ID data

- Sex: female
- Age: 46y/o
- Birthday: 1955/10/13

Chief Complain

- Right upper quadrate abdominal tenderness for one month.

Present illness (1)

- This 46 years old female patient was in a healthy condition until one month ago when she suffered from RUQ abdominal tenderness. She went to the nephrology OPD for help on 2000/12/20, and the abdominal ultrasound was arranged. The right renal tumor (79*59mm) was noted, and then she was referred to the urology OPD for further evaluation.

Present illness (2)

- At urology OPD, the IVP and abdominal CT were arranged. The IVP showed that downward displacement of the right kidney due to upper pole renal tumor or right adrenal mass, and the abdominal CT revealed that a well-defined soft tissue mass with heterogeneous contrast enhancement at the medial aspect of the right upper pole kidney, so she was admitted to our ward for further evaluation and management.

Present illness (3)

- Throughout whole course of disease, no fever, no nausea, and no vomiting were found. The pain didn't relate to the meal. Besides, there were no Murphy’s sign and CV angle knocking pain.

Past Medical History

- Hypertension: denied
- Diabetes mellitus: denied
- Drug and food allergy: denied

Past Surgical History

- Mixed hemorrhoid s/p hemorrhoidectomy on 2000/01/31 at TMUH.
- Breast fibroadenoma s/p excision years ago.

Personal History

- Smoke: denied
- Drink: denied
- Betel nuts: denied

Family History

- Not contributory

Physical Examination

- Consciousness: clear
- HEENT: grossly normal
- Chest: breathing sound clear
- Heart: RHB without murmur
- Abdomen: RUQ tenderness, no rebounding pain
- Back: no CV angle knocking pain
- Extremity: freely movable, no pitting edema

Laboratory Data

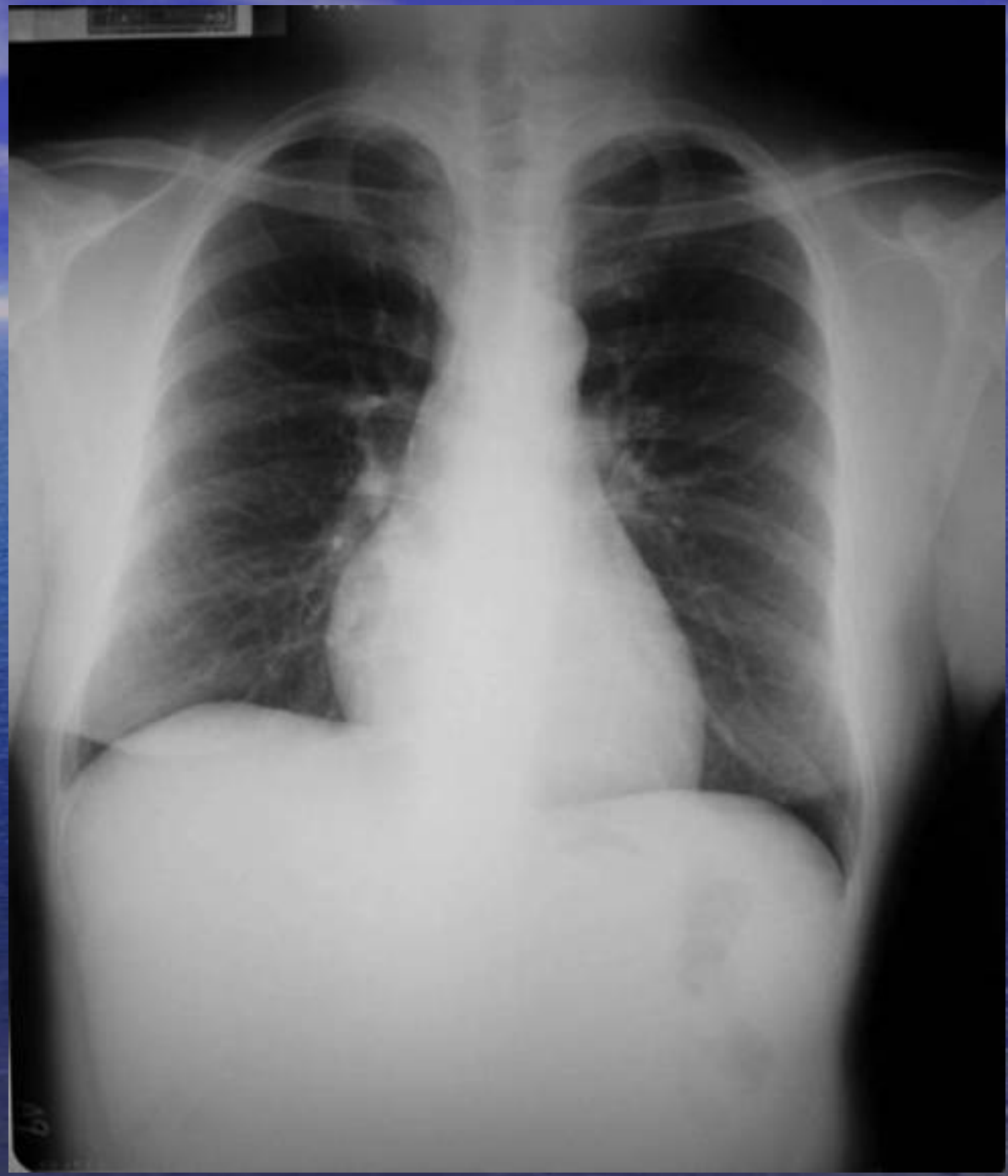
- CBC/DC: WNL
- SMA: WNL
- U/A: WNL

Image study

- 2000/12/23 Abdominal Ultrasound
- 2000/12/23 IVP
- 2000/12/28 Abdominal CT
- 2001/01/02 Angiography

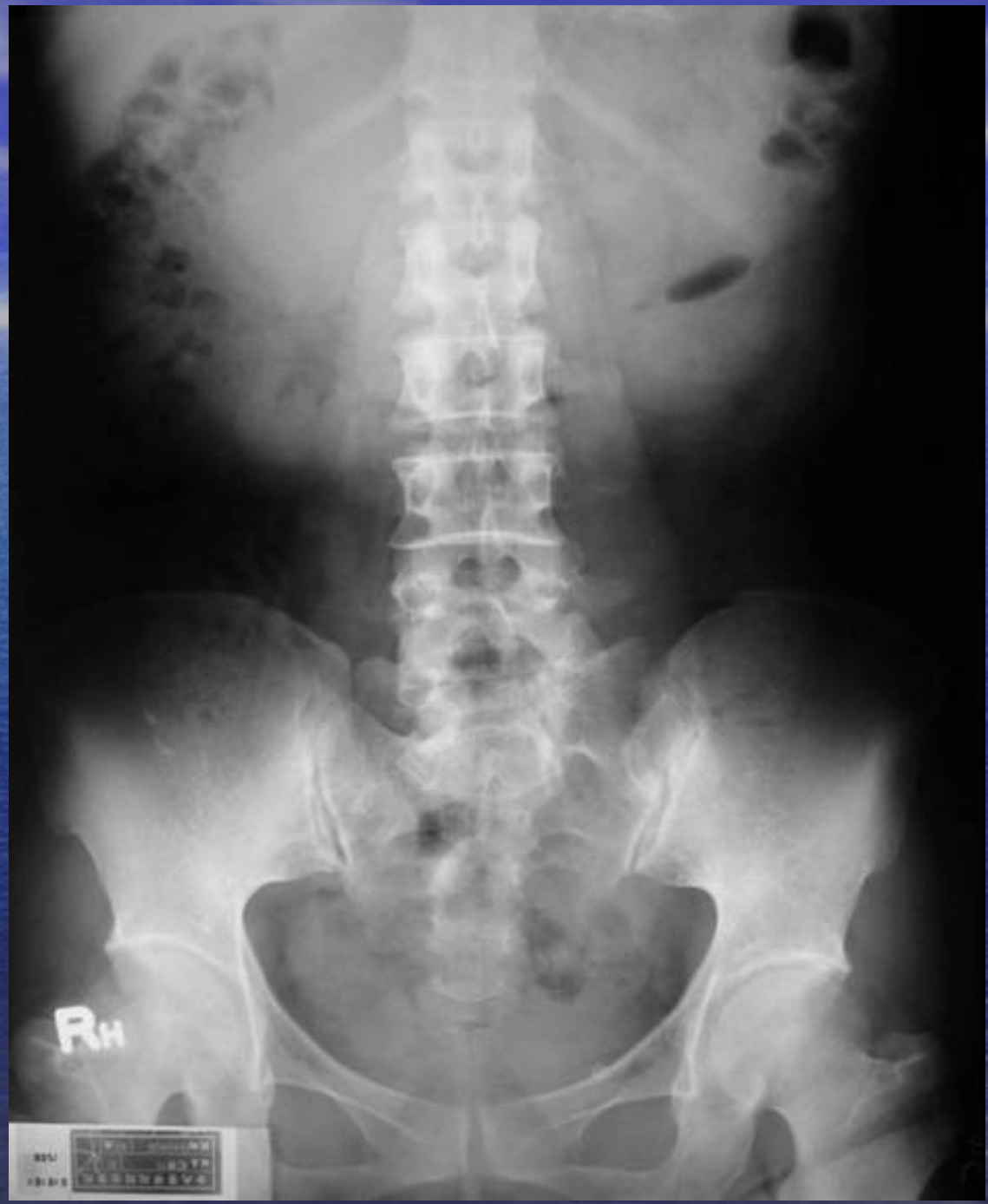
CXR

- No definite active lung lesion, no significant abnormality in heart and diaphragm.

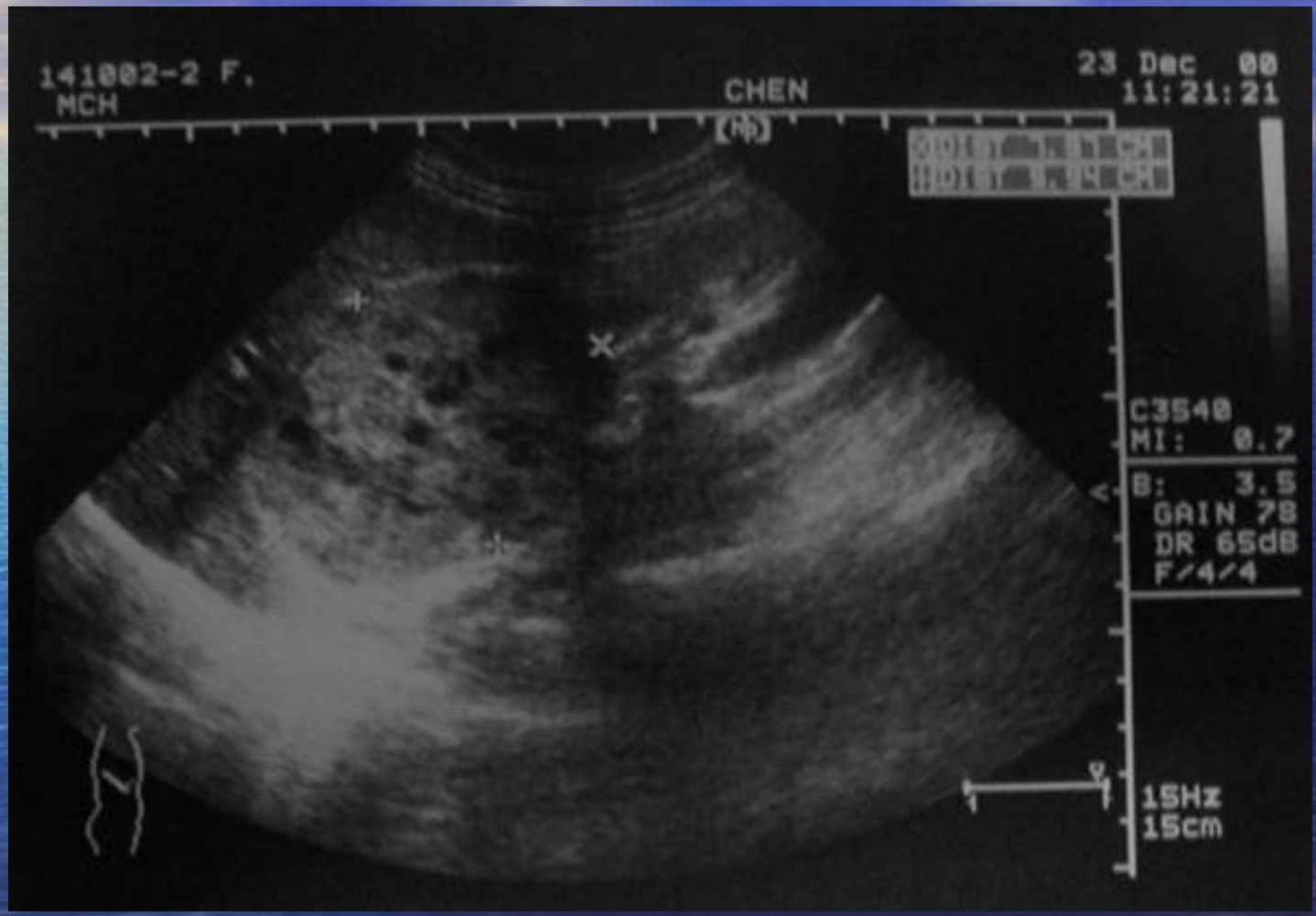


KUB

- There are well visualization of the bilateral psoas line with no other particular findings.

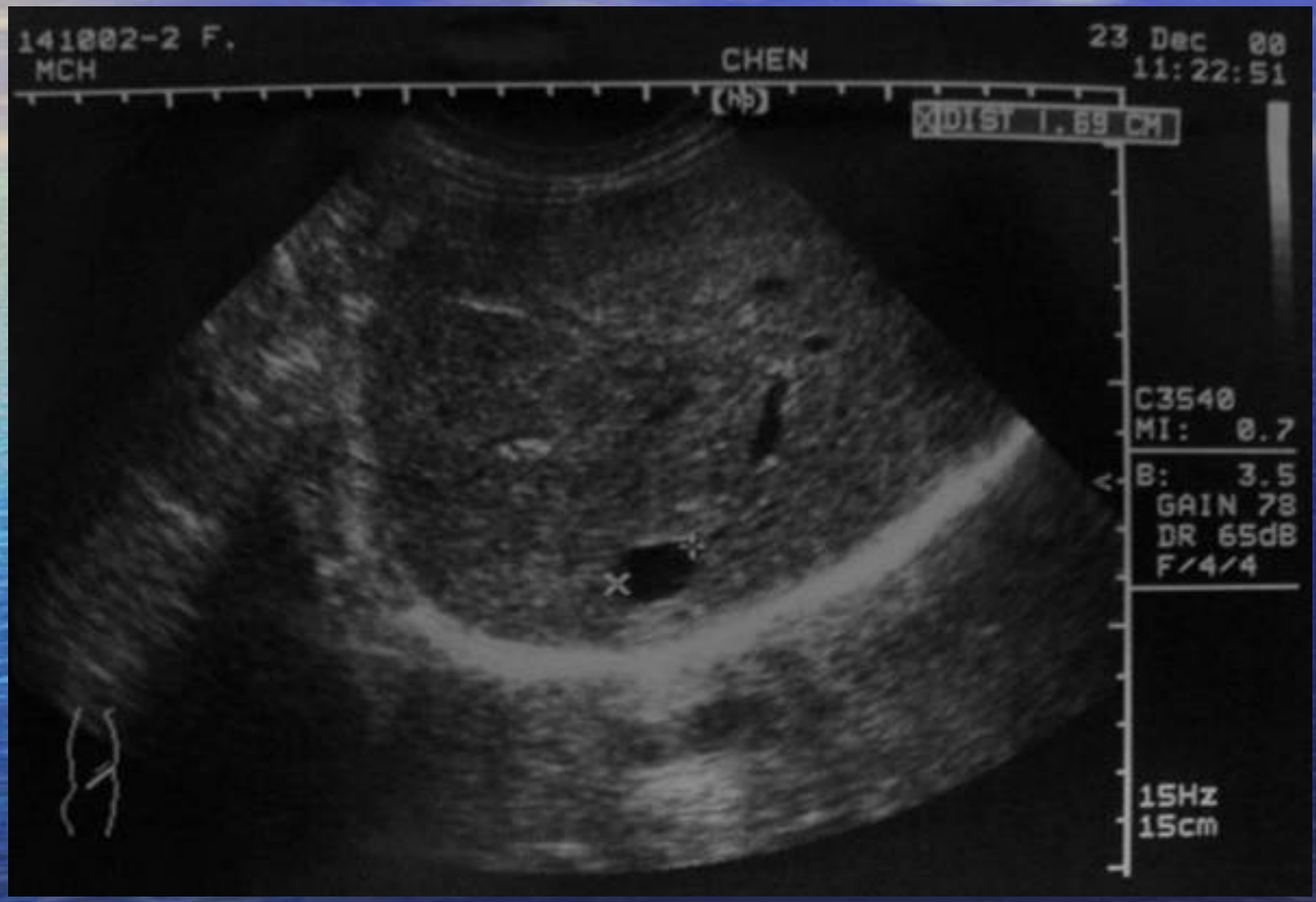


Abdominal Ultrasound 2000/12/23 (1)



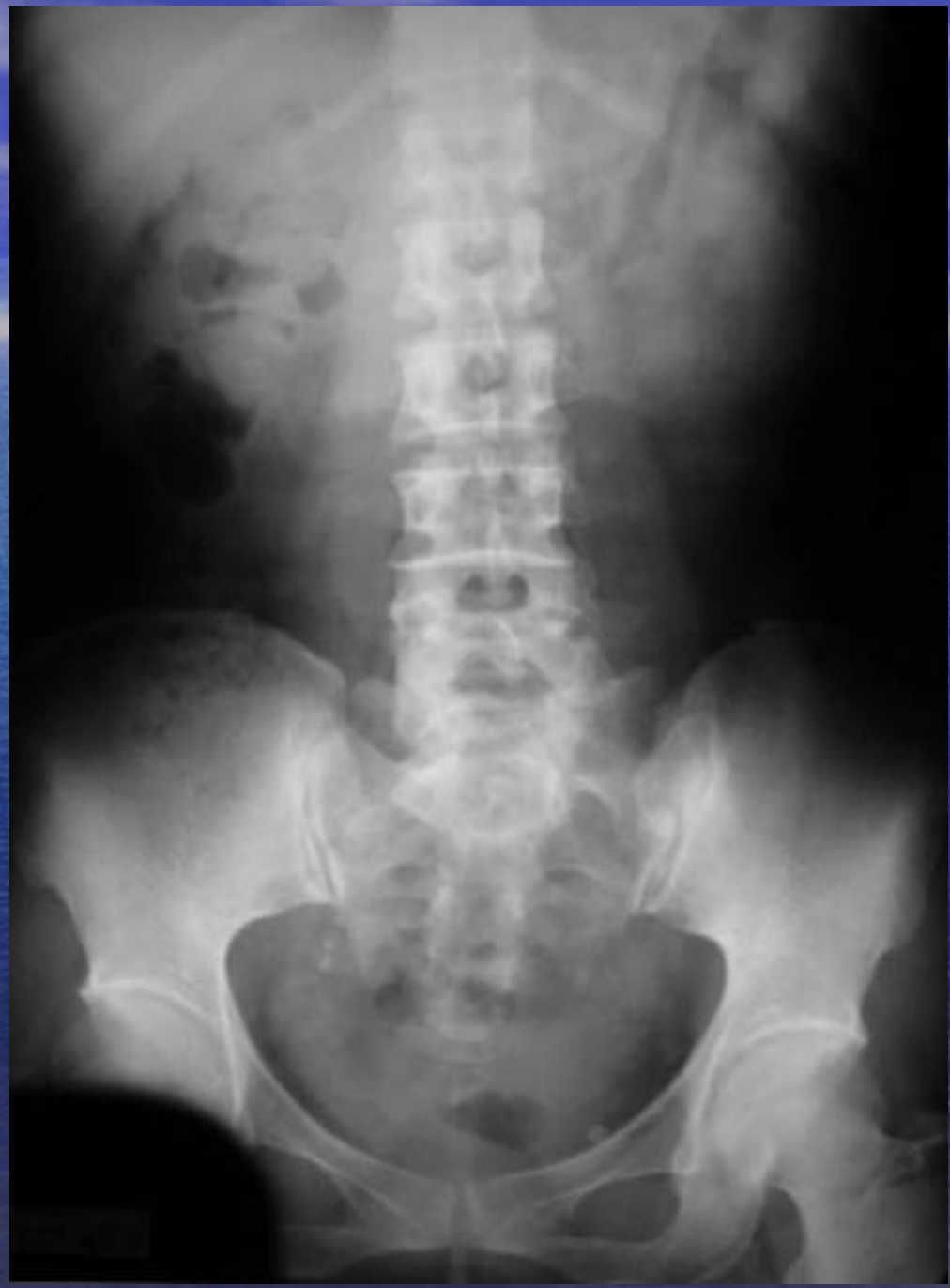
- A isoechoic tumor, diameter 79*59mm, was noted at upper pole of right kidney.

Abdominal Ultrasound 2000/12/23 (2)

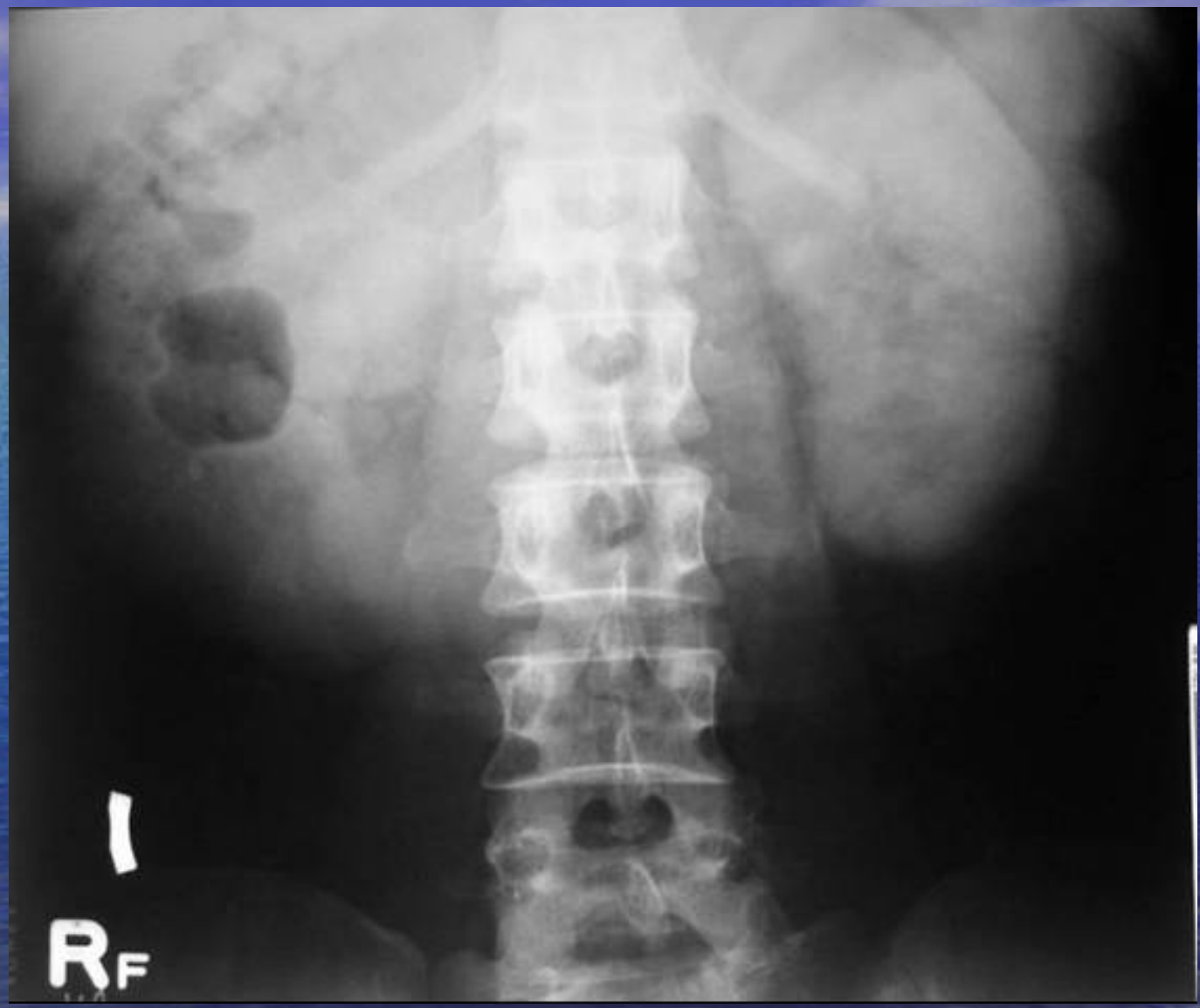


- A cystic lesion in right lobe, diameter 17mm.

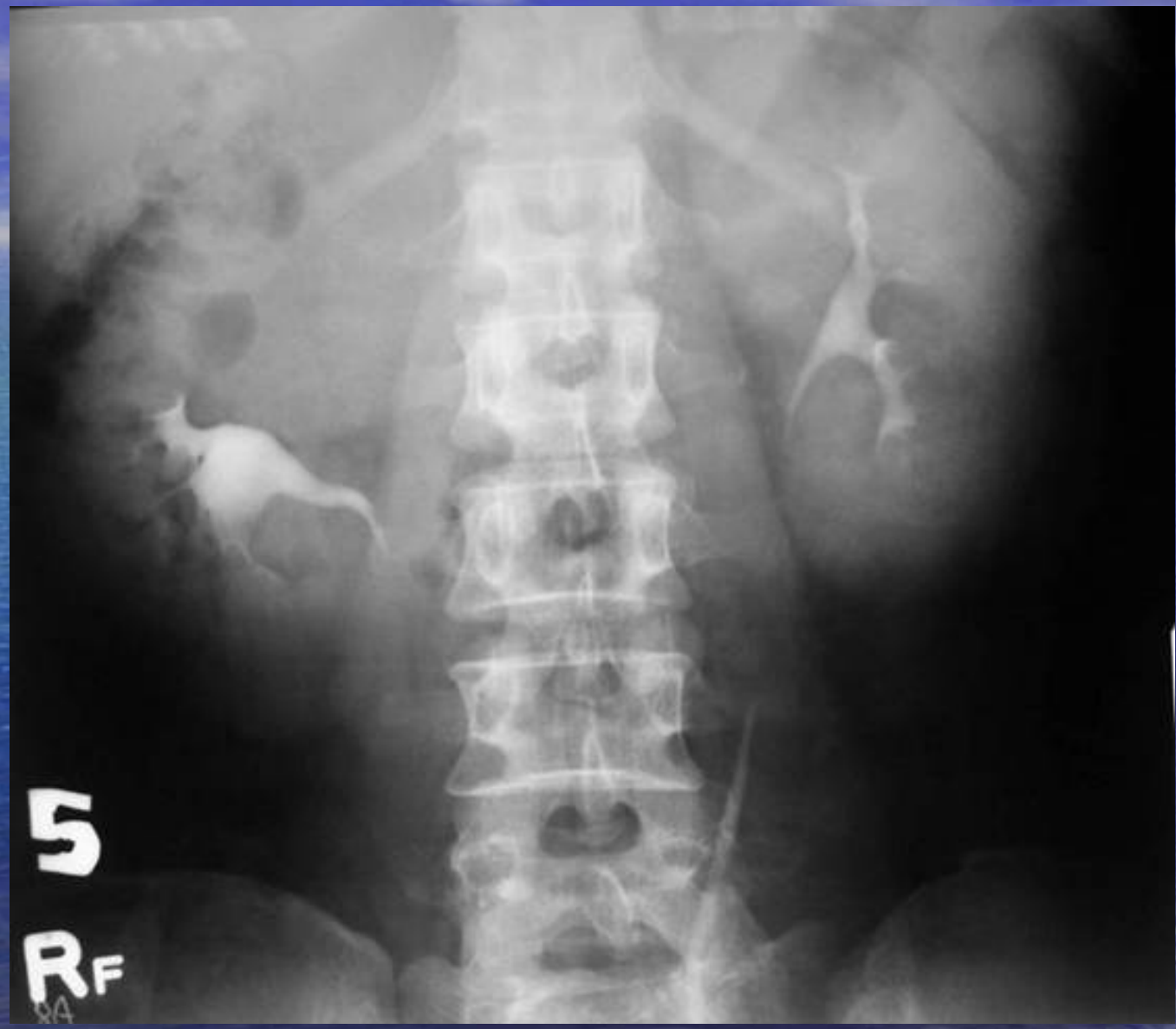
IVP (1)



IVP (2)

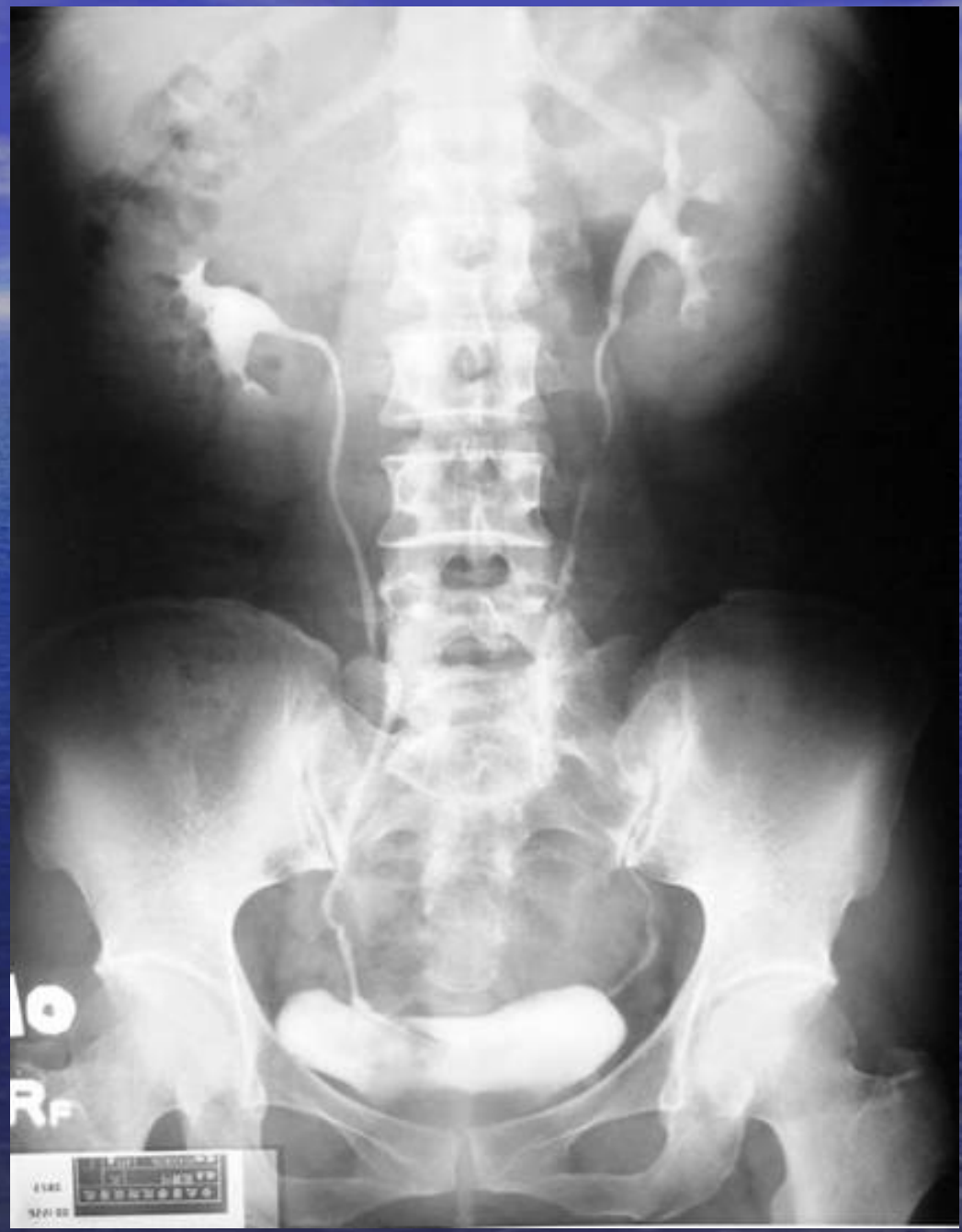


IVP (3)

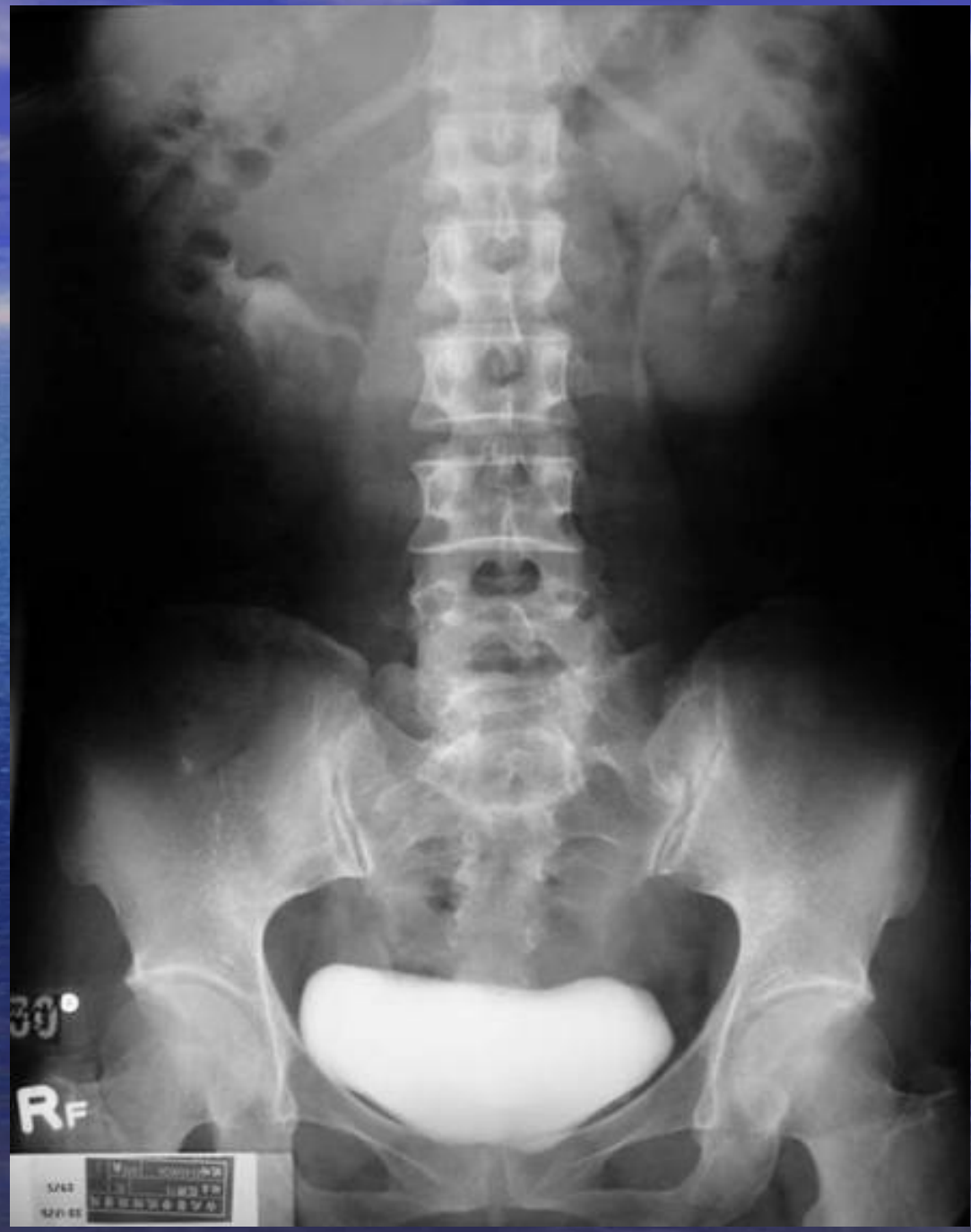


IVP (4)

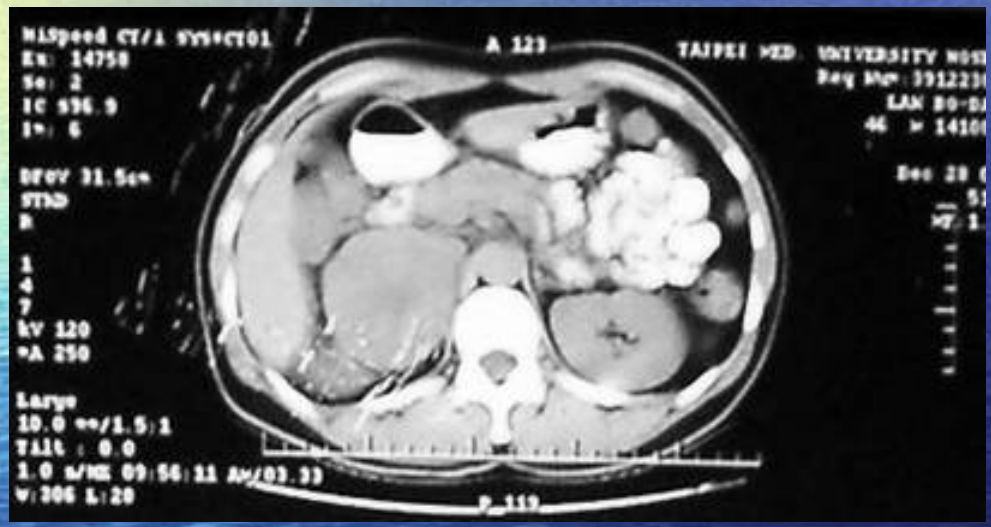
- Downward displacement of the right kidney due to upper pole renal tumor or right adrenal mass

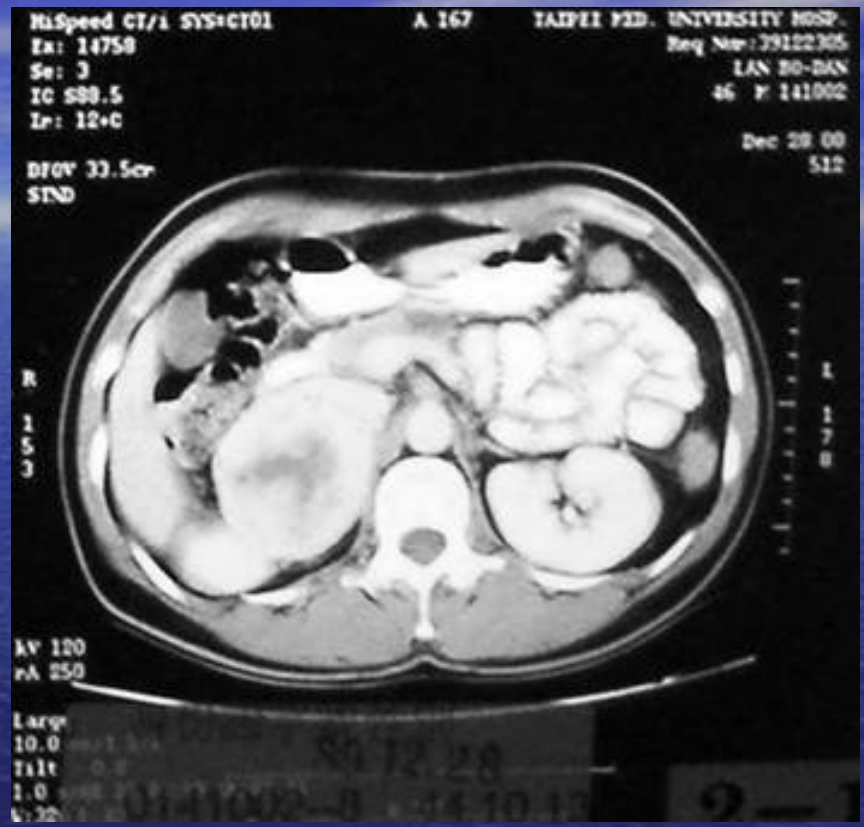


IVP (5)



Abdominal CT 2000/12/28 (1)



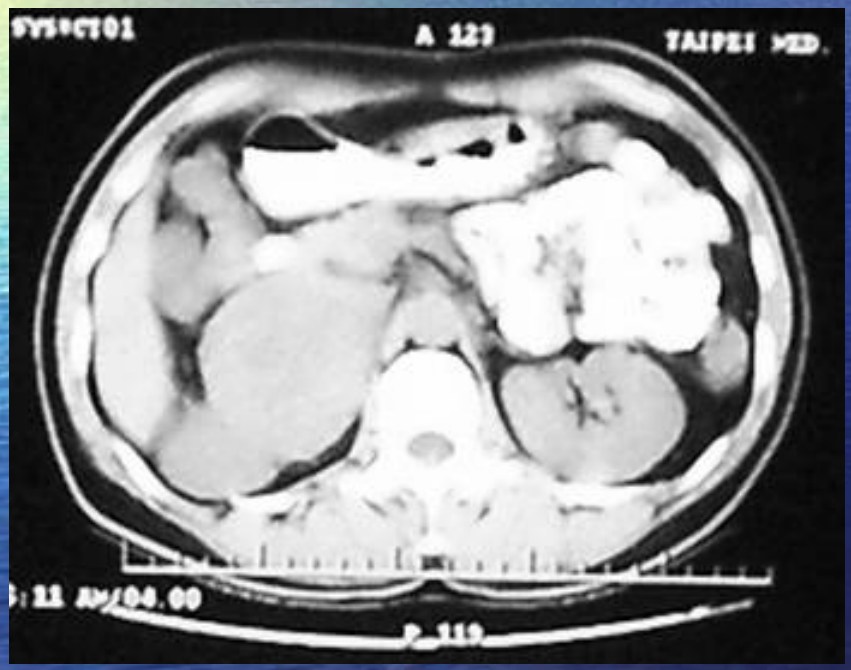


Pre-enhanced

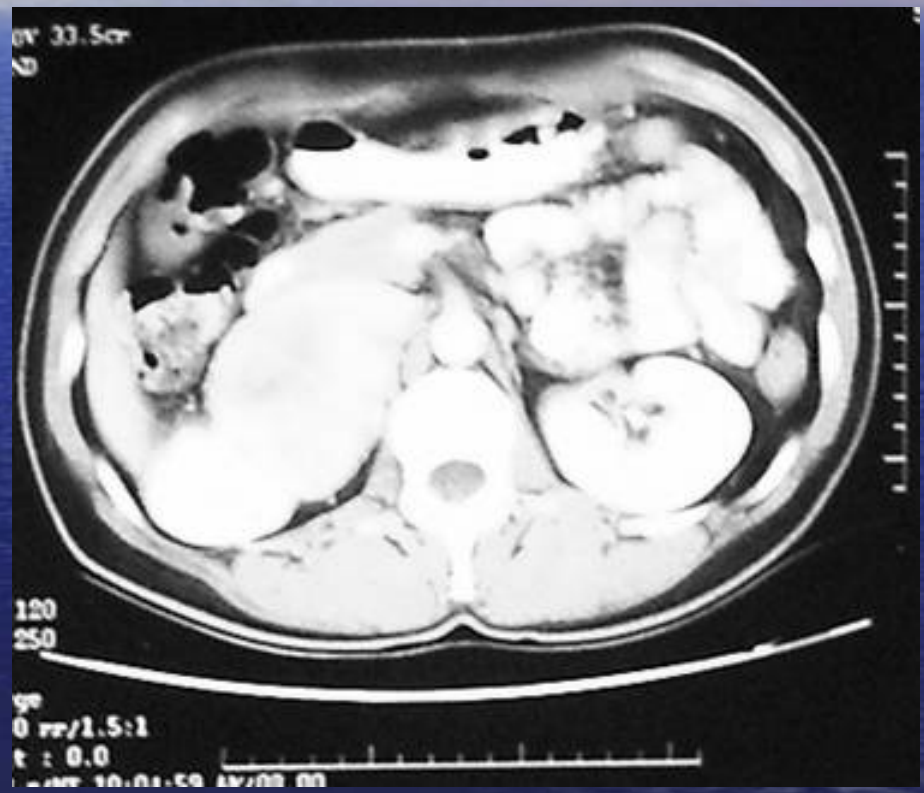
Post-enhanced

- A well-defined soft tissue mass (8*7cm) with heterogeneous contrast enhancement (central necrosis) was noted within the right renal fossa at the medial aspect of the right upper pole kidney.

Abdominal CT 2000/12/28 (2)

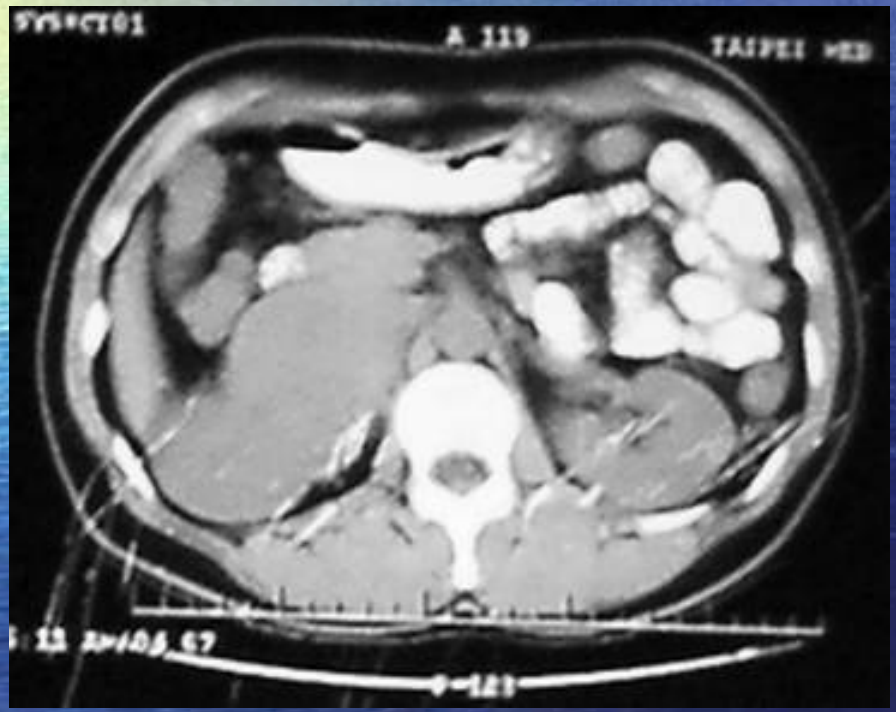


Pre-enhanced

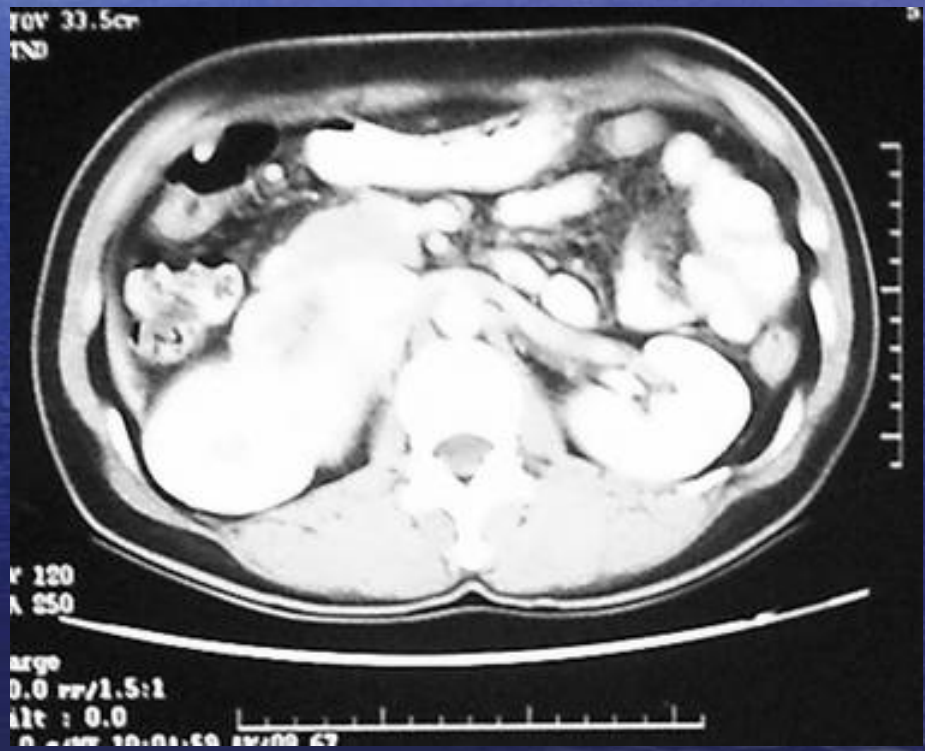


Post-enhanced

Abdominal CT 2000/12/28 (3)

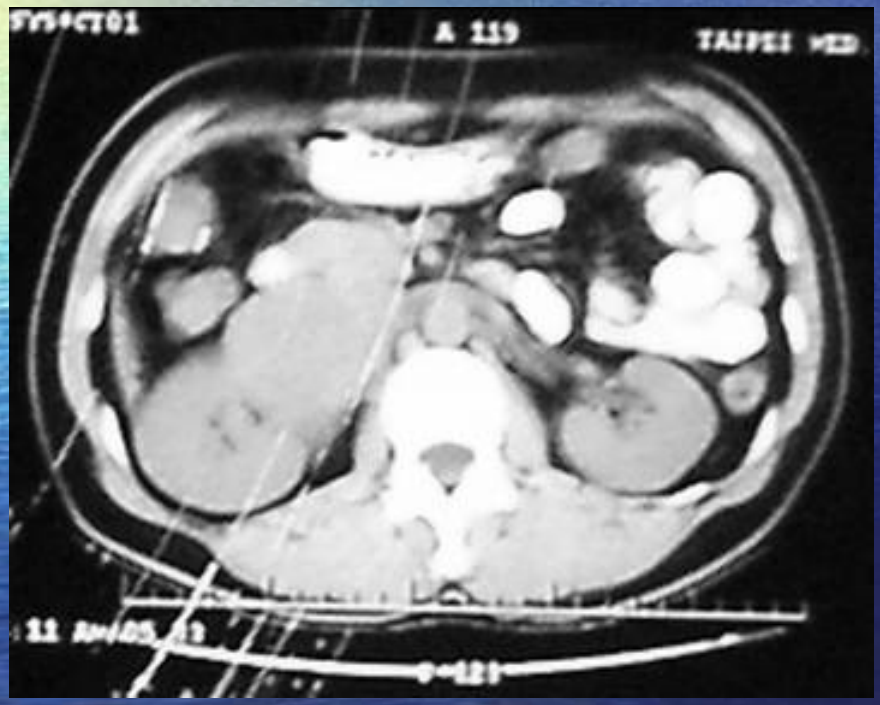


Pre-enhanced

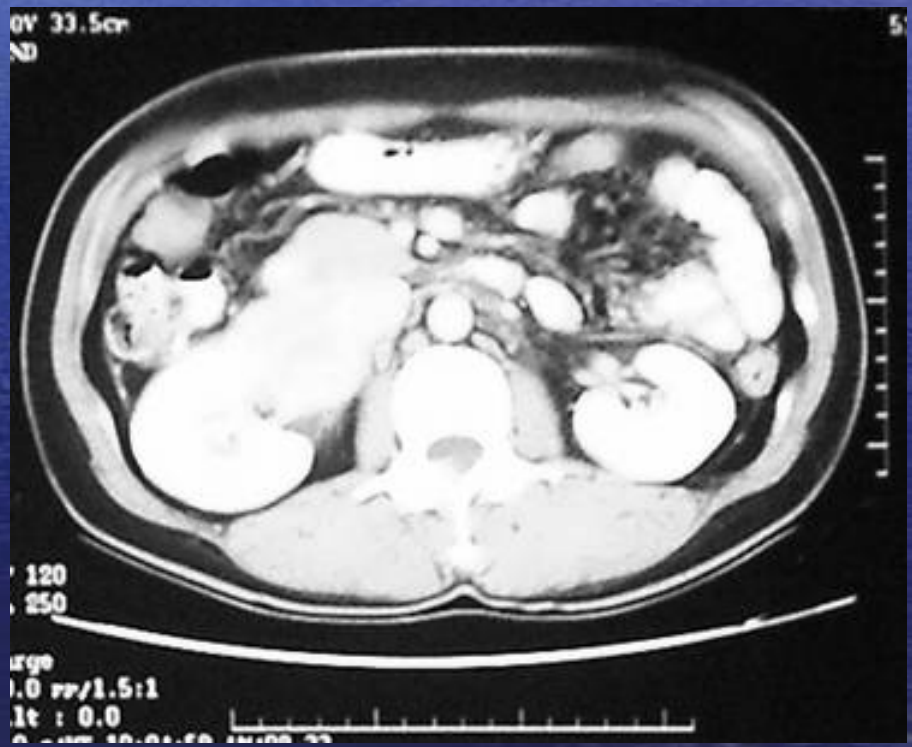


Post-enhanced

Abdominal CT 2000/12/28 (4)

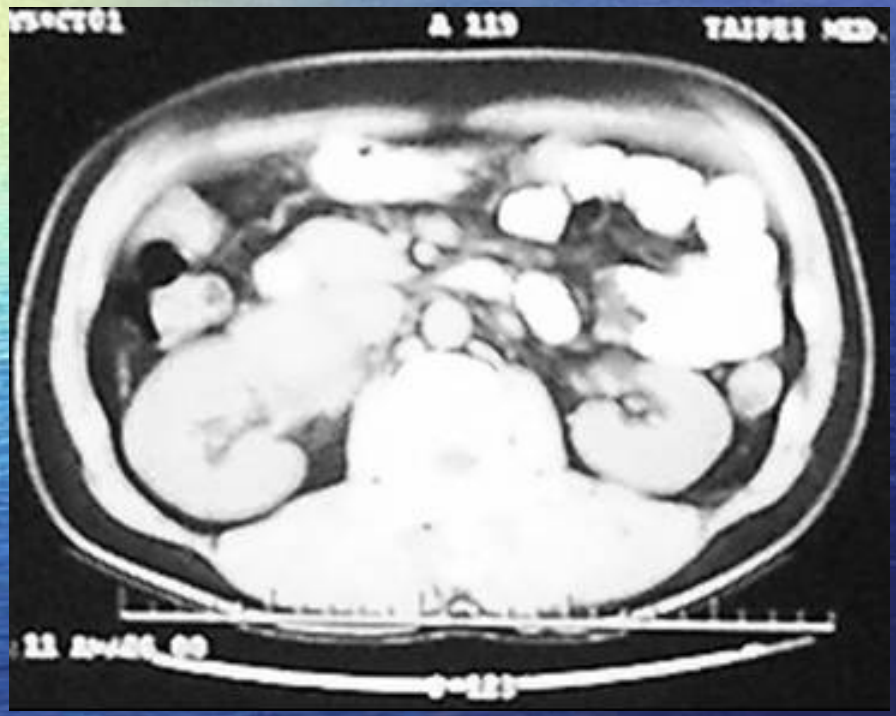


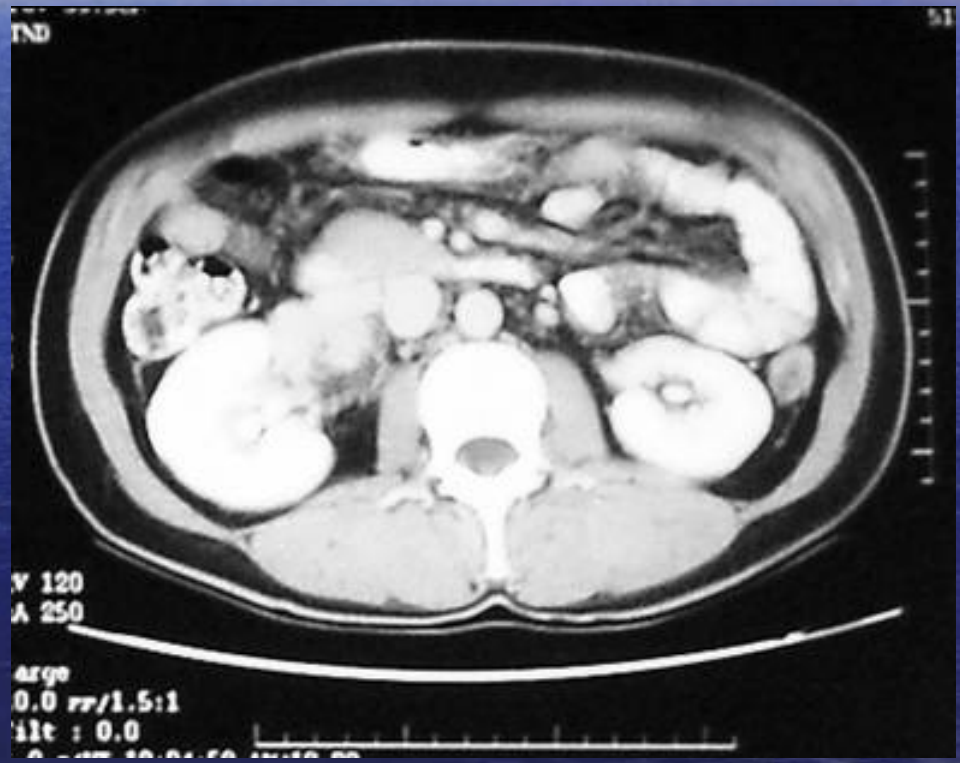
Pre-enhanced



Post-enhanced

Abdominal CT 2000/12/28 (5)

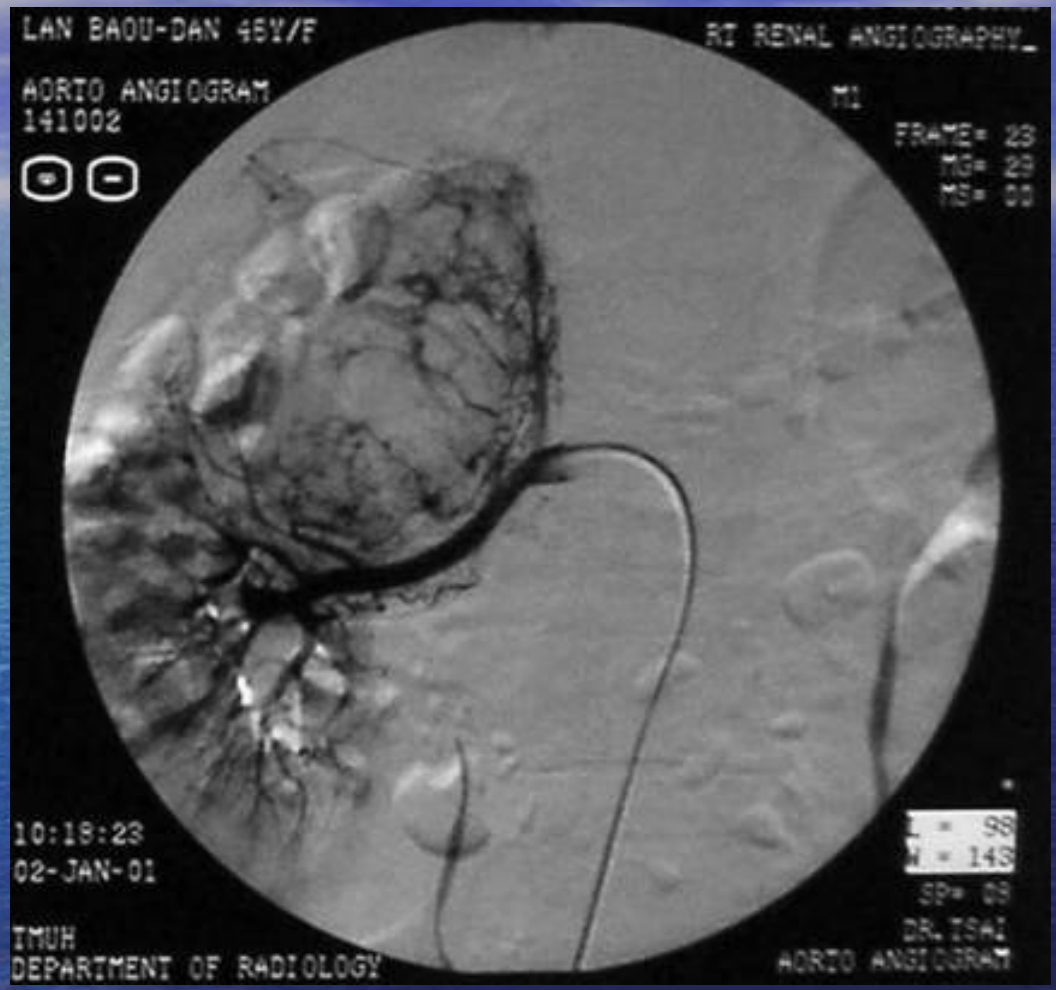




Pre-enhanced

Post-enhanced

Angiography 2001/01/02



- A hypervascular tumor mass noted at right suprarenal region, which is mainly supplied from the inferior adrenal artery of right renal artery.

Summary of image finding (1)

- CXR:
  - No definite active lung lesion
  - No significant abnormality in heart and diaphragm
- KUB:
  - Well visualization of bilateral psoas lines
  - No other particular finding
- Abdominal Ultrasound:
  - A isoechoic tumor, diameter 79*59mm, was noted at upper pole of right kidney.
  - A cystic lesion in right lobe, diameter 17mm.

Summary of image finding (2)

- IVP
  - Downward displacement of the right kidney due to upper pole renal tumor or right adrenal mass
- Abdominal CT
  - A well-defined soft tissue mass with heterogeneous contrast enhancement was noted within the right renal fossa at the medial aspect of the right upper pole kidney.
- Angiography
  - A hypervascular tumor mass noted at right suprarenal region, which is mainly supplied from the inferior adrenal artery of right renal artery.

Differential Diagnosis

- Renal cell carcinoma
- Pheochromocytoma
- Adrenal adenoma
- Adrenal carcinoma
- Adrenal metastasis

Renal cell carcinoma (1)

- Abdominal Ultrasound
  - Isoechoic, hyperechoic, hypoechoic relative to the renal parenchyma
  - Primarily to differentiate solid masses from simple cysts

Renal cell carcinoma (2)

- IVP
  - Mass effect on the collecting system
  - Distort of the renal contour
  - Enlargement of a portion of the kidney
  - Calcification
  - Most RCCs are less attenuating than surrounding renal parenchyma

Renal cell carcinoma (3)

- CT
  - Pre-enhanced
    - May isoattenuating, hypoattenuating, hyperattenuating relative to the kidney
    - Amorphous, internal calcification
  - Post-enhanced
    - Usually solid, necrosis
    - Sometimes cystic mass, with thick septa and wall nodularity
    - May completely solid and highly enhancing mass

Renal cell carcinoma (4)

- Angiography
  - Noninvasive cross-sectional imaging has replaced angiography
  - Occasionally use if the origin of a tumor (renal v.s. adrenal) is not certain. In these patients, selective injection of the renal and adrenal arteries may be necessary

Pheochromocytoma (1)

- Abdominal Ultrasound
  - Ultrasound has largely been replaced by CT and MRI, and it is limited as a result of the effects of overlying bowel gas, especially the left adrenal gland
  - For differentiating cystic lesions from solid lesions in the adrenal gland
- IVP
  - Large adrenal masses may compress and deform the upper pole of the kidney

Pheochromocytoma (2)

- CT
  - Large tumors (often $>$ 3cm)
  - Round or oval masses
  - Necrosis, hemorrhage, and fluid-fluid levels
  - Inhomogeneous
  - Calcification is rare
- Angiography
  - increased vascularity in the tumors

Adrenal adenoma (1)

- Abdominal Ultrasound
  - No specific finding for adrenal adenoma
  - Ultrasound of the adrenal glands may be performed to evaluate abdominal masses in infants and children
- IVP
  - Old adrenal hemorrhage or commonly calcified adrenal neoplasm
  - No significant role in the diagnosis of adrenal adenoma

Adrenal adenoma (2)

- CT
  - Well-circumscribed mass lesions
  - Homogeneous
  - Heterogeneous can be observed when a lipid-rich adenoma and a lipid-poor adenoma coexist
- Angiography
  - No specific finding

Adrenal carcinoma (1)

- Abdominal Ultrasound
  - Homogeneous when small
  - Heterogeneous with cystic areas when the tumor grows as a result of hemorrhage and necrosis
- IVP
  - Mass effect on the ipsilateral superior pole of the kidney, usually displacing the upper pole of the kidney laterally and, when large enough, inferiorly

Adrenal carcinoma (2)

- CT
  - Large mass ($>$ 5cm)
  - Central necrosis or hemorrhage
  - Heterogeneous enhancement (greatest at the periphery)
  - Invasion in to adjacent structures
  - The imaging findings of large pheochromocytoma and metastasis may be identical

Adrenal carcinoma (3)

- Angiography
  - Hypovascular masses
  - The predominant arterial supply to the adrenal gland and to the adrenal carcinoma is the superior adrenal artery of the inferior phrenic artery

Adrenal metastasis (1)

- Abdominal Ultrasound
  - Solid lesions with heterogeneous echogenicity
  - Hypoechoic
- IVP
  - Plain-film imaging of adrenal masses is limited
  - Large masses often are indistinguishable from renal lesions

Adrenal metastasis (2)

- CT
  - Focal masses or distortion of the contour of the adrenal gland
  - Homogeneous when $<$ 3cm
  - Heterogeneous when larger lesions with central necrosis or hemorrhage
  - May invade contiguous organs such as kidney
- Angiography
  - It is not useful in the diagnosis

Surgical intervention

- 2001/01/05
  - Right nephrectomy
  - Right adrenectomy
  - Pre-op diagnosis: R/O adrenal tumor

Pathology

- Paraganglioma
  - The tumor shows a picture of paraganglioma composed of polygonal and spindle shaped tumor cells arranged in solid nests invested by interconnecting hypervascular stroma

Discussion (1)

- Background
  - The tumors arise from the chromaffin cells of the adrenal medulla and are associated with increased catecholamine production.
  - Pheochromocytoma: arise from adrenal gland
  - Paraganglioma: arise from other location (ex: mediastinum, along the aorta, and in the pelvis)

Discussion (2)

- Detecting the tumors is important
  - HTN is cured with the removal of the tumor
  - Familial disorder
  - 10% of pheochromocytoma are malignant
- 10% tumor
  - 10% risk of malignancy
  - 10% of the tumors are bilateral (sporadic)
  - 10% are extra-adrenal
- Early detection may reduce the risk of metastasis

Discussion (3)

- Pathophysiology
  - 80~90% are sporadic
  - Familial disorder
    - MEN IIa: AD, pheochromocytoma, medullary thyroid carcinoma, parathyroid hyperplasia or adenoma
    - MEN IIb: AD, pheychromocytoma, medullary thyroid tumor, mucosal neuroma
    - Neurofibromatosis
    - Pheochromocytoma may be inherited, as they are 10% of patients. Familial cases these tend to occur in younger patients

Discussion (4)

- Tumors are larger than 3cm when seen
- They are highly vascular, and larger tumors are prone to hemorrhage and necrosis, even they are benign
- Malignancy is depends on the metastasis
- Metastasis have been reported in the lymphatic tissue, lung, liver, bones, and brain
- Prognosis affective facotr
  - Vascular invasion
  - Local or distant metastasis
  - DNA ploidy (diploidy is more benign)
- The risk of malignancy is lower in patients with familial tumors than in patients with sporadic tumors

Discussion (5)

- Frequency: the true incidence is unknown
- Mortality/Morbidity: although rare, pheochromocytomas can result in serious morbidity or mortality if they remain unrecognized.
  - The most severe complication is pheochromocytoma crisis
    - Shock, DIC, seizures, rhabdomyolysis, acute renal failure, and death
  - Other complication
    - Reactivation of Graves disease or transient thyrotoxicosis
    - hypercalcemia due to tumor secretion of a parathyroid hormone-related protein
    - Noncardiogenic pulmonary edema
    - Acute abdomen
    - Renal infraction
- The high risk of provoking a hypertensive crisis during the manipulation of an adrenal gland

Discussion (6)

- Race: no racial predilection
- Sex: the ♂-to-♀ ratio is almost equal
- Age:
  - Sporadic: 30~50 y/o
  - Familial disorder: in children

Discussion (7)

- Clinical details
  - Episodic or sustained hypertension
  - Triad
    - Headache
    - Palpitation
    - diaphoresis
  - Post-voiding loss of consciousness
  - Indication for imaging of the adrenal gland
    - New or worsening DM due to impaired glucose regulation
    - Hypertensive crisis after anesthesia, surgery, or treatment with medication
    - History of multiple endocrine problems
  - Laboratory tests
    - Serum or urine catecholamine levels
    - Urinary VMA and metanephrine levels

Discussion (8)

- Preferred Examination
  - CT: quick and relatively inexpensive, and it offers good spatial localization
  - MRI: more specific for pheochromocytoma than CT, but some patients cannot tolerate it
  - ^{131}I-MIBG: lower sensitivity, but it is useful for detection of extra-adrenal tumors and metastatic deposits

Discussion (9)

- X-ray
  - Limited value
  - IVP: large adrenal masses may compress and deform the upper pole of the kidney incidentally may be discovered

Discussion (10)

- CT
  - Findings
    - Large tumor (often $>$ 3cm)
    - Round or oval masses
    - Necrosis, hemorrhage, fluid-fluid levels
    - Inhomogeneous
  - Administration of the contrast agent may precipitate a hypertensive crisis in an unmedicated patient

Discussion (11)

- MRI
  - Findings
    - T1-weighted image
      - Hyopintense or isointense relative to the liver
    - T2-wieghted image
      - Highly intense
  - MRI obtained with gadolinium-diethylnetriamine pentaacetic acid (DTPA), tumors demonstrate brisk and prolonged enhancement
  - In 20~33% of patients, T2-weighted images show atypical finding
  - Comparable high signal intensity may been in some necrotic adrenal metastasis and adrenal cysts. These lesions cannot always be distinguished from pheochromocytoma on MRI

Discussion (12)

- Ultrasound
  - It is limited as a result of the effects of overlying bowel gas, especially in the left adrenal gland
  - It is limited to differentiate cystic lesions from solid lesions in the adrenal gland
- Angiography
  - Increased vascularity in the tumor
  - Angiography is hazardous without premedication, and a hypertensive crisis can result

Discussion (13)

- Intervention
  - Prompt surgical referral for excision because patients are at significant risk for lethal complication such as hypertensive crisis and adrenal hemorrhage
  - Biopsy does not need to performed, and it can be dangerous because hypertension is triggered by direct manipulation of adrenal gland