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KARNATAKA RADIOLOGY EDUCATION PROGRAM DR KUSHALI, DNB RESIDENT NH

## CASE I:

## A 31yr old male presented to the OPD with the following symptoms:

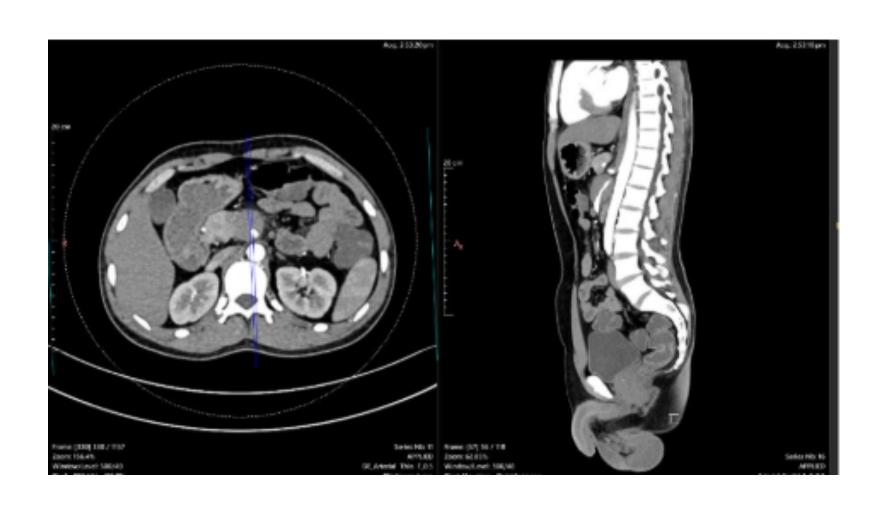
- Dyspepsia- on and off for last 2 months
- No vomiting
- No loose stools or PR bleeding
- Weight loss- 7kgs
- No significant past or family history.

#### **O/E**:

No abnormal signs elicited.

Multiple lab tests done in Bengal revealed no abnormal findings.

### KEY FINDINGS:



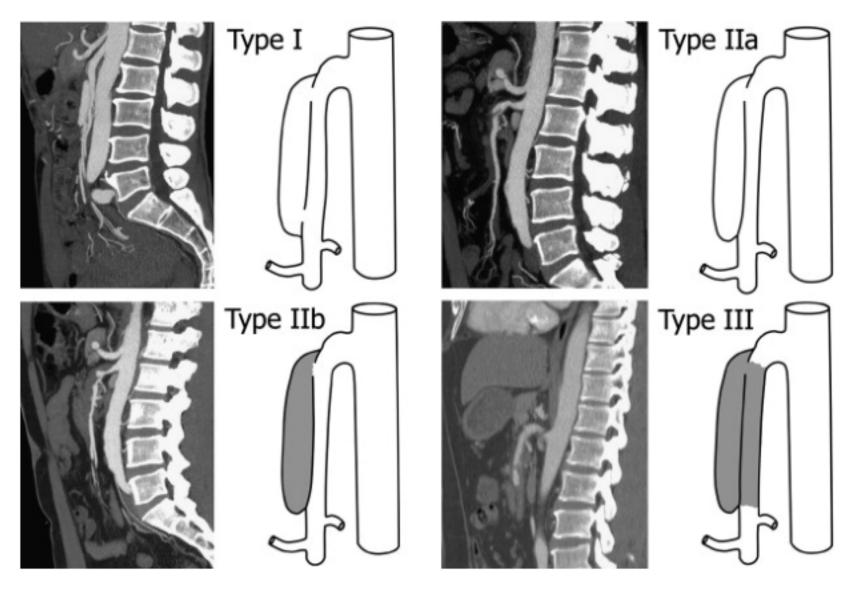
#### DIFFERENTIAL DIAGNOSIS:

- 1. Spontaneous isolated superior mesenteric artery dissection (SISMAD).
- 2. Segmental medial arteriolysis (SAM).
- 3. Perivascular cuffing secondary to manifestation of other disease.
- 4. Mycotic aneurysm.
- 5. Inflammatory aneurysm.

## I. SISMAD (Spontaneous isolated superior mesenteric artery dissection):

- SISMAD occurs mainly in male patients in their fifth decade.
- The underlying causes of SISMAD are not related to atherosclerotic risk factors such as hypertension.
- In patients with SISMAD, after conservative treatment, a benign clinical course is seen, even without anticoagulation or anti-platelet use. No progression of dissection is observed on follow-up CT.
- For patients without signs of acute bowel ischaemia or SMA rupture, conservative treatment without anticoagulation therapy is the first-line treatment.

Coexisting medical conditions	
Smoking (current and ex-smoker)	12 (38)
Hypertension	10 (31)
Intraabdominal cancer <sup>a</sup>	6 (19)
Diabetes mellitus	1 (3)
Ischaemic heart disease	1 (3)
Hypercholesterolaemia	4 (13)
Cerebrovascular disease	1 (3)
Abdominal aortic aneurysm	2 (6)
Marfan's syndrome	1(3)



Angiographic categorisation of SISMAD based on cross-sectional and sagittal views of CT angiography

## II. SEGMENTAL ARTERIAL MEDIOLYSIS:

Original Research | Vascular and Interventional Radiology | February 15, 2018

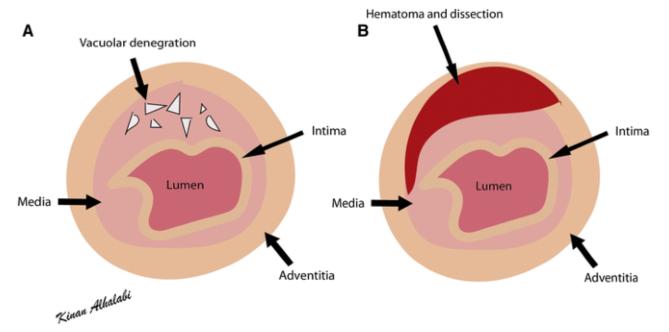
#### Segmental Arterial Mediolysis: Abdominal Imaging of and Disease Course in 111 Patients

Authors: Salien G. Naidu, Christine O. Menias, Rahmi Oklu, Robert S. Hines, Kinan Alhaiabi, Gerges Makar, Fadi E. Shamoun, Stanislav Henkin, and Robert D. McBane | Author INFO & AFFILIATIONS

- Segmental arterial mediolysis (SAM) is a rare noninflammatory vasculopathy that tends to affect the renal, mesenteric, and iliac arteries.
- It is often characterized by dissections, aneurysms, or stenoses that can be found incidentally on imaging or can present acutely with end-organ ischemia or life-threatening bleeding.
- The underlying histological process is lysis of the smooth muscle of the outer media of the arterial wall <sup>5</sup>, resulting in intramural hemorrhage, saccular or dissecting aneurysms, thrombosis and hemorrhage.
- Vs FMD Distribution; Age.
- Vs Mycotic aneurysm Pattern of involvement; segmentation.
- Vs Cystic Medial Necrosis Territory; Association.

Characteristic	Value
Age (y), median (range)	51 (23–87)
Male sex	79 (71)
Medical history	
Hypertension	55 (50)
Hyperlipidemia	36 (32)
Diabetes mellitus	3 (3)
History of smoking	37 (33)
Presentation	
Abdominal pain	82 (74)
Flank pain	23 (21)

Characteristic	No. (%) of Patients
Renal artery	52 (47)
Superior mesenteric artery	51 (46)
Cellac trunk	51 (46)
Number of different arteries involved	
1	48 (43)
2	35 (32)
Imaging finding	
Dissection	95 (86)
Aneurysm	63 (57)





## II. PERIVASCULAR CUFFING SECONDARY TO OTHER PATHOLOGY:

## Perivascular Cuffing as the Sole Imaging Finding of Pancreatic Cancer

Moran Drucker Iarovich MD, Yael Inbar, MD, Sara Apter MD, Eli Konen MD MHA, Eyal Klang MD, and Marrianne Michal Amitai MD

Department of Radiology, Sheba Medical Center, Tel Hashomer, Israel

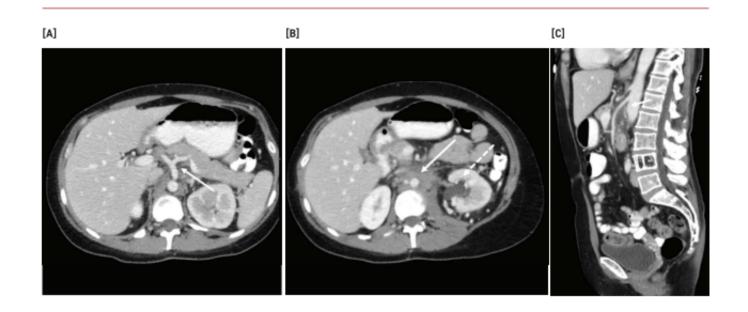
Their study included 14 patients. Key summary points of their findings is summarised below:

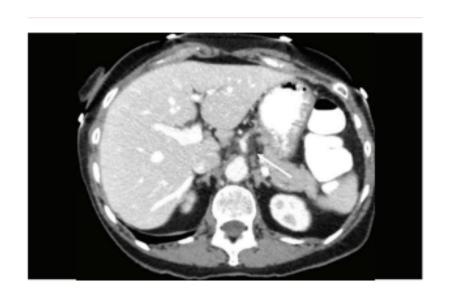
- 1. 3 PDAC patients, 7 Vasculitis patients and 4 RPF patients were included.
- 2. <u>CF</u>: Abdominal Pain 10/14 patients, weight loss 5/24 pts.
- 3. <u>Investigations</u>: None of the patients showed elevated Tumour marker at the time of study (incl CA 19-9). RPF & Vasculitis patients showed: \( \gamma \) APR, \( \gamma \) mildly Complements & + Autoantibodies.

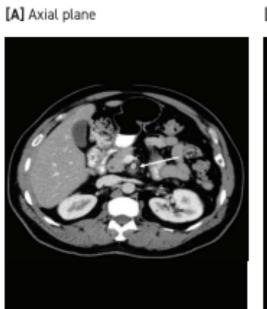
#### 4. Radiology:

- i. PDAC group: Isolated circumferential perivascular cuffing of SMA or coeliac trunk. No pancreatic mass or other specific findings.
- ii. Vasculitis Group: Aortitis + Iliac/ SMA (2/7), Isolated aortitis (1/7), Mid SMA (1/7), coeliac (2/7), Coeliac + proximal SMA( 1/7).
- iii. RPF Group: Iliac & common aorta (4/4), Renal (2/4) & HUN(3/4)
- No statistically significant difference in length of involvement in either of the cases. RPF demonstrated increased wall thickness as compared to other 2 groups.

Group	Age in years	Clinical presentation	Pertinent available laboratory exams	Follow-up in months	Disease course and procedures	Patient outcome
	55	abdominal pain, weight loss, heartburn	amylase 72 IU/I	52	CT, US, EUS, MRI (11/2010-1/2011) negative for a mass. Laparoscopic investigation with intra-surgical US confirmed presence of tumor, followed by a Whipple procedure (02/2011) and chemotherapy	Died due to PDAC
PDAC	72	abdominal pain, constipation	CA 19-9, CEA normal	19	CT, PET CT (11/2015-1/2016) negative for a mass (5/2016) Pulmonary nodule, biopsy was taken confirming PDAC pathology, followed by chemotherapy	Died due to PDAC
	55	abdominal pain, weight loss	CA 19-9, CEA normal, CRP normal, IgG4 normal	16	CT, MRI negative for a mass (9-12/2016). EUS+FNA X2 (latest 5/2017) negative for malignancy. Explorative laparoscopy (7/2017) with tissue sampling surrounding celiac axis confirmed unresectable PDAC. Chemotherapy	Died due to PDAC
	65	back and left flank pain, radiating to groin	WBC 11K, ESR 50, c-ANCA +3	22	Primarily treated as UTI with no relief of symptoms. PET CT on later course of disease demonstrated uptake in lungs and kidneys lesions. Lung lesion biopsy Wegener granulomatosis	Lost to follow- up
	74	History of prostatic cancer with radiation treatment left lower limb swollenness, DVT ruled out	CRP 55.4 mg/l, negative IgG4	12	Family history of autoimmune diseases, including SLE, SAPHO, aortitis. Steroids treatment was initiated, later followed by methotrexate, with good response. Symptoms relapse with treatment discontinuation	Lost to follow- up
	57	Anemia	none	24	Multiple rectal varices on colonoscopy, CT referral to rule out portal vein thrombosis	Lost to follow- up
Vasculitis	71	Right knee monoarthritis, concomitant facial and forearms rash, fever	CRP 106.1 mg/l, elevated liver enzymes, positive ANA, mild rise in C4 41.4 mg/dl, positive anti cardiolipin	18	Steroids treatment was initiated, with good response	Continues follow-up
	65	abdominal pain, nausea, vomiting	WBC 18K, CRP 46.1 mg/l, mildly elevated C3, C4, hemoglobin 9.4 g/dl	18	Initially treated as acute cholecystitis, no relief of symptoms, CT features compatible with vasculitis. Symptoms improved during admission	No recurrence of symptoms
	71	abdominal and back pain	WBC 11K, CRP 72.9 mg/l	20	Symptoms improved during admission, MRI conducted 6 months post-discharge demonstrated resolution of imaging findings	No recurrence of symptoms
	83	abdominal pain, diarrhea. Crohn's disease patient	WBC 6K, CRP 29.5 mg/L	54	Episodic abdominal pain concluded unrelated to underlying Crohn's disease. Improvement under steroids. CT features compatible with vasculitis were consistent during follow-up	Continues follow-up
ş	59	abdominal pain, weight loss	WBC 9K, CRP 100,3 mg/l, negative IgG4, elevated C3 197 mg/dl (top-180)	22	Improvement under steroids treatment	Continues follow-up
Retro-peritoneal fibrosis	31	abdominal and back pain, weight loss	WBC 18K, CRP 156,7 mg/l, mildly elevated d-dimer 436ng/ml	30	Improvement under steroids treatment, later changed to DMARD (mabthera). Later diagnosed with concomitant ankylosing spondylitis	Continues follow-up
Retro-perit	49	abdominal pain, weight loss, lower limb pain, erectile dysfunction	ESR 100, ANA positive, mildly elevated C4 64 mg/dl	28	Improvement under steroids treatment, later changed to DMARD (Imuran+ methotrexate, followed by mabthera)	Continues follow-up, occasional relapses
	79	Abdominal and back pain	WBC 8K, CRP 10 <sub>4</sub> 3 mg/L	72	RPF proven by biopsy	Continues follow-up









[B] Sagittal plane

#### IV. MYCOTIC ANEURYSM:

- Frequently found in atypical locations. The most common sites are:
  - 1. Thoracic and abdominal aorta
  - 2. Abdominal visceral arteries
  - 3. Lower extremity arteries (femoral artery is the most frequently involved and often associated with IV drug abuse)
  - 4. Intracranial arteries: typically more peripheral than Berry aneurysms.

#### • IMAGING:

- 1. Centric aneurysmal sac in an odd location for atheromatous disease
- 2. Often wild, multilobulated appearance
- 3. Interruption of arterial wall calcification
- 4. Adjacent soft tissue stranding
- 5. Adjacent collection +/- gas
- 6. Hazy aortic wall with rupture
- 7. Retroperitoneal para-aortic fluid collection and vertebral erosion
- 8. Thrombus formation within a false lumen after aneurysmal rupture

Mycotic aneurysm of the sup	perior and
inferior mesenteric artery	

inferior mesenteric arte	ery	Streptococcus	47 (18)
Ali Kordzadeh MBBS, MSc, MD ♀ 窗, James Watsor	n BSc, MBBS,	Alpha-hemolytic (viridans) streptococcus	28 (11)
Yiannis P. Panayiotopolous MD, MS, PhD		Staphylococcus	28 (11)
		Candida tropicalis	2.7(1)
Age, years (range)	36 (14-92)	Brucella melitensis	2.7(1)
Gender	73% male (n= 28) vs 11% female (n= 11)	Investigative modality	
Signs and symptoms		CT or CTA	57.8 (22)
Abdominal pain (epigastrium or lower quadrant)	65 (25)	Ultrasonography	23 (9)
Low-grade fever	60 (23)	Intraoperative arteriography	18 (7)
Malaise	26 (10)	Magnetic resonance imaging	5.2 (2)
		Exploratory laparotomy	10.4 (4)
Weight loss	23 (9)	Management	
Nausea and vomiting	20 (8)	Aneurysmectomy without bypass	39 (15)
Anorexia	15 (6)	(Simultaneous bowel resection)	10.5 (4)
Fatigue	13 (5)	Aneurysmectomy with GSV bypass	18 (7)
Lower back pain	10 (4)	Endoaneurysmorrhaphy	10.5 (4)
Postprandial pain	10 (4)	Endovascular embolization	2.6(1)
Joint pain	7.8 (3)	Ligation (distal and proximal)	8(3)
Peritonitis (ruptured)	5.2 (2)	No intervention	2.7(1)
·(·aptaica)	(-)	Autopsy	8(3)

Microorganism

Significant male predominance Median age (36-48) Pentad of abdominal pain, pyrexia of unknown origin (PUO), malaise, weight loss and nausea



#### "Suspect SMA/ IMA Mycotic aneurysm"

- Start board spectrum Anti-Microbial therapy (bactericidal antibiotics over bacteriostatic antibiotics).
  - Obtain blood cultures.
  - 3. Echocardiogram in suspicious cases.
- 4. Computed Tomography angiography (CTA).

#### Inferior mesenteric mycotic artery aneurysm

Superior mesenteric mycotic artery aneurysm

Aneurysmectomy & antimicrobial therapy for 6 weeks. Aneurysmectomy only if viable collateral flow was assess perioperatively & antimicrobial therapy for 6 weeks (upon specimen culture).

"Associated with bowel resection"

Aneurysmectomy and GSV bypass followed by antimicrobial therapy for 6 weeks (upon specimen culture)

"High evidence"

Endovascular approach in selective cases after initiation of antimicrobial therapy for 6 weeks.

"Very few selective cases, evidence is lacking".



#### V. INFLAMMATORY ANEURYSM:

- Inflammation and fibrosis extend into the periaortic tissue and may entrap adjacent retroperitoneal structures such as the ureters or duodenum. There is some overlap with retroperitoneal fibrosis.
- The circumference of the aneurysm wall is involved with inflammation, but for unknown reasons, the periaortic fibrosis is most prominent around the anterior and lateral walls of the aneurysm with relative sparing of the posterior wall.
- Vs MYCOTIC ANEURYSM: Gas in abdominal wall, Saccular Vs Fusiform, Perivascular cuffing/ Mantle sign

#### **TREATMENT:**

- Corticosteroids and other immunosuppressive drugs have been found to decrease symptoms and the degree of periaortic inflammation and fibrosis.
- Definitive treatment of the aneurysm is performed via EVAR or open surgical repair.



# FINAL DIAGNOSIS: INFLAMMATORY PSEUDOTUMOUR

GROSS EXAMINATION	Two grey-white linear cores are identified each measuring 1.5 cm in length, entire tissue is processed in a single paraffin block.		
MICROSCOPIC EXAMINATION	Abdominal mass; Core biopsy:		
	Biopsy cores show a scierosing lesion with lymphohisticcytic infiltrate and rare eosinophils.		
	Focal areas show vague nests of atypical cells with pericellular clearing Several cells show crushing artefacts.		
	Definite RS-like cells are not identified on morphology.		
	Plasma cells are not evident.  Few hypertrophic nerve twigs are noted.		
IMPRESSION	Abdominal mass; Core biopsy:		
	Sclerosing lesion		
	Differentials to consider are several and broad and include neoplasms across lineages.		
	Immunohistochemistry is necessary for further characterization (>10 markers)		
IHC REPORT	ADDENDUM added on 20.01.2025 by Dr. Vidya		
	By immunohistochemistry, EMA and CD138 highlight the plasma cells which are polytypic on kappa and lambda immunostains.		
	CD68, S100, CD3 and CD20 highlight the inflammatory background		
	composed of predominantly T cells and histocytes.		
	Screening with CD30 (performed twice), PAXS and EBV is negative for any definite RS cells.		
	Desmin, Myogenin, p16, Synaptophysin, Pan cytokeratin, Inhibin, SALL-		
	<ol> <li>CD117 do not contribute to any evidence of tumor.</li> </ol>		
	ALK1 is negative.		
	Main differentials considered included inflammatory pseudotumor,		
	IgG-G4 related disease and Nodular sclerosis Hodgkin Lymphoma.		
	There is no evidence of stortform fibrosis, abiliterative phiebitis or unequivocal presence of 85 cells in this biopsy		
	Based on morphology and the above immunohistochemistry results,		
	possibility of an inflammatory pseudotumor may be considered in an appropriate clinical context.		
	Harrison at the both discounts of analysis and allege and allege and		

However, this is a diagnesis of exclusion and clinico-radiological correlation is suggested along with Serum tgG-G4 levels.

Alpha Feto Protein (Afp) ng/mi. jan 18,2025, 8:32 AM	2.02	111
CA -19 -9	<1.4	H
U/mL lan 18,2025, 8:32 AM		
per 10,0002, 0.30 AM		
Carcino Embryonic Antigen (CE	2.14	Ш
ng/mL		
Jan 18,2025, 8:32 AM		
SERUM IGG 4	0.073	Ш
g/l		
Jan 18,2025, 8:32 AM		

## INFLAMMATORY PSEUDOTUMOUR:

- The term "inflammatory pseudotumour" has been used to describe a wide range of reactive and neoplastic lesions:
- 1. Inflammatory myofibroblastic tumour (IMFT)
- 2. Pseudosarcomatous myofibroblastic proliferations of the genitourinary tract,
- 3. Infectious and reparative processes, and
- 4. Inflammatory pseudotumour of lymph node, spleen and orbit.

INFLAMMATORY SPINDLE CELL LESIONS			
Neoplastic	Reactive		
Inflammatory myofibroblastic tumor	Inflammatory pseudotumor		
Epithelioid inflammatory myofibroblastic sarcoma	IgG4-related disease		
Inflammatory fibroid polyp			
Inflammatory well-differentiated liposarcoma			
Inflammatory dedifferentiated liposarcoma			

- Although most articles do not make a difference between them, and IMT is included in the "Inflammatory Pseudotumor" term, IMT and IPT have to be regarded as separate pathological entities.
- Inflammatory pseudotumor: IPT is a reactive lesion, never recurs after resection, and does not metastasize (it has no metastatic potential)
- Inflammatory Myofibroblastic Tumor: IMT is a neoplasm, with a high recurrence rate after excision and low metastatic potential. Shows a wide anatomical distribution.

ALK gene translocation can be detected in 50% of IMTs, but it is not an essential diagnostic criterion

- IMFTs have been reported to occur mainly in children and young adults. IMFTs in the pediatric patient have clinical importance because the lesions often mimic malignant neoplasms, such as sarcomas, lymphomas, or metastases.
- Inflammatory pseudotumour more common affects the lung, where it was considered a reparative postinflammatory condition rather than a neoplastic process. But it has been reported to occur in nearly every site in the body.
- Histologically, the lesion is composed of myofibroblastic spindle cells, accompanied by plasma cells, lymphocytes, and eosinophils.
- Immunohistochemically, the myofibroblastic spindle cells can be positive for vimentin (99%), smooth muscle actin (92%), muscle-specific actin (92%), desmin (69%), cytokeratin (36%), CD68 (KP-1) (24%), and CD30 (Ki-1) (6%). Cytoplasmic reactivity for ALK has also been demonstrated in approximately 50% of these lesions.

Location	Common Imaging Findings	
Lung	Single peripheral, well-defined, lobulated mass with lower lobe predominance, heterogeneous enhancement	
Mediastinum	Mediastinal mass encasing various structures, often heterogeneous	
Trachea and bronchi	Well-defined endoluminal tracheobronchial mass or heterogeneous lobulated or exophytic endoluminal mass	
Heart	Heterogeneous enhancement similar to that of other cardiac tumors	
Orbit	Focal or diffuse enhancing disease involving the orbit; associated retrobulbar fat infiltration or edema, bone destruction, intracranial extension	
Brain	Mass in the brain parenchyma or attached to the dura or both	
Sinonasal cavity	Typically aggressive on CT images with associated osseous involvement including erosion, remodeling, and sclerosis	
Liver	Nonspecific, single or multifocal circumscribed mass	
Spleen	Typically a large, well-circumscribed single mass lesion	
Biliary	Infiltrating hilar lesion and intrahepatic ductal dilatation, similar to cholangio- carcinoma	
Gastrointestinal tract	Ulceration, wall infiltration, and extraluminal extension similar to findings of malignancy	
Mesentery	Well-circumscribed or an infiltrative ill-defined mass extending to adjacent bowel; variable enhancement (no enhancement, heterogeneous or peripheral enhancement)	
Urinary bladder	Polypoid enhancing intraluminal mass, submucosal mass with or without perivesicular fat involvement	
Kidney	Low-attenuation mass on CT images, hypoechoic or hyperechoic mass on ultrasound images	
Adrenal gland	Nonspecific solid mass	