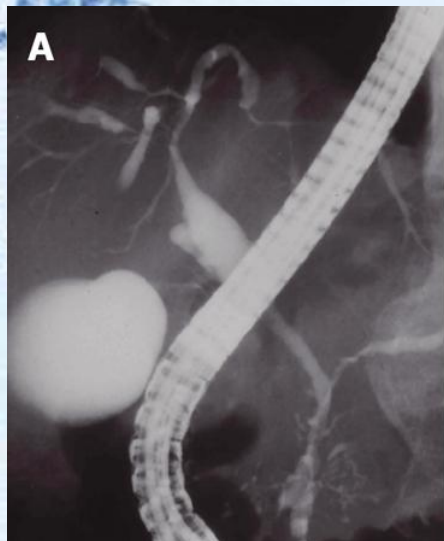
IgG4相关性硬化性胆管炎

- 肝内外胆管弥漫性/节段性狭窄，伴上游胆管扩张
- 有研究者根据胆管狭窄分布模式分为四型：仅胆总管下段受累；肝内外胆管弥漫受累；肝门区和胆总管下段受累；仅肝门区胆管受累

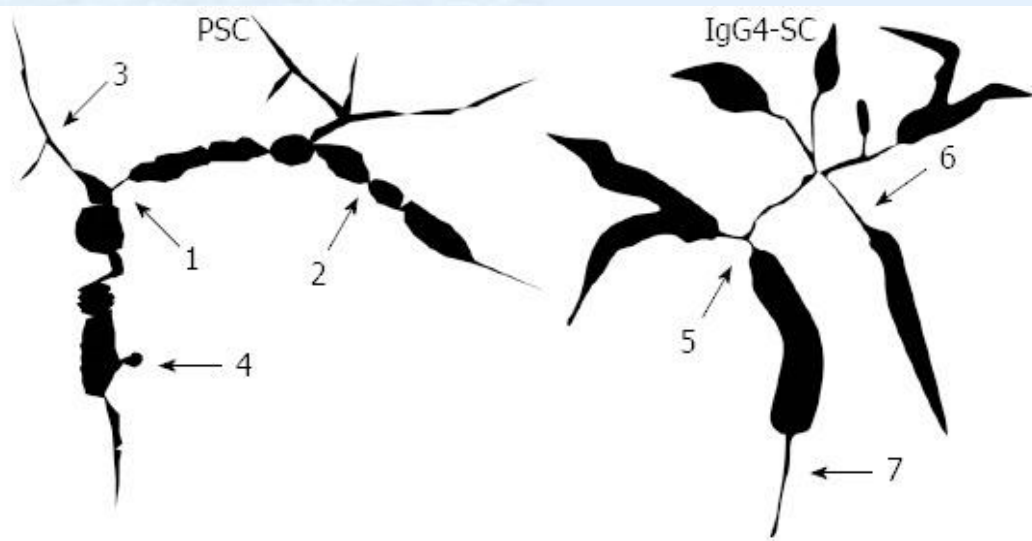


IgG4相关性硬化性胆管炎

- 临床和影像学特征类似原发性硬化型胆管炎 (primary sclerosing cholangitis, PSC)



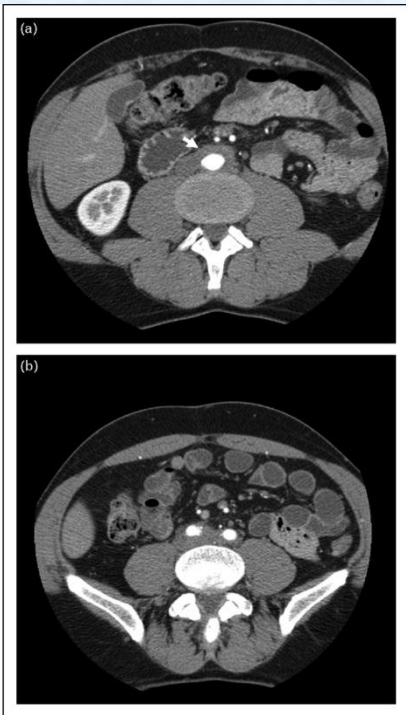
肝内胆管狭窄，与PSC类似
胆管壁可见IgG4+浆细胞浸润



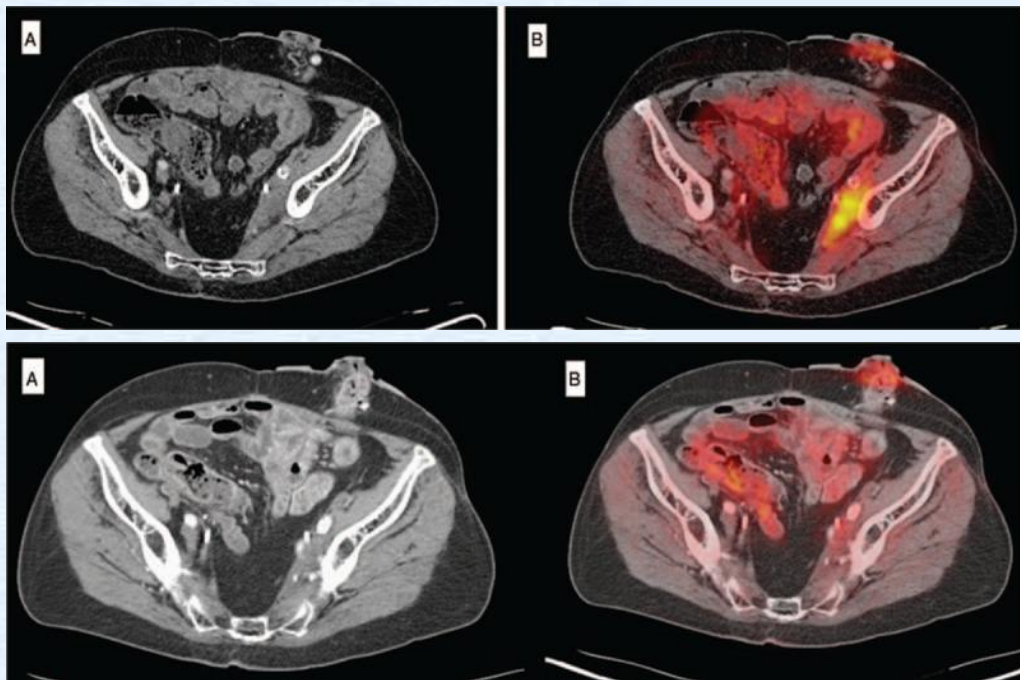
腹膜后纤维化

- 炎症性和纤维性腹膜后组织，常包绕输尿管或腹部器官，分特发性及继发性
- 有研究显示：58.8%的RPF患者同时存在血清IgG4升高、组织病理学提示IgG4-RD
- 最常见的IgG4-RD亚型之一

腹膜后纤维化

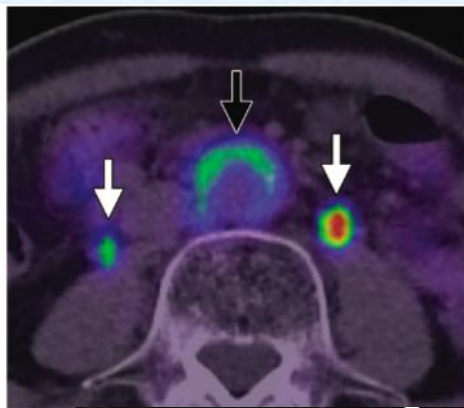


Computed tomographic angiogram revealing (a) an extensive soft tissue density that surrounds the inferior abdominal aorta. (b) This soft tissue density extends superiorly and inferiorly surrounding the common iliac arteries.

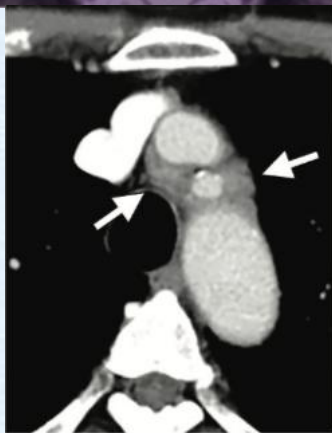


男，65岁，右腹痛、恶心、呕吐、无尿、左下肢水肿1月余
既往结肠癌术后+放化疗后20年，哮喘、肾结石、高血压
利妥昔单抗治疗后范围略缩小，代谢程度减低

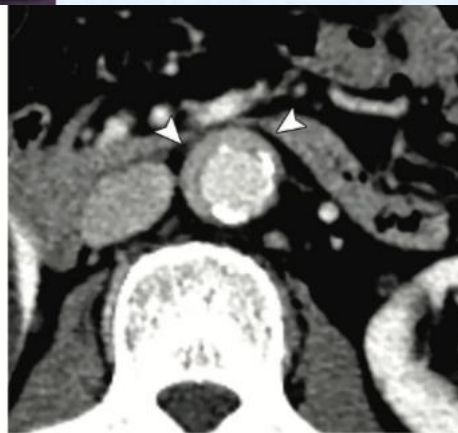
血管



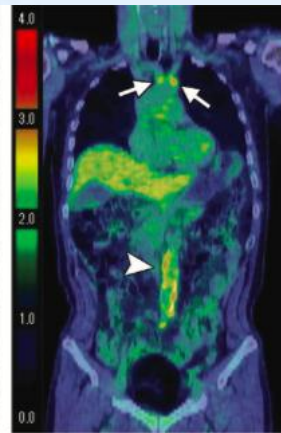
- 上图：男，75岁，IgG4相关性腹主动脉炎
- 下图：男，69岁，IgG4相关性动脉炎和动脉周围炎



a.

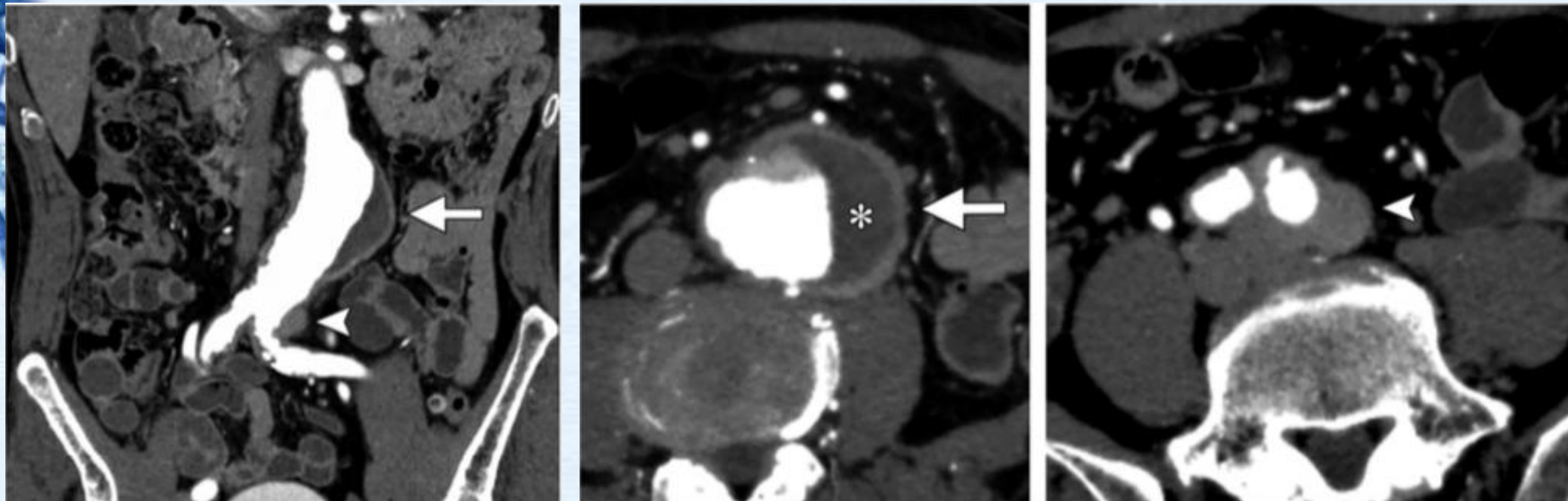


b.



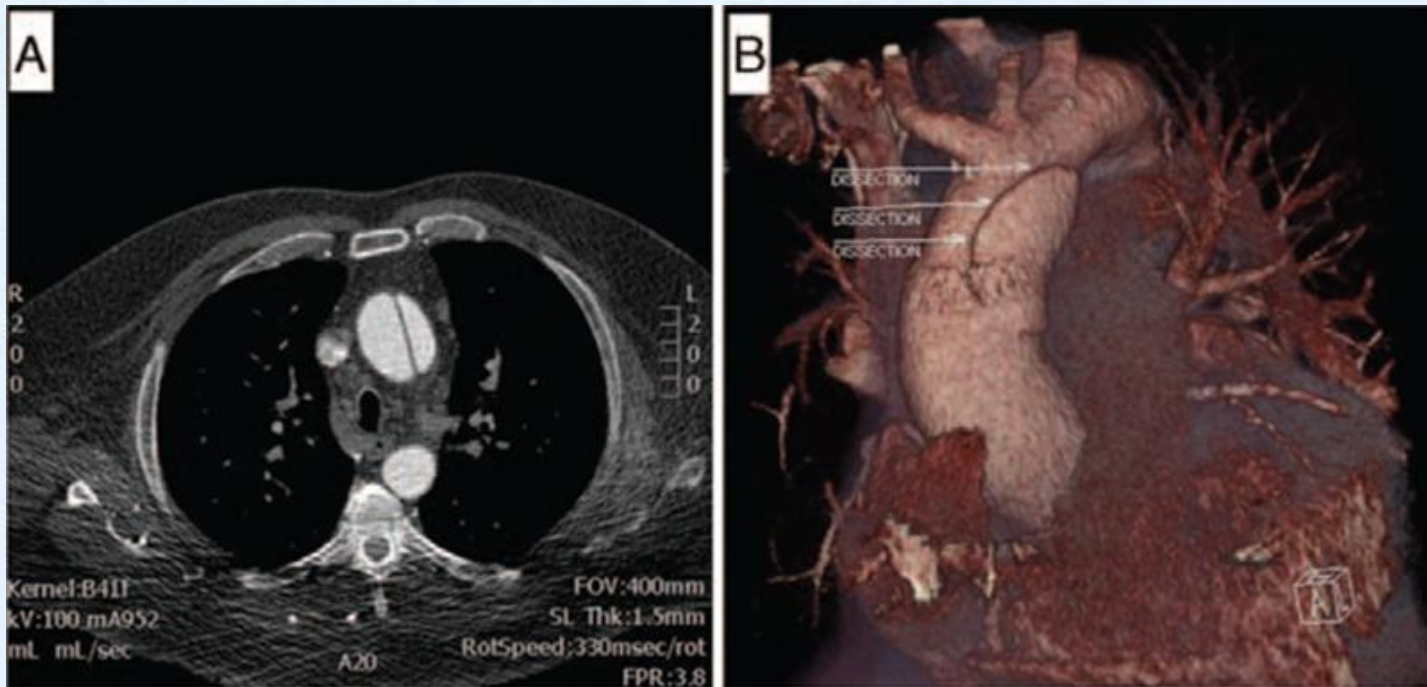
c.

血管



- 男，77岁，IgG4相关性炎性主动脉瘤

血管



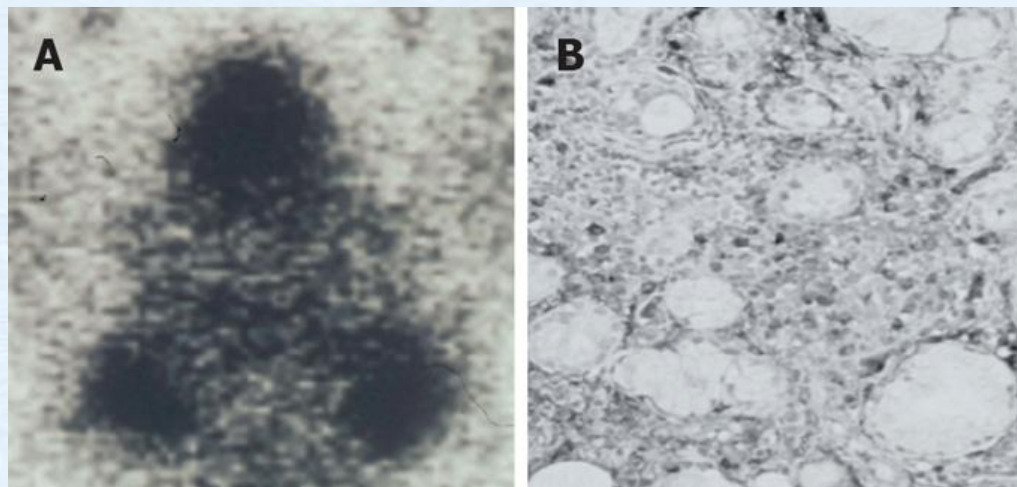
- 男，65岁，既往不明原因纵隔淋巴结肿大，术前CT：升主动脉瘤及主动脉夹层。
- 血清IgG4水平升高，病理提示IgG4-RD。诊断为IgG4相关性动脉夹层。

唾液腺、泪腺

- 可表现为泪腺、腮腺、下颌下腺肿大，一般为双侧
- 泪腺肿大合并腮腺和下颌下腺肿大时，称为IgG4相关性Mikulicz病



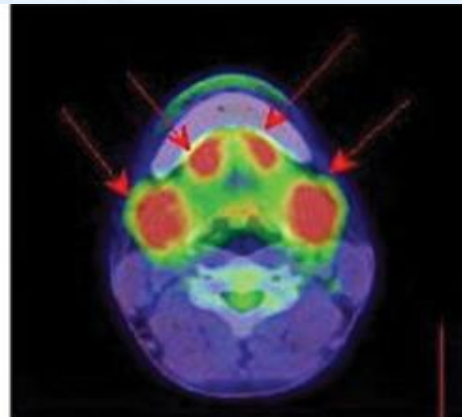
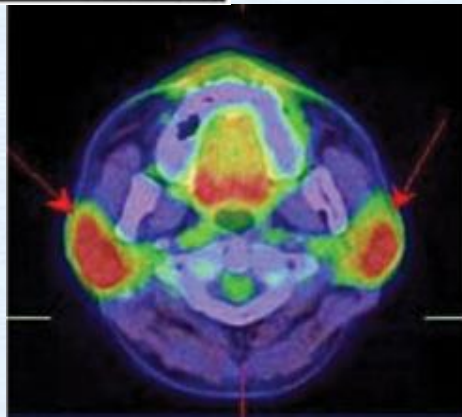
Her condition, once misdiagnosed as Sjögren's syndrome or termed 'Mikulicz disease' or 'Küttner tumor', is now known as IgG4-related sialadenitis. IgG4-related sialadenitis is part of the larger spectrum of IgG4-related systemic disease.



女，26岁，双侧下颌下腺和肿胀3月

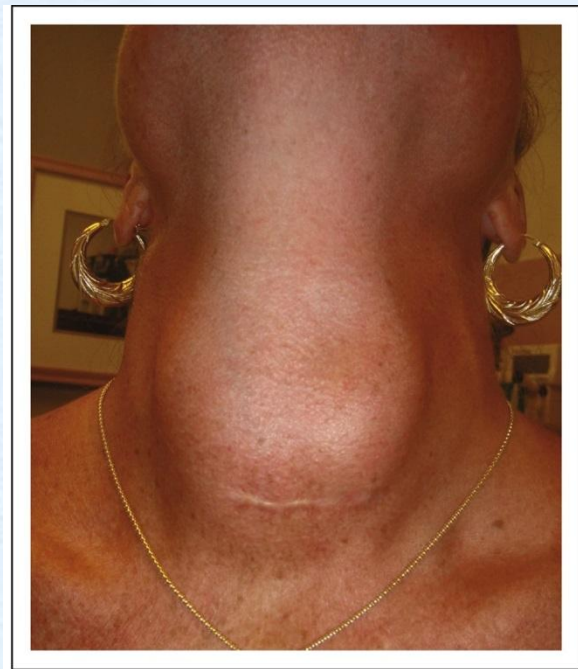
双侧下颌下腺肿胀（颌）

唾液腺、泪腺



甲状腺

- 桥本氏甲状腺炎
- Hashimoto's thyroiditis (HT): 罕见, 表现为硬性甲状腺肿, 可引起临近组织受压相关症状 (如呼吸困难、吞咽困难、声音嘶哑)



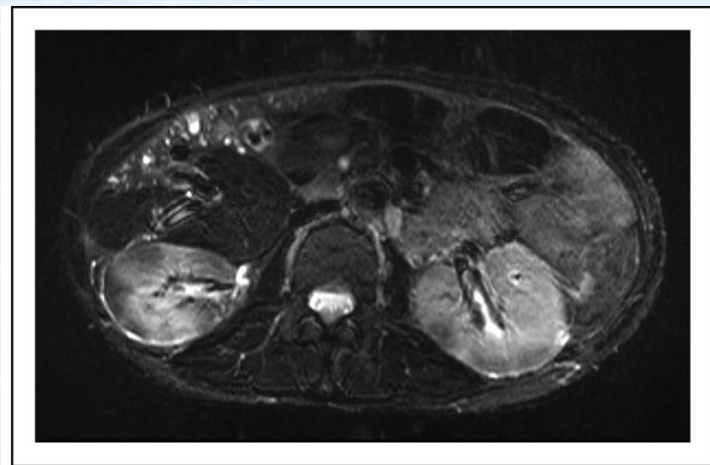
A large, woody goiter in a patient with a history of biliary tract disease, a pulmonary nodule, and lacrimal gland enlargement, all caused by IgG4-related systemic disease. Biopsy of the thyroid gland during the inflammatory (prefibrotic) phase of this patient's Riedel's thyroiditis also demonstrated classic histopathologic features of IgG4-related systemic disease.

肾脏

- 肾脏受累表现：蛋白尿、血尿、肾功能下降、异常影像学表现
- 炎性假瘤、小管间质性肾炎

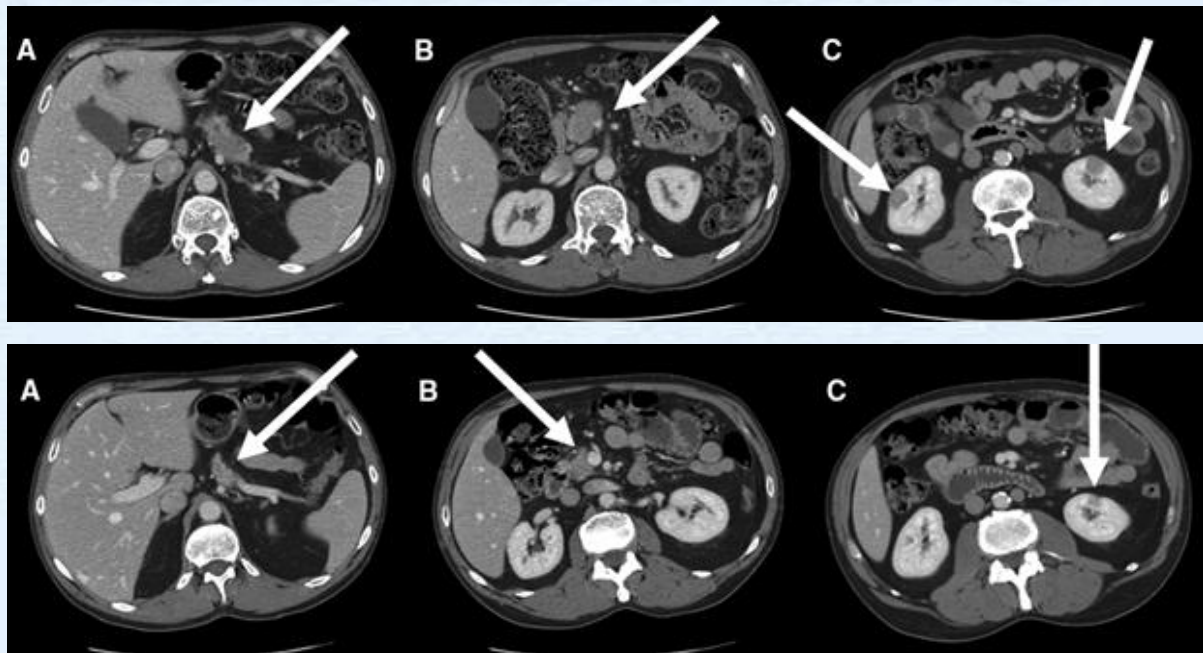


双肾多发低密度灶；左肾盂壁明显增厚，内壁光滑



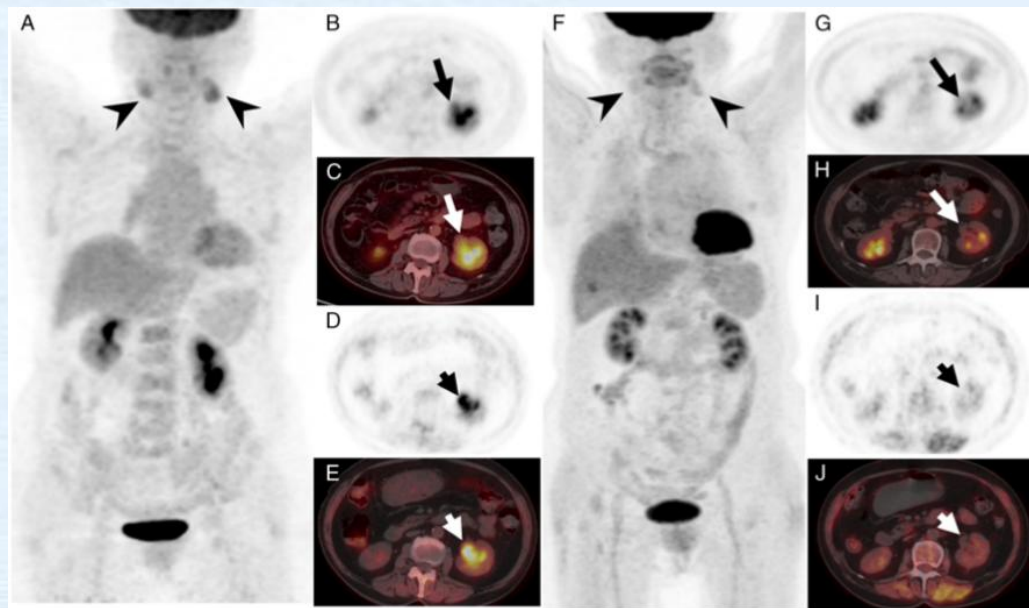
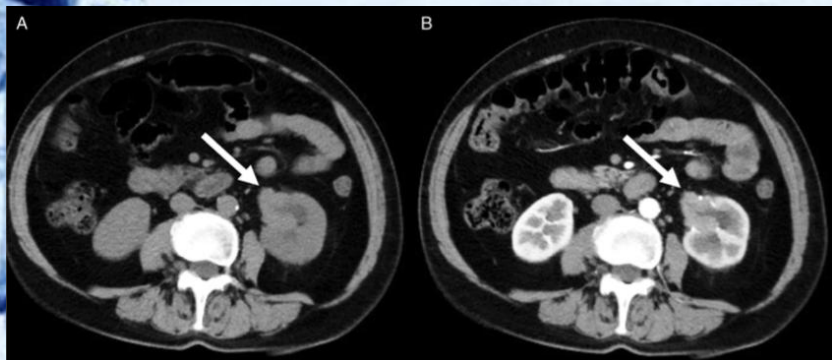
This patient also had IgG4-associated cholangitis and salivary gland swelling. The patient's kidney pathology revealed dense tubulointerstitial lymphoplasmacytic infiltrates and abundant IgG4+ plasma cells.

肾脏



男，52岁，体重下降2月，超声提示胰腺占位
结合血清IgG4水平及胰腺、肾穿刺病理，考虑IgG4-RD

肾脏



男，70岁，双侧颌下腺肿大。
血清IgG4↑□，颌下腺穿刺：IgG4-RD

激素治疗前， $SUV_{delayed}$ 9.3

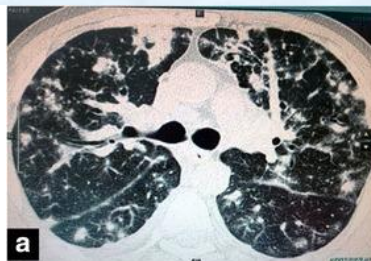
激素治疗后

肺

- 可无症状，或表现为咳嗽、咯血、呼吸困难、胸膜炎、胸痛
- CT分型：实性结节型；磨玻璃密度型；肺泡间质型；支气管血管型

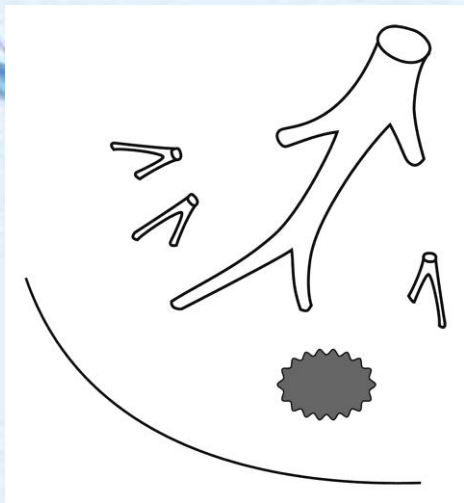
Chest CT findings

Nodules and masses with spiculated borders
Alveolar consolidations with air bronchograms
Ground glass opacities \pm reticular opacities
Hilar or mediastinal lymphadenopathy
Thickening of the bronchovascular bundles
Thickening of the interlobular septa

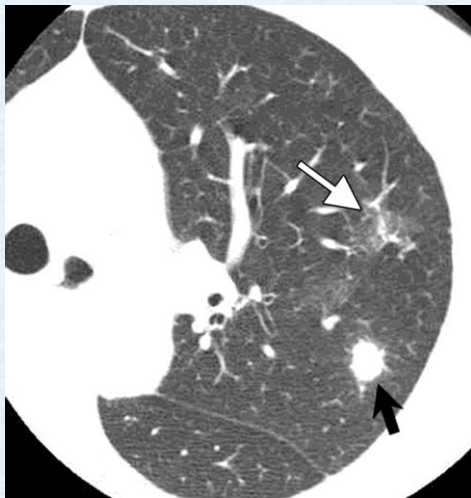


肺

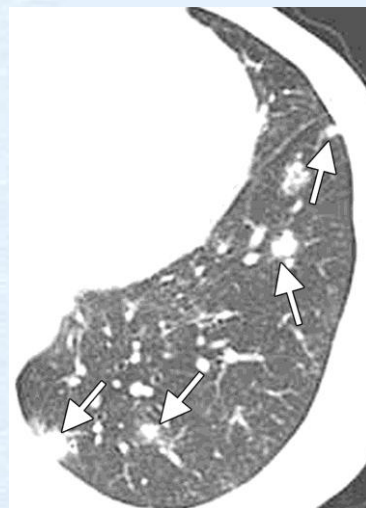
- 实性结节型



实性结节型



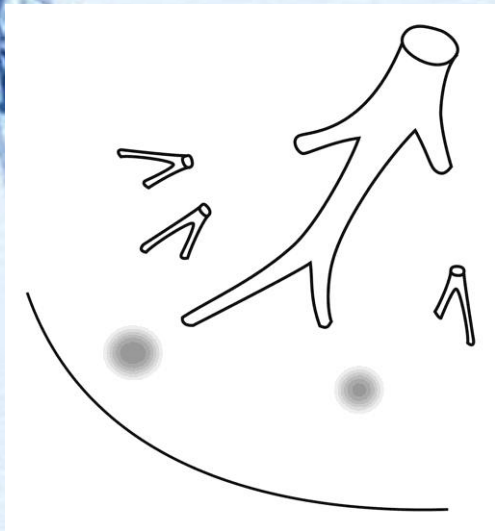
男，76岁，左肺上叶结节及磨玻璃密度影



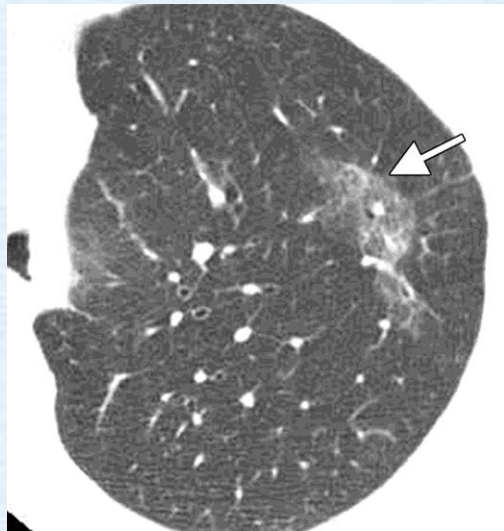
男，59岁，左肺上叶多发结节

肺

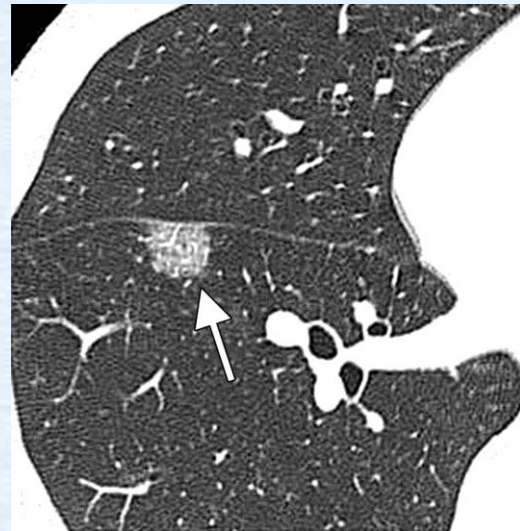
- 磨玻璃密度型



磨玻璃密度影型



男，76岁，左肺下叶磨玻璃密度影



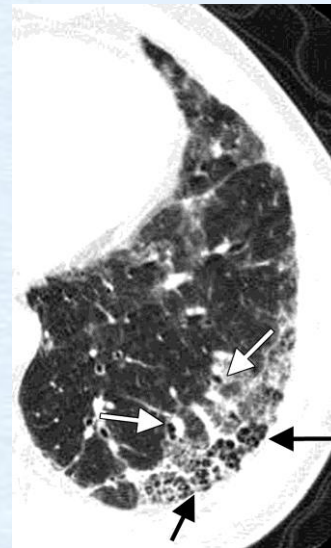
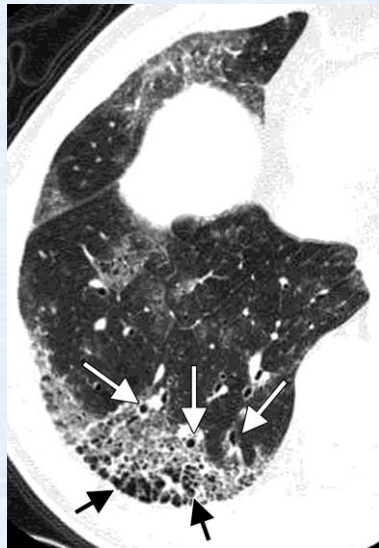
男，43岁，双肺多发GGO

肺

- 肺泡间质型



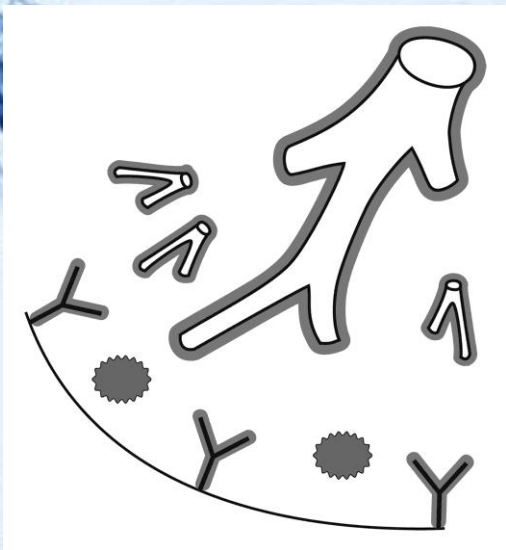
肺泡间质型



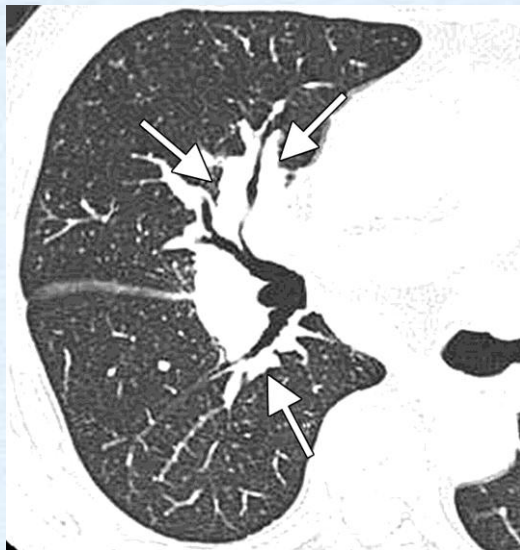
男，59岁，双肺下叶蜂窝状，伴支气管扩张、GGO

肺

- 支气管血管型



支气管血管型

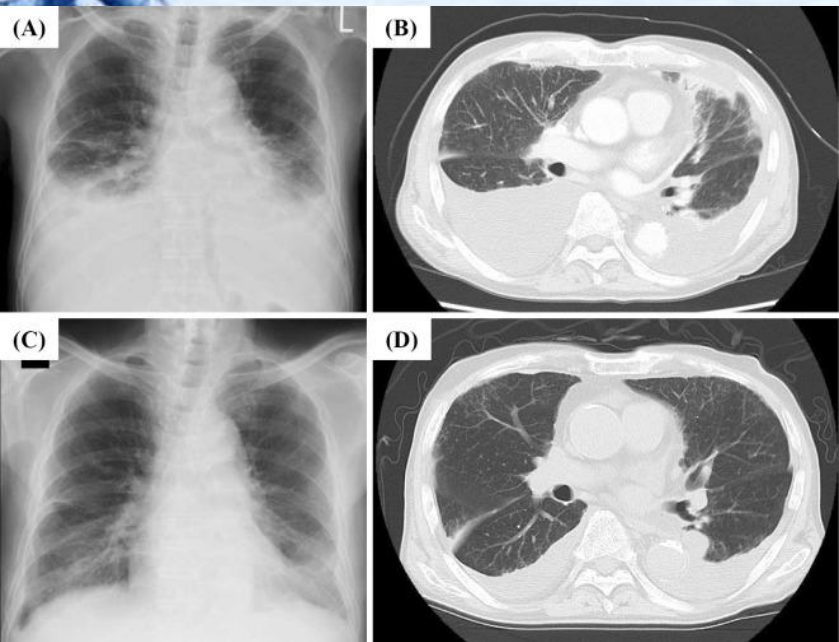


男，70岁，右肺门支气管血管束增粗

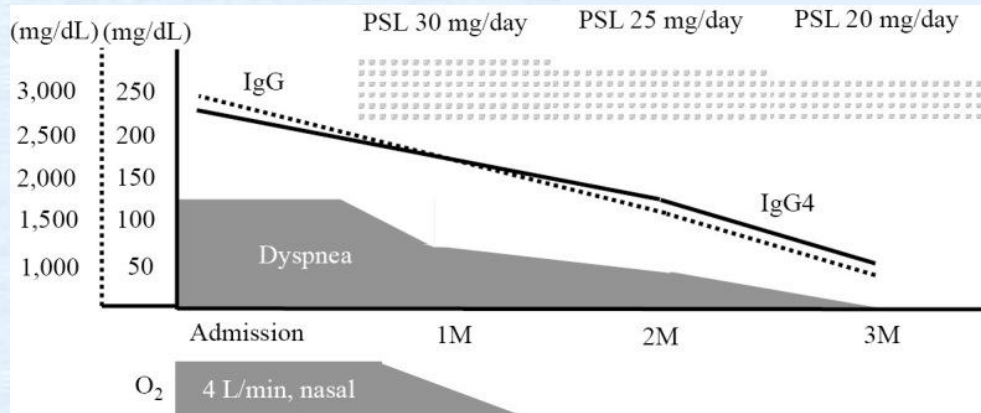


右肺支气管血管束增粗，小叶间隔增厚

胸膜



- 男，81岁，呼吸困难
- CT：双肺胸膜下磨玻璃密度灶、左肺实变灶；纵隔淋巴结肿大
- IgG4水平升高，病理提示IgG4-RD
- 激素治疗后，双侧胸腔积液吸收



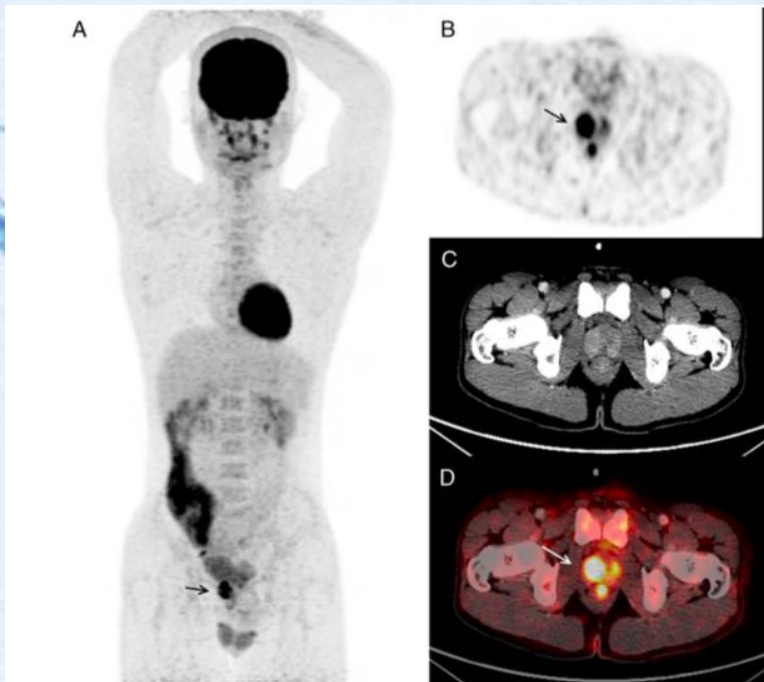
淋巴结肿大

- 80%的AIP患者具有淋巴结肿大，有时是IgG4-RD患者的唯一临床表现
- 五种病理类型，共同特征为：IgG4+浆细胞弥漫分布

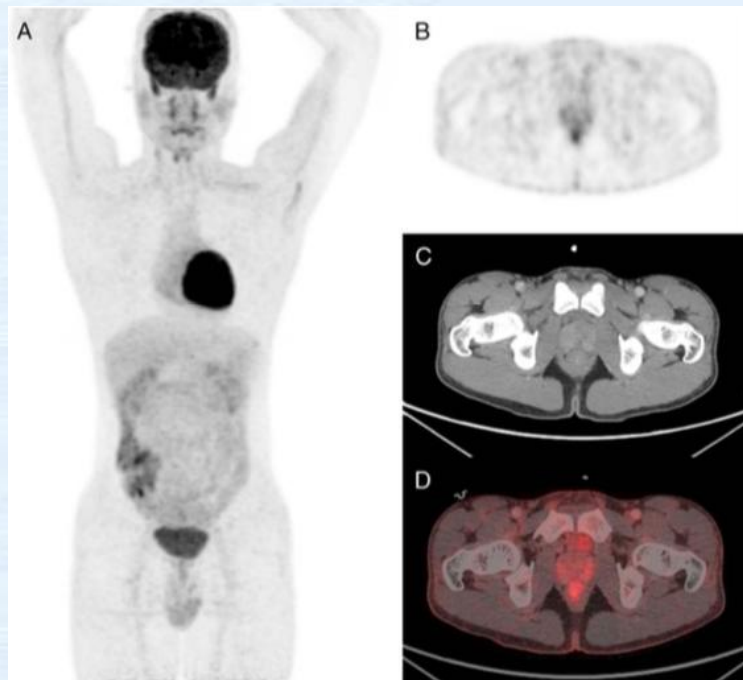
Type I	Morphology resembling Castleman's disease
Type II	Reactive follicular hyperplasia
Type III	Interfollicular plasmacytosis and immunoblastosis
Type IV	Progressive transformation of germinal centers
Type V	Inflammatory pseudotumor

前列腺

男，20岁，不明原因发热、腹痛、体重下降。血清IgG4水平升高



激素治疗前



激素治疗后

小结

- 系统性疾病，好发于老年男性，组织病理学特征为受累组织大量IgG4阳性浆细胞浸润，血清IgG4水平升高。激素治疗有效
- 累及器官可有胰腺、胆管、胆囊、唾液腺、腹膜后、肾、肺、前列腺。一些病例中，可有炎性假瘤（如肝、肺、垂体）
- 诊断需结合临床、生化、影像、组织病理学检查，需与肿瘤鉴别
- PET/CT：有助于诊断、鉴别诊断，指导活检部位；监测疗效



谢谢!