General History(1)

Mr. 邱 was referred to our URO OPD by Ching-May hospital on 2003/02/08 for performing pelvic CT scanning.

General History(2)

Mr. 邱 was born in Taipei on 1972/12/27 and is still dwelling in Taipei now. He is a business man and had had married for once but had divorced.

According to Mr. 邱 himself, he had had been quite well before, until half year ago prior to his coming to our hospital, when urination difficult first developed.

General History(3)

At the first few months of the illness, he still was able to urinate though it had become more and more difficult to make it.

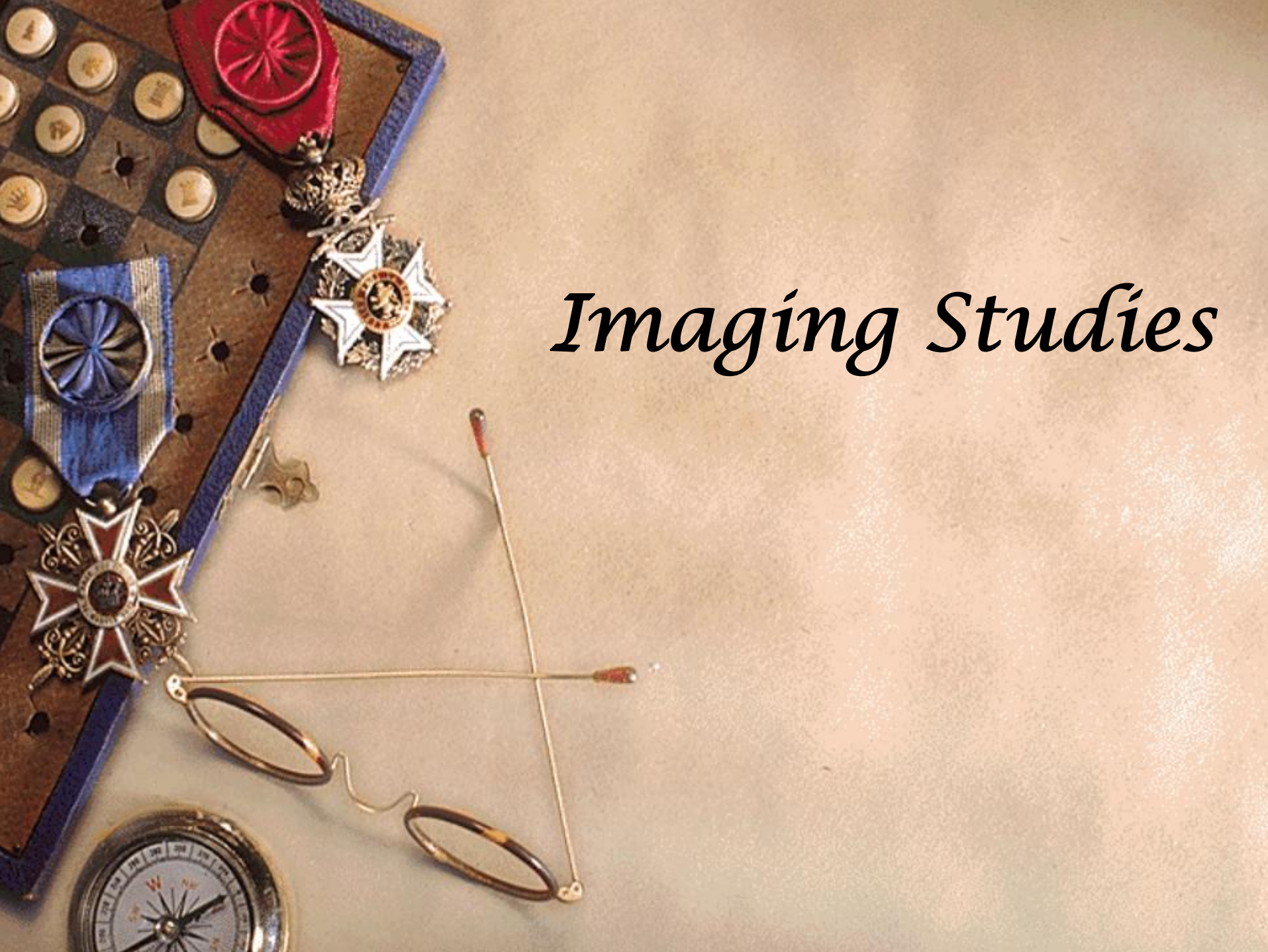
Not until February in this year, when he couldn't urinate eventually, did the fully retained bladder bring him to Ching-May hospital.

General History(4)

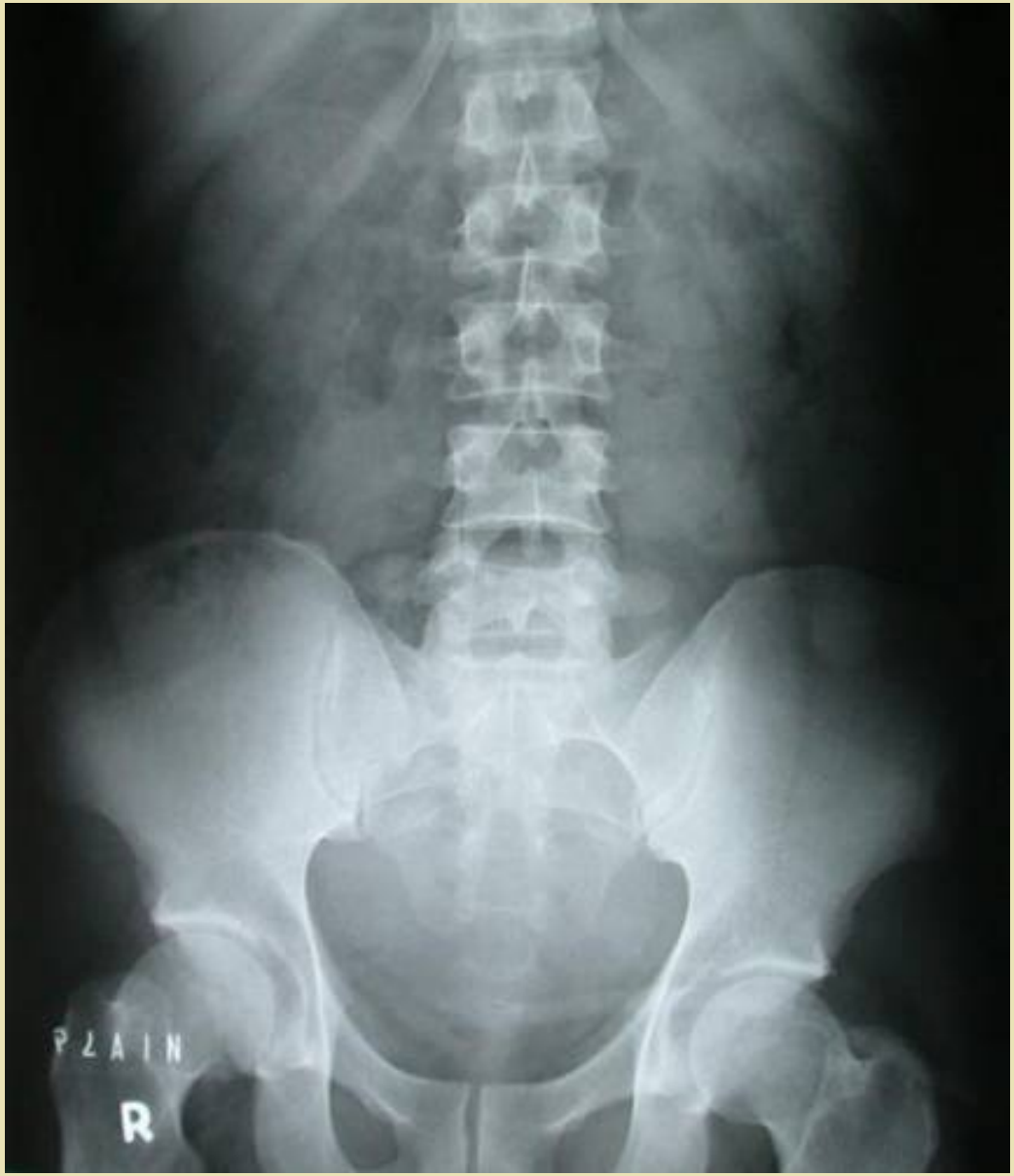
At Ching-May hospital, ICP was performed but was failed due to severe stricture of the urethra. Hence, cystoscpoe was performed alternatively, which revealed that an acute angle made the urethra to be obstructed.

However, because the cystoscpoe was still failed to pass into the bladder, Mr. 邱 was referred to our hospital for further evaluation and managements.

X


Imaging Studies

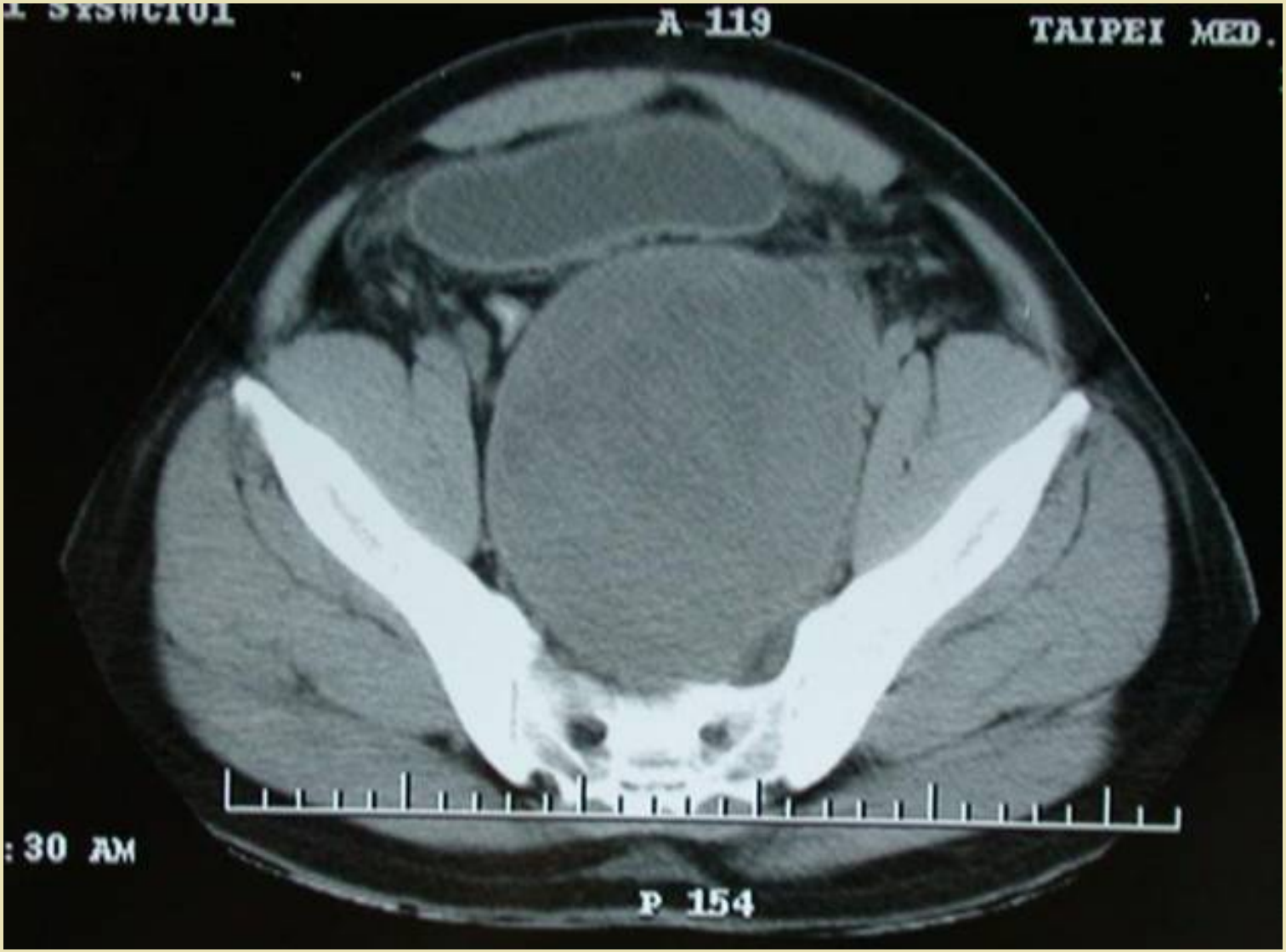
KUB



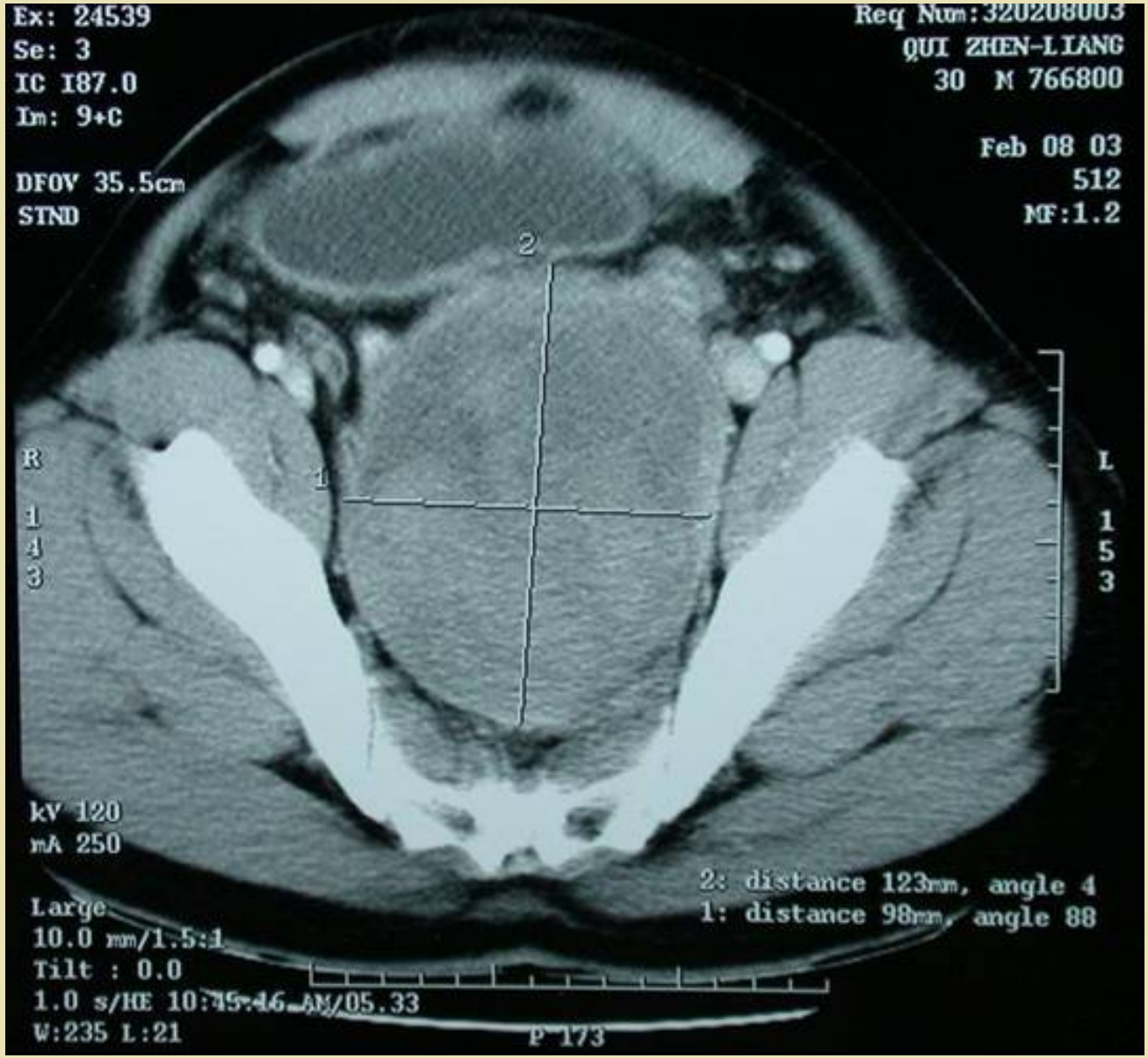
KUB

- Right hydronephrosis and hydroureter are noted.
- Relatively increased radiopaque density at left pelvic cavity region.
- Right-side displacement of urinary bladder is noted.

Pre Contrast Pelvic CT



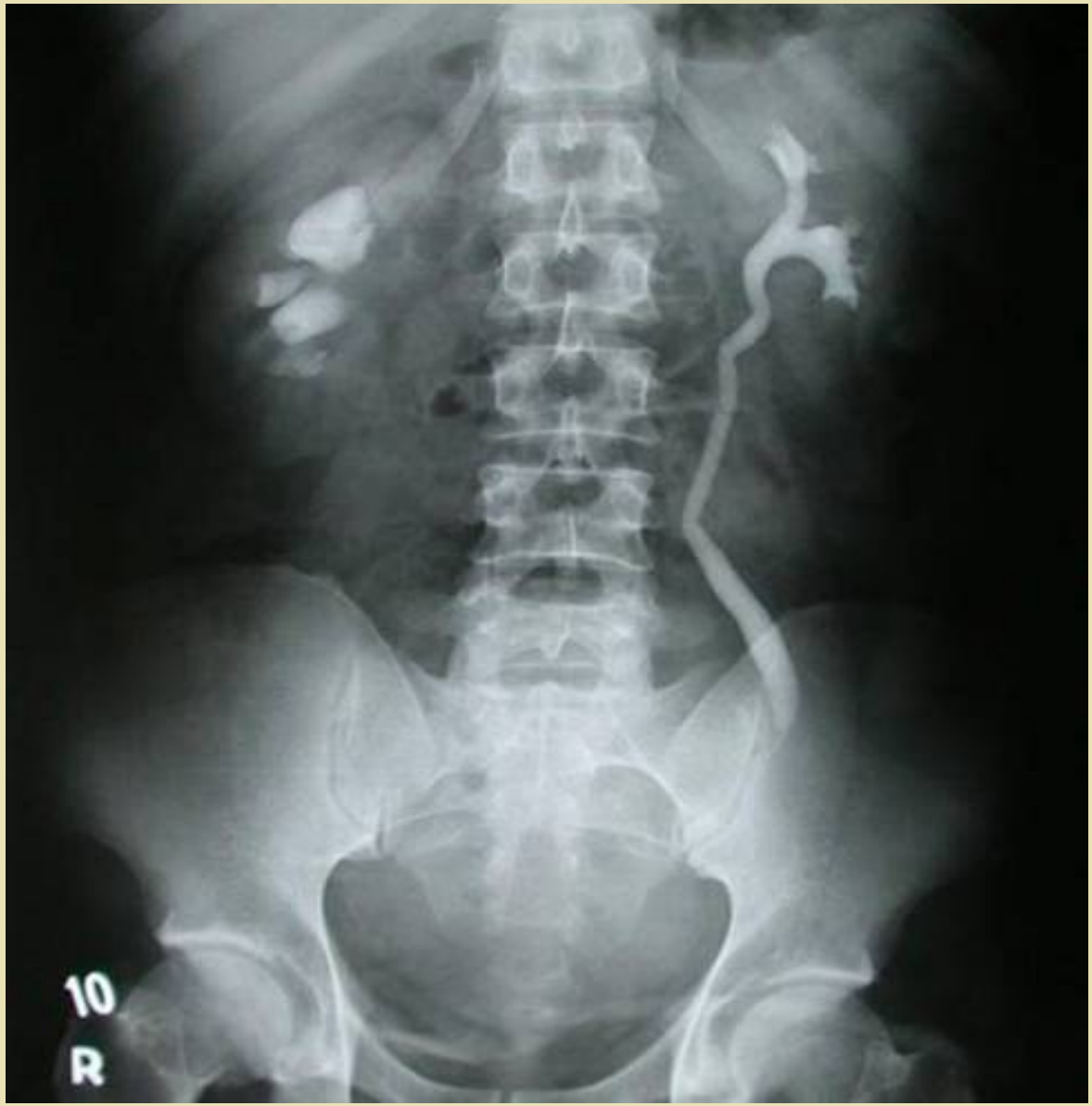
Post Contrast Pelvic CT



Pelvic CT(1)

- A large, encapsulated mass (9.8 cm x 12.3 cm) situated at the pelvic cavity just at presacral location.
- The mass has an soft tissue density on pre-contrast image and exhibits heterogenous enhancement.

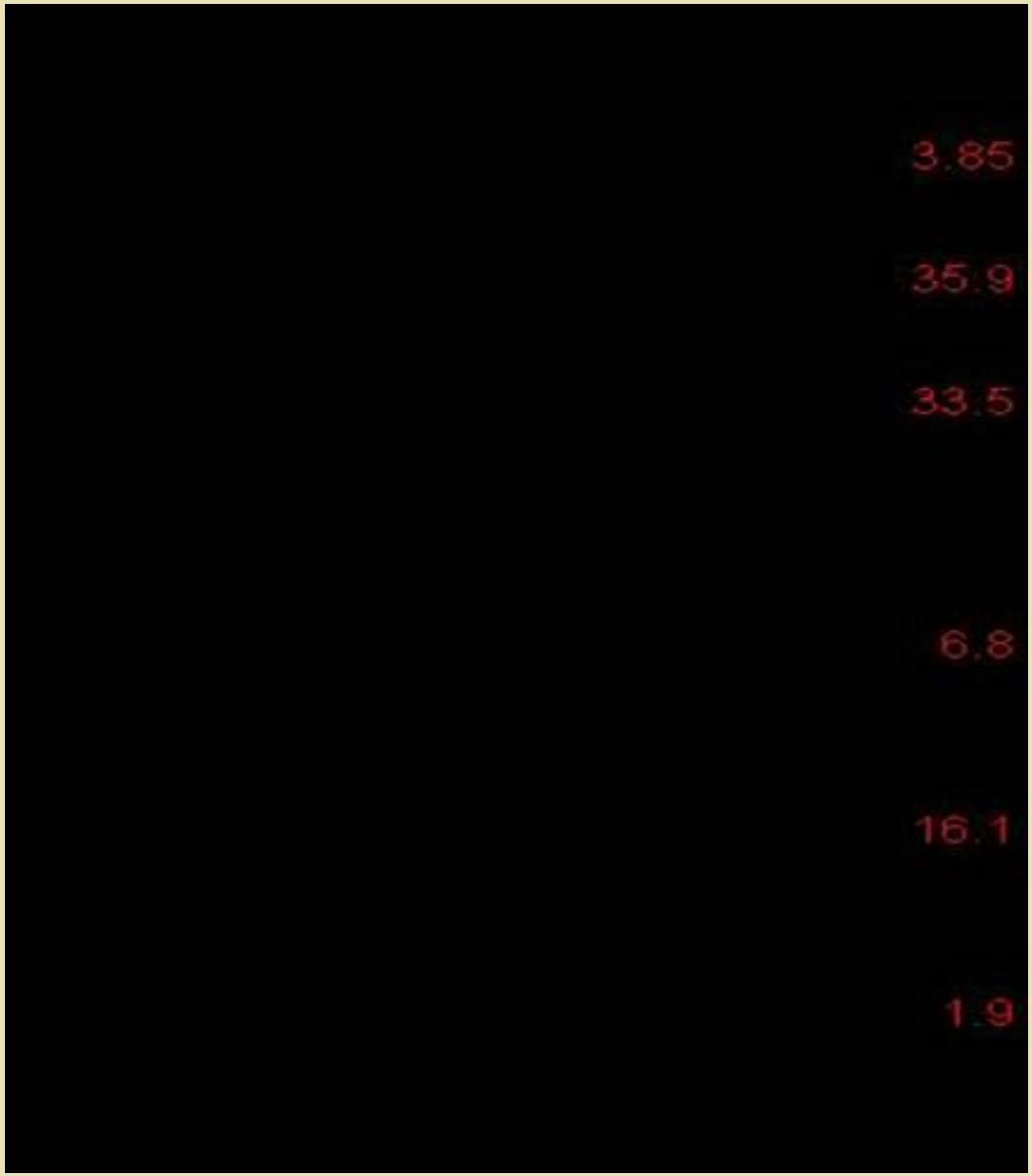
R’t Hydronephrosis



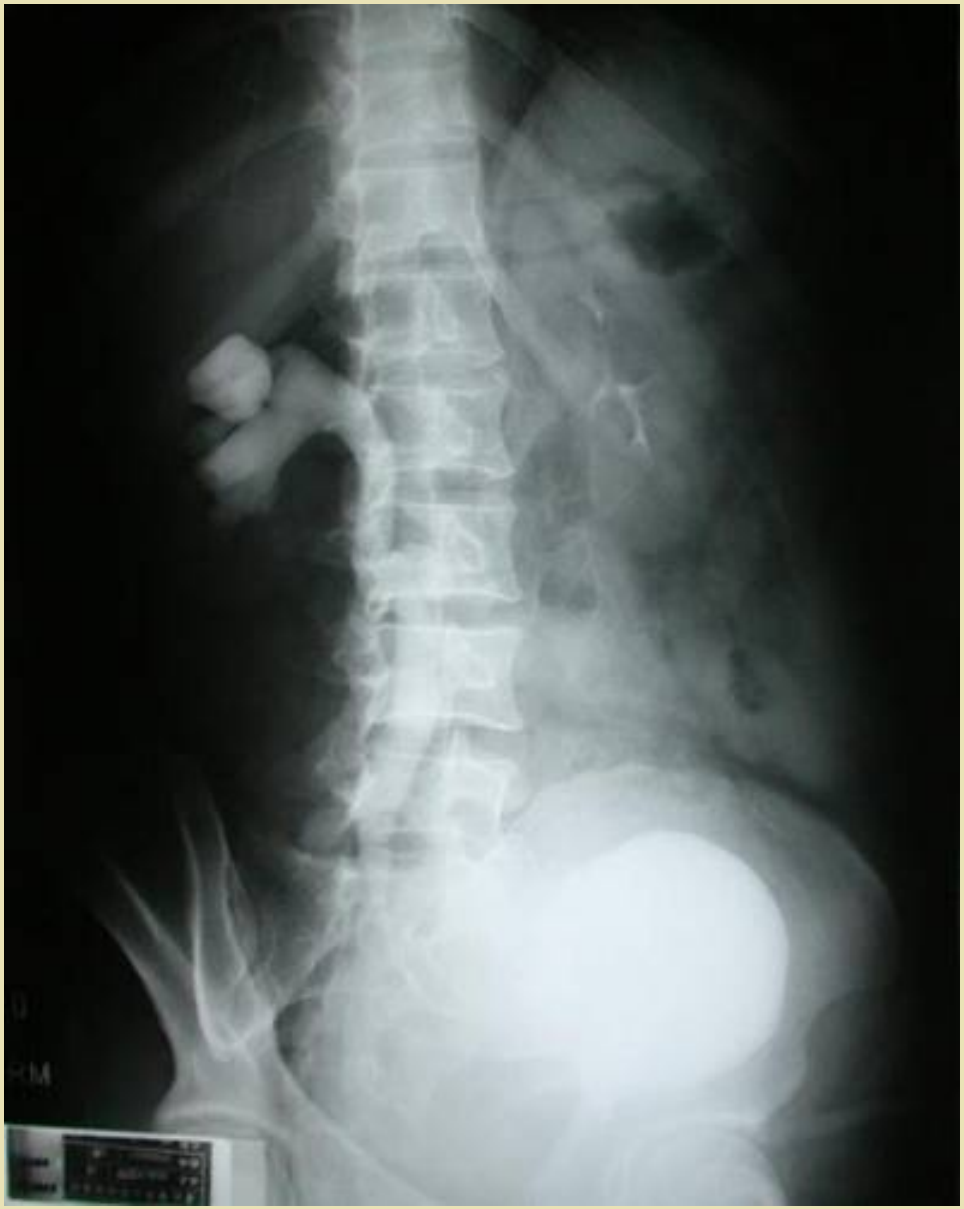
Pelvic CT(2)

- The mass results in anterior to right side displacement of the urinary bladder, complicated with hydroureter and hydronephrosis on the right side.
- The mass also results in lateral displacement of bilateral lower ureters.

IVP 10thmin



IVP 30thmin



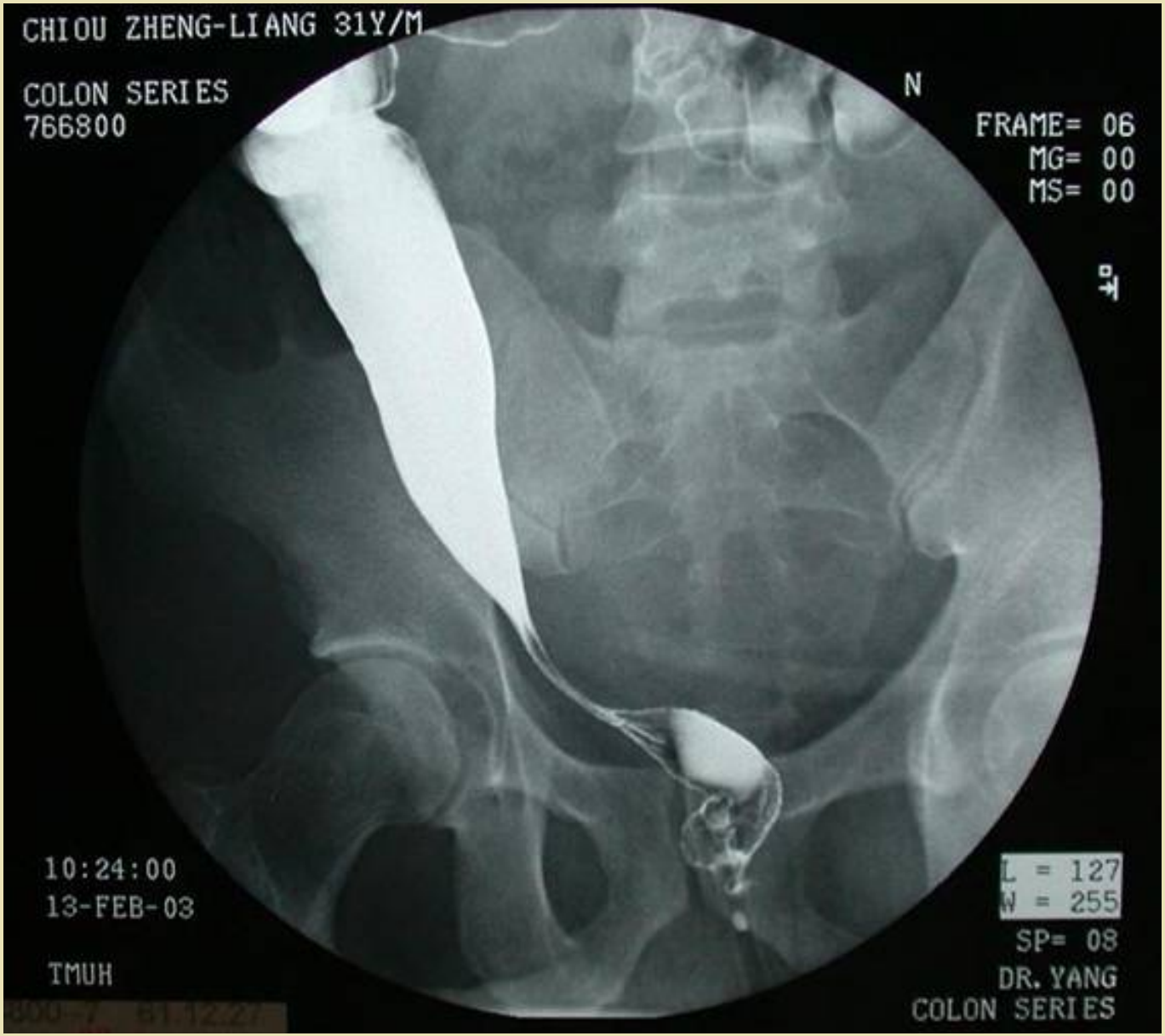
IVP

- Right hydronephrosis and right hydroureter, and with dilated left ureter.
- Both ureters are lateral displaced and the urinary bladder is anteriorly displaced.

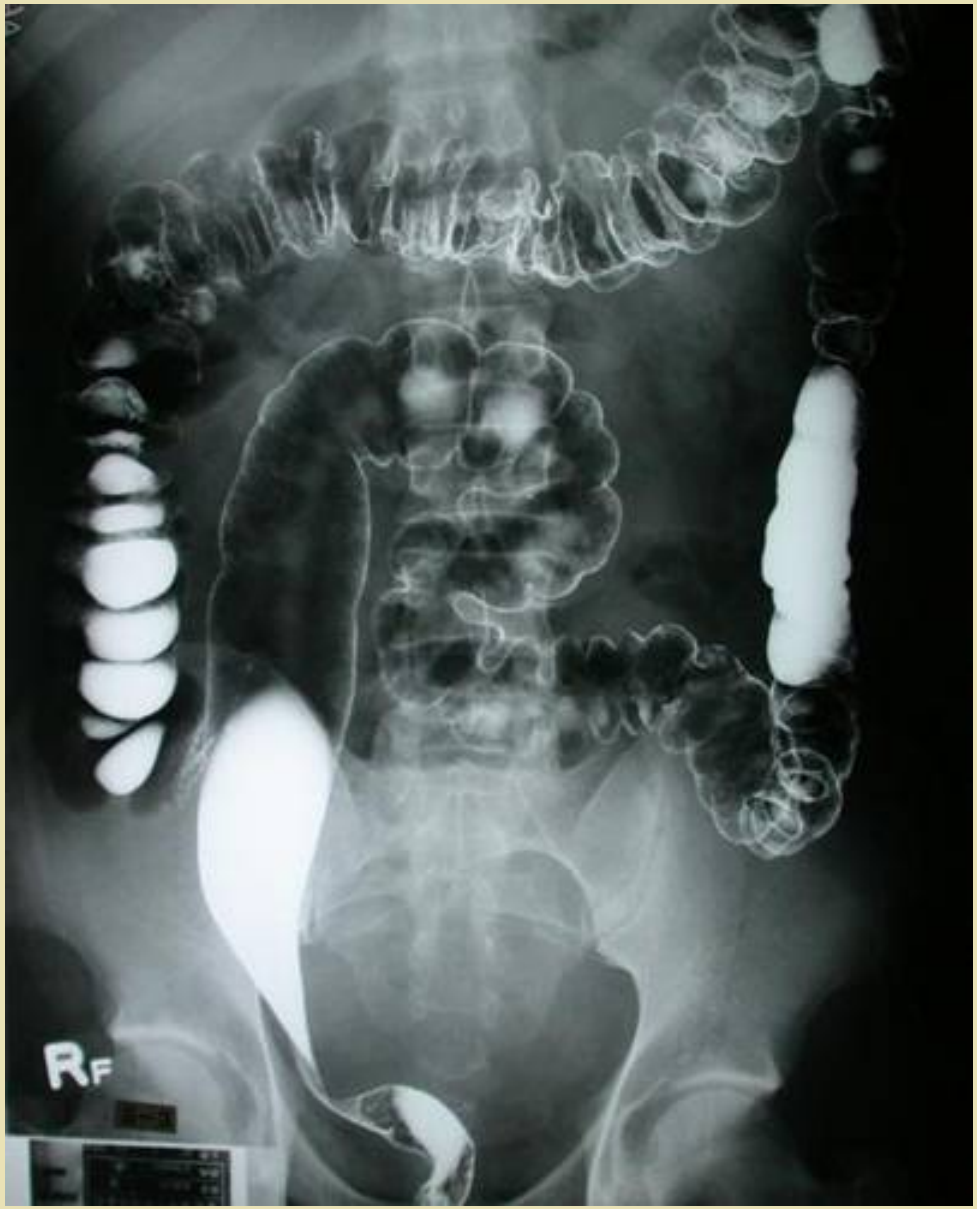
Bone Scan

- Whole body bone scan revealed no evident of primary bone lesion or secondary bone metastasis.

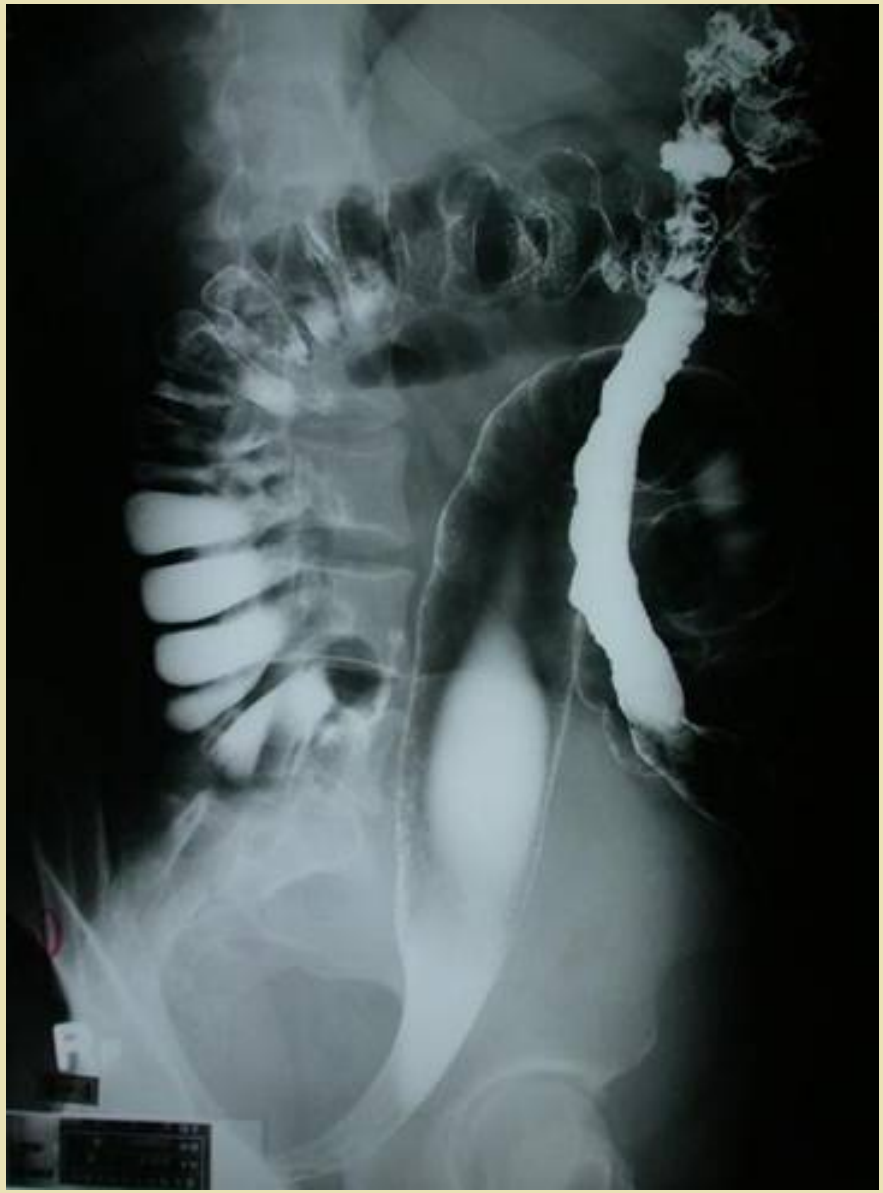
Barium Enema Pelvic View



Barium Enema AP View



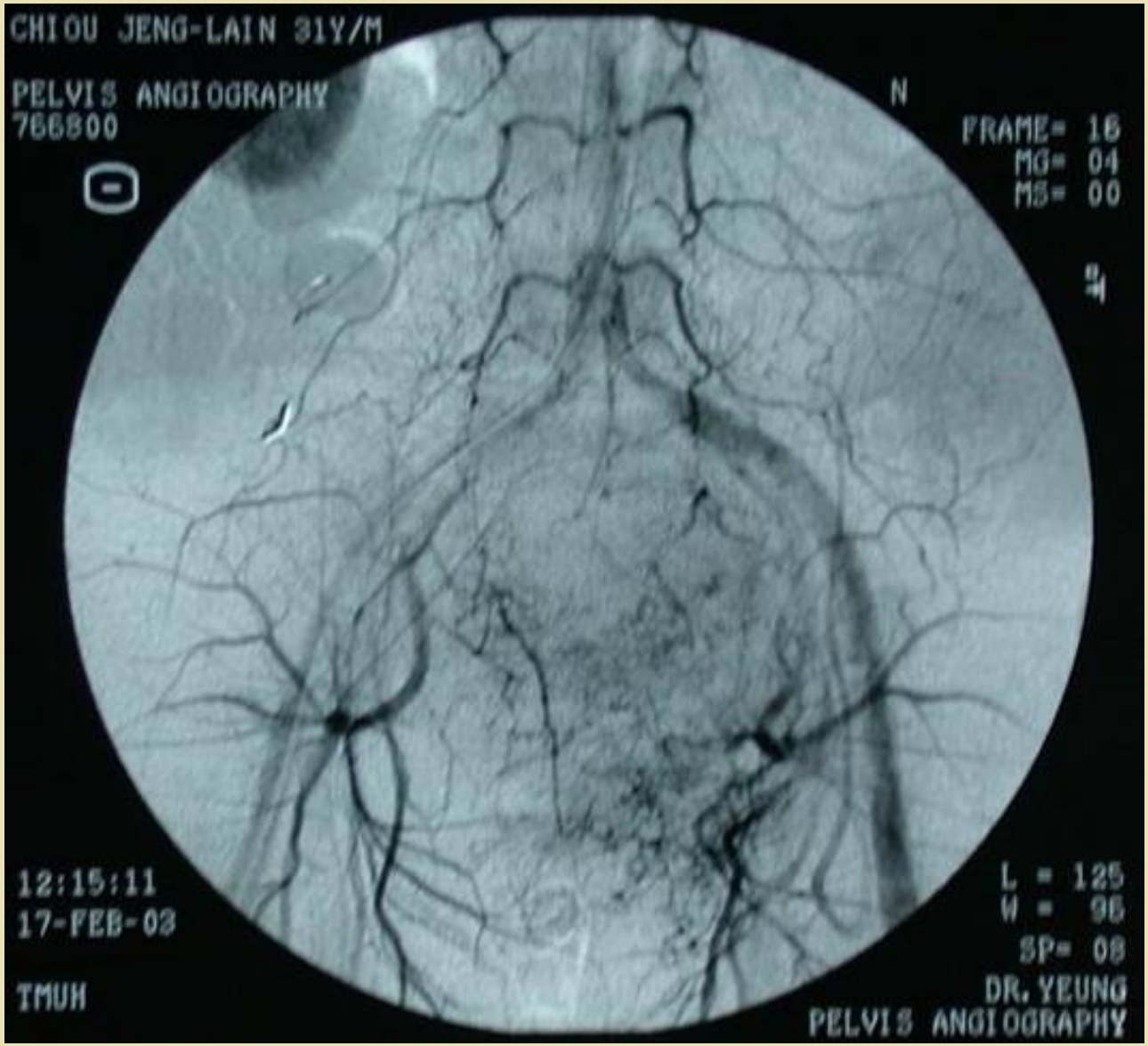
Barium Enema Lateral View



Barium Enema

- Marked widening of the presacral space, with anterior displacement of the rectum.
- The sigmoid colon is displaced upwardly and right laterally.
- Smooth passage of barium without colon obstruction.

Pelvic Angiography



Pelvic Angiography

- There is a hypervascular tumor stains lesion in the left side pelvic cavity, supplied by the branches of the bilateral internal iliac arteries, mainly from the left side.


Differential
Diagnosis

Differential Diagnosis

- The followings are the possible causes:
  - Leiomyosarcoma
  - Liposarcoma
  - Fibrosarcoma
  - Malignant fibrous histiocytoma
  - Neurogenic tumor

Radiologic Characteristics of

Leiomyosarcoma

Leiomyosarcoma X-ray(1)

- Plain radiographs usually offer little in the evaluation of GISTs.
- In the chest, esophageal GISTs can appear as a soft tissue mass in the lower two thirds of the mediastinum.
- In the abdomen, the soft-tissue mass may cause deformation of the gastric air shadow or displace loops of bowel.
- Abdominal films may depict an obstructive bowel pattern. If necrotic, collections of air can be visualized within the tumor (Shojaku, 1997).

Leiomyosarcoma X-ray(2)

- Regardless of the location of GISTs, barium-enhanced images demonstrate predominantly intramural masses with potential exophytic components.
- The tumor margins usually are smooth, but with ulceration, some surface irregularity is present.
- As with other intramural masses, the tumor borders form right or obtuse angles with the adjacent visceral wall. The intraluminal surfaces often have well-defined margins.

Leiomyosarcoma X-ray(3)

- Because the tumors are intramural but extramucosal, the overlying mucosa can be intact.
- In the stomach, this location results in a preserved areae gastricae pattern over the tumor mass. However, overlying mucosal ulcerations are often present; they are more common in malignant GISTs. These ulcerations fill with barium, causing a bull's eye or target-lesion appearance.

Leiomyosarcoma X-ray(4)

- If necrosis and cavitation are present, barium may fill the inner parts of the tumor mass.
- At times, the mass is entirely exophytic, and thus, it is not appreciated during contrast-enhanced examination.
- Barium images outline the intraluminal portion of this tumor; frequently, a substantial exophytic extension is present.
- Double-contrast images show abnormalities in 80% of cases (Pidhorecky, 2000).

Leiomyosarcoma CT(1)

- CT should be performed with both oral and intravenous contrast materials.
- Smaller GISTs appear as smooth, sharply defined intramural masses with homogenous attenuation.
- Contrast enhancement may be rim-like or uniform. Occasionally, dense focal calcifications are present.
- Larger GISTs with necrosis appear as heterogeneous masses with enhancing borders of variable thickness and irregular central areas of fluid, air, or oral contrast attenuation that reflect necrosis.

Leiomyosarcoma CT(2)

- Overlying mucosal ulcerations and extension into nearby structures may be present.
- CT is also sensitive for the detection of metastatic liver, peritoneal, lung, and bone lesions.
- The diagnosis of GIST can be suggested in the presence of a large, complex, intestinal mass with liver lesions but without significant lymphadenopathy.
- Liver lesions can be hypervascular or appear as cystic multilocular lesions with fluid-fluid levels (Levine, 1996; Miettinen, 2001; Shojaku, 1997; Buckley, 1998; Lehnert, 1998; Miettinen, 2000).

Leiomyosarcoma CT(3)

- CT scanning has good sensitivity for the detection of GISTs, and it can show abnormalities in 87% of cases (Pidhorecky, 2000).

Leiomyosarcoma MRI

- GISTs appear as sharply delineated, heterogeneous masses with cystic and necrotic areas.
- The masses tend to be isointense relative to skeletal muscle on T1-weighted images and hyperintense on T2-weighted images.
- Signal intensity voids are present if gas is present within areas of necrotic tumor (Levine, 1996; Shojaku, 1997; Tervahartiala, 1998).

Leiomyosarcoma Ultrasound

- On sonograms, larger GISTs appear as complex masses with cystic and solid components, which are consistent with their tendency to necrose (Shojaku, 1997; Tervahartiala, 1998).
- Endoscopic ultrasonography can be valuable in the evaluation of GISTs. The tumors appear as hypoechoic masses that are contiguous with the fourth hypoechoic layer of the GI wall, which corresponds to the muscularis propria.

Leiomyosarcoma Angiography

- Angiography demonstrates a relatively well-circumscribed hypervascular lesion with central avascularity.
- They have large feeding arteries and draining veins, and they show intense tumor staining (Miettinen, 2000).

Radiologic Characteristics of

Liposarcoma

Liposarcoma X-ray

- Radiographic findings are seldom diagnostic, and the images may demonstrate a nonspecific soft-tissue mass. Frequently, no fat is detectable. Rarely, calcification is present.
- Abdominal radiographs in patients with retroperitoneal tumors may reveal a soft tissue–displacing gas-filled structure and effacement of the normal fat planes.
- Sensitivity and specificity of radiographs is low in liposarcoma.

Liposarcoma CT(1)

- CT scans demonstrate 3 distinct patterns:
  - An enhancing, solid, inhomogeneous, poorly defined, infiltrating mass.
  - A mixed-pattern tumor with foci of fat interspersed in high-attenuating tissue.
  - A pseudocystic water-density tumor.

Liposarcoma CT(2)

- Liposarcomas of the myxoid type, the mixed myxoid and round cell type, the round cell type, and the pleomorphic type are usually poorly defined, with attenuation values of 12-38 HU and varying degrees of contrast enhancement. Calcification is detectable in as many as 12% of the tumors.
- Malignant fibrous histiocytoma, leiomyosarcoma, and desmoid tumors may have appearances indistinguishable from liposarcoma, particularly of the myxoid, mixed myxoid and round cell, round cell, and pleomorphic types.

Liposarcoma MRI(1)

- Most liposarcomas appear well defined on MRIs, mostly with lobulated margins.
- Well-differentiated liposarcomas are mainly composed of fat with septations or nodules and are hyperintense on T2-weighted images.
- After the administration of contrast material, well-differentiated liposarcomas may enhance minimally or not at all.

Liposarcoma MRI(2)

- Most myxoid tumors have linear or lacy amorphous foci of fat. Some myxoid tumors may appear cystic on nonenhanced MRIs, but they are usually enhancing after the administration of contrast agents.
- Pleomorphic tumors show a markedly heterogeneous internal structure and moderate contrast enhancement.
- The malignancy grade is believed to increase in parallel with tumor heterogeneity and contrast enhancement.

Liposarcoma MRI(3)

- Well-differentiated liposarcomas may be differentiated from other types of tumors on the basis of their largely lipomatous appearances.
- Gadolinium-enhanced imaging is important in differentiating myxoid liposarcomas from benign cystic tumors.

Liposarcoma Ultrasound(1)

- Liposarcomas are usually hyperechoic.
- Retroperitoneal liposarcomas are highly reflective, although this feature may be absent when the tumor is poorly differentiated.
- The finding of a solid retroperitoneal mass that demonstrates a heterogeneous echo pattern with an echo-poor center usually suggests a sarcoma.
- The central echo-poor area is usually the result of hemorrhage or necrosis because the tumors tend to outgrow their blood supply.

Liposarcoma Ultrasound(2)

- A well-differentiated peripheral liposarcoma is usually hyperechoic and may be indistinguishable from a lipoma. However, Doppler studies reveal that a liposarcoma is more vascular than a lipoma.
- The remaining 3 varieties of liposarcoma appear as a heterogeneous soft-tissue mass with no distinguishing characteristics.

Liposarcoma Ultrasound(3)

- Vascular tumors (eg. Hemangiopericytomas) can be highly reflective, presumably because of the numerous tissue interfaces with multiple vascular walls.
- Distinguishing poorly differentiated lipo-sarcomas from other types of retroperitoneal or peripheral masses is not always possible. A peripheral well-differentiated liposarcoma may have the appearance of a lipoma.

Liposarcoma Angiography

- Liposarcomas are usually hypovascular to moderately vascular, and they cause displacement of the major vessels, particularly the inferior vena cava. Venous filling may occur early, and the veins may be dilated and tortuous.
- Displacement of the kidneys and arteries is seen in all except very small retroperitoneal tumors.
- Angiography cannot help in differentiating lipo-sarcomas from other types of sarcomas.
- Moreover, both benign and malignant retro-peritoneal tumors can be avascular.

Radiologic Characteristics of

Fibrosarcoma

Fibrosarcoma X-ray(1)

- Plain radiographs of the involved anatomic region are needed to evaluate for primary or secondary involvement of bone.
- Typically an osteolytic area of destruction with a permeative or moth-eaten appearance is present. Little periosteal reaction or reactive sclerosis is depicted.

Fibrosarcoma X-ray(2)

- For bony lesions, plain radiographs often greatly assist with determining diagnosis, location, size, and local extent of involvement.
- For soft tissue masses, size often can be estimated, any bone involvement can be seen, and intra-lesional content (matrix) can sometimes be determined.

Fibrosarcoma CT

- Density of fibrosarcoma is similar to that of surrounding normal muscle.
- Signs of fracture or impending fracture may be seen, and the tumor can be more accurately localized.
- CT scanning of the chest may be appropriate. CT scan is very sensitive for metastatic disease.

Fibrosarcoma MRI

- MRI may be the best overall study for soft tissue masses and for detecting the intraosseous and extraosseous extent of many bony sarcomas.
- MRI is useful in providing information about the local extent, lesion size, and involvement of the neurovascular structures. Fibrosarcoma of bone typically has extraosseous extension.

Fibrosarcoma Bone Scan

- Bone scanning using technetium Tc^{99m} is a very useful adjunct in the evaluation of tumor stage.
- It helps to detect bone metastatic disease or polyostotic disease.
- For fibrosarcoma, it has been mostly supplanted by MRI. The limitation with bone scanning is that it often is nonspecific.

Radiologic Characteristics of

Malignant Fibrous Histiocytoma

Malignant Fibrous Histiocytoma
X-ray(1)

- Radiographs may reveal a nonspecific soft tissue mass, often greater than 5 cm in diameter.
- Deep intramuscular tumors often lie adjacent to the diaphysis of a long bone.
- Secondary osseous involvement, including periosteal reaction, cortical erosion, and pathologic fracture, is uncommon but suggestive of MFH.

Malignant Fibrous Histiocytoma
X-ray(2)

- Calcification or ossification can be detected in 5-20% of patients.
- Calcifications within the tumor may be punctate, curvilinear, and/or poorly defined.
- Heterotopic bone formation may be present in the periphery of the mass.

Malignant Fibrous Histiocytoma
CT(1)

- CT typically reveals a nonspecific, large, lobulated, soft tissue mass of predominantly muscle density, with nodular and peripheral enhancement of solid portions.
- Central areas of low attenuation may be present, corresponding to myxoid regions, old hemorrhage, or necrosis.
- Fat attenuation is not observed in the tumors, which can distinguish tumors from some well-differentiated liposarcomas.

Malignant Fibrous Histiocytoma
CT(2)

- CT may be used to evaluate potential internal matrix and/or cortical erosion.
- Retroperitoneal tumors manifest as heterogeneous masses with areas of hemorrhage and/or necrosis and occasionally focal or diffuse coarse calcifications (approximately 10%). The tumors may invade the abdominal musculature but do not invade the renal veins or inferior vena cava.

Malignant Fibrous Histiocytoma
MRI(1)

- MRI typically reveals an intramuscular mass with heterogeneous signal intensity on all pulse sequences.
- As with other soft tissue neoplasms, the signal intensity pattern is nonspecific, usually low to intermediate on T1-weighted images and intermediate to high on T2-weighted images.
- Regions of prominent fibrous tissue (high collagen content) may demonstrate low signal on both T1-weighted and T2-weighted images.

Malignant Fibrous Histiocytoma
MRI(2)

- Calcification may present as foci of low signal on both T1-weighted and T2-weighted sequences.
- Consider subacute hemorrhage when regions of high signal are noted on both T1-weighted and T2-weighted images.
- Areas of necrosis demonstrate a signal pattern similar to fluid.
- As with CT, solid components of MFH typically reveal nodular and peripheral enhancement.

Malignant Fibrous Histiocytoma
MRI(3)

- Tumor margins appear relatively well defined on MRI. A low signal intensity margin may be observed, representing a pseudocapsule.
- False Positives/Negatives: The radiologist should keep in mind that the diagnosis of MFH is made using histopathology, not imaging; however, MRI remains invaluable for delineating tumor extent.

Malignant Fibrous Histiocytoma
Ultrasound

- Sonography typically reveals a well-defined heterogeneous mass that contains hyperechoic areas of cellularity and hypoechoic regions of necrosis.
- Appearance of tumors on ultrasound is nonspecific; however, sonography may be used to evaluate tumor volume.
- Retroperitoneal tumors tend to appear as hypoechoic solid masses with scattered regions of heterogeneity.

Malignant Fibrous Histiocytoma
Angiography

- Angiographic findings are nonspecific.
- The tumor may be hypovascular or, more commonly, hypervascular with early venous return.
- Similarly, retroperitoneal tumors may be either hypovascular or hypervascular, with blood supply from the lumbar, celiac, iliac, renal, renal capsular, and/or inferior adrenal arteries.

Radiologic Characteristics of

Neurogenic Tumor

Neurogenic Tumor

- Benign nerve sheath tumours are subdivided into Schwannoma and neurofibroma. Malignant nerve sheath tumours are known by a confusing number of different names, usually including the terms either malignant or sarcoma.
- On MRI, Schwannomas typically present as a small fusiform mass with tapering margins.
- We will discuss in the last section.

Operation Findings and Pathologic Findings

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5.8

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OP Finding

- Pre-op and post-op diagnosis
  - Pelvic tumor, R/O malignancy.
  - Right side hydronephrosis.
- OP method
  - Pelvic tumor excision.
  - Right side D-J insertion.
- OP Finding
  - A large tumor between urinary bladder and rectum displacing bladder anteriorly and sigmoid colon laterally with severe adhesion to sacrum.

Pathology(1)

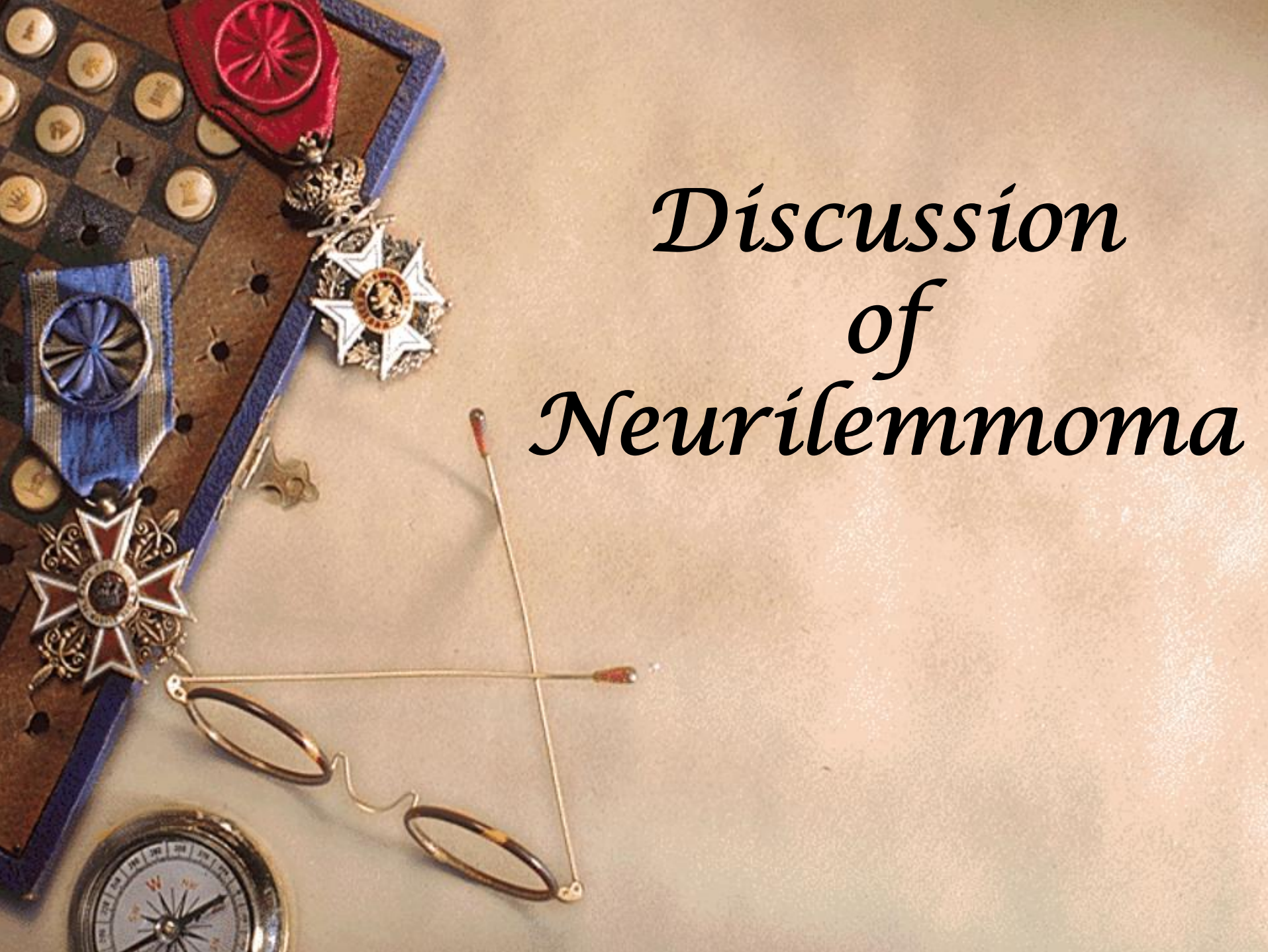
- Soft tissue, retroperitoneum, excisional biopsy, 14 x 10.5 x 10 cm. in size, fixed in formalin.
- Grossly, it is a well-defined, elastic and flesh-colored to tan tumor.
- On cut, hemorrhage and cystic change are seen focally. No necrosis is found.

Pathology(2)

- Microscopically, it shows a picture of ancient neurilemmoma, composed of spindle cells in a fibrillary or myxoid background.
- No Lymphoplasma cell aggregation are seen.
- Marked degeneration with hyalinized thickening of vascular wall, cystic degeneration and nuclear atypia are seen.
- No mitoses is found. No evidence of malignancy is seen.

Pathology(3)

- Immunohistochemical studies reveal positive staining S-100, and negative of actin, desmin, or CD 117.
- Ancient neurilemmoma.



Discussion of Neurilemmoma

Neurilemmoma (Schwannoma)

- Neurilemmomas are benign encapsulated tumors of the nerve sheath.
- Their cell of origin is thought to be Schwann cells derived from the neural crest.
- These masses usually arise from the side of a nerve, are well encapsulated, and have a very unique histologic pattern.

Causes

- The cause for the growth of these neoplasms is unknown.
- Neurilemmoma can be associated with von Recklinghausen disease, and, when associated, multiple tumors often are present.

Epidemiology

- No racial or sex predilection exists.
- Neurilemmomas affect persons aged 20-50 years.
- The head and flexor surface of the upper and lower extremities and the trunk are common locations in decreasing order.

Location(1)

- Within the cranial vault, the most common location is in the cerebellopontine angle, where they are attached to the vestibular branch of the eighth nerve.
- Patients often present with tinnitus and hearing loss, and the tumor is often referred to as an acoustic neuroma.

Location(2)

- Elsewhere within the dura, sensory nerves are preferentially involved, including branches of the trigeminal nerve and dorsal roots.
- When extradural, schwannomas are most commonly found in association with large nerve trunks, where motor and sensory modalities are intermixed.

Symptoms(1)

- The benign lesion presents essentially with cosmetic deformity, a palpable mass, and/or symptoms similar to a compressive neuropathy.
- Neurologic symptoms tend to present late. Symptoms can be vague, with an average interval of up to 5 years before the diagnosis is established.
- Neurilemmoma is the most common neurogenic tumor.

Symptoms(2)

- Tenderness to palpation is often present.
- When involving the C7 nerve root, neurilemmoma has been described as a cause of thoracic outlet syndrome. Lesions in the sciatic nerve can mimic discogenic low back pain.
- These benign tumors can cause a functional deficit because of local pressure on the nerve of origin.

Symptoms(3)

- Some may involve the spinal nerve roots and present with symptoms that mimic that of herniated disk disease of the spine.
- In the extremities, presentation can be either an symptomatic mass or mild localized pain and paresthesia due to pressure on the nerve of origin.
- Masses are slow growing and can be present for months to years without symptoms.
- The average time from onset of symptoms to diagnosis is 5.5 years.

Symptoms(4)

- Lesions in proximal nerves may cause distal symptoms. If these masses occur in well-defined compartments (eg, wrist, ankle), they can present as carpal tunnel syndrome or tarsal tunnel syndrome.
- In the unusual case in which resection would lead to a significant functional deficit, these benign lesions can be merely observed.

Imaging Study(1)

- Plain radiograph findings generally are not specific. The rare intraosseous lesion presents as a benign-appearing well-circumscribed lesion.
- Massive bony destruction may be present, especially when the lesion involves the sacrum.

Imaging Study(2)

- MRI is particularly useful and shows a usually round or oval mass with a moderately bright signal on T1-weighted images and a bright heterogeneous signal on T2-weighted images.
- The mass is usually less than 2.5 cm in size. The lesion enhances uniformly with gadolinium contrast.

Pathology(1)

- These benign tumors arise from the neural crest-derived Schwann cell and are associated with neurofibromatosis type 2.
- Tumors are firm, gray masses but may also have areas of cystic and xanthomatous change.
- On microscopic examination, tumors show a mixture of two growth patterns.

Pathology(2)

- Because the lesion displaces the nerve of origin as it grows, silver stains demonstrate that axons are largely excluded from the tumor, although they may become entrapped in the capsule.
- The Schwann cell origin of these tumors is borne out by their S-100 immunoreactivity.

Pathology(3)

- The diagnostic criteria for neurofibromatosis type 1 (von Recklinghausen's disease) include café-au-lait spots, one plexiform neurofibroma, or more than one neurofibroma.
- Neurofibromatosis type 2 (Central neurofibro-matosis) is a multisystem genetic disorder associated with bilateral vestibular schwannomas, spinal cord schwannomas, meningiomas, gliomas, and juvenile cataracts with a paucity of cutaneous features, which are seen more consistently in neurofibromatosis type 1 (NF1).

Malignant Potential

- Malignant degeneration is extremely rare but was described by Yousem et al in 1985.
- Primary malignant tumors of this cell type do exist, but histologically, they are very distinct from this benign counterpart.

Prognosis

- Recurrence is unlikely with complete resection. Patients usually have rapid and complete relief of pain, with excellent long-term results.
- Rare descriptions exist of malignant change in long-standing benign tumors of this type, usually in patients with an underlying diagnosis of neurofibromatosis. Malignant change is extremely rare in isolated lesions.