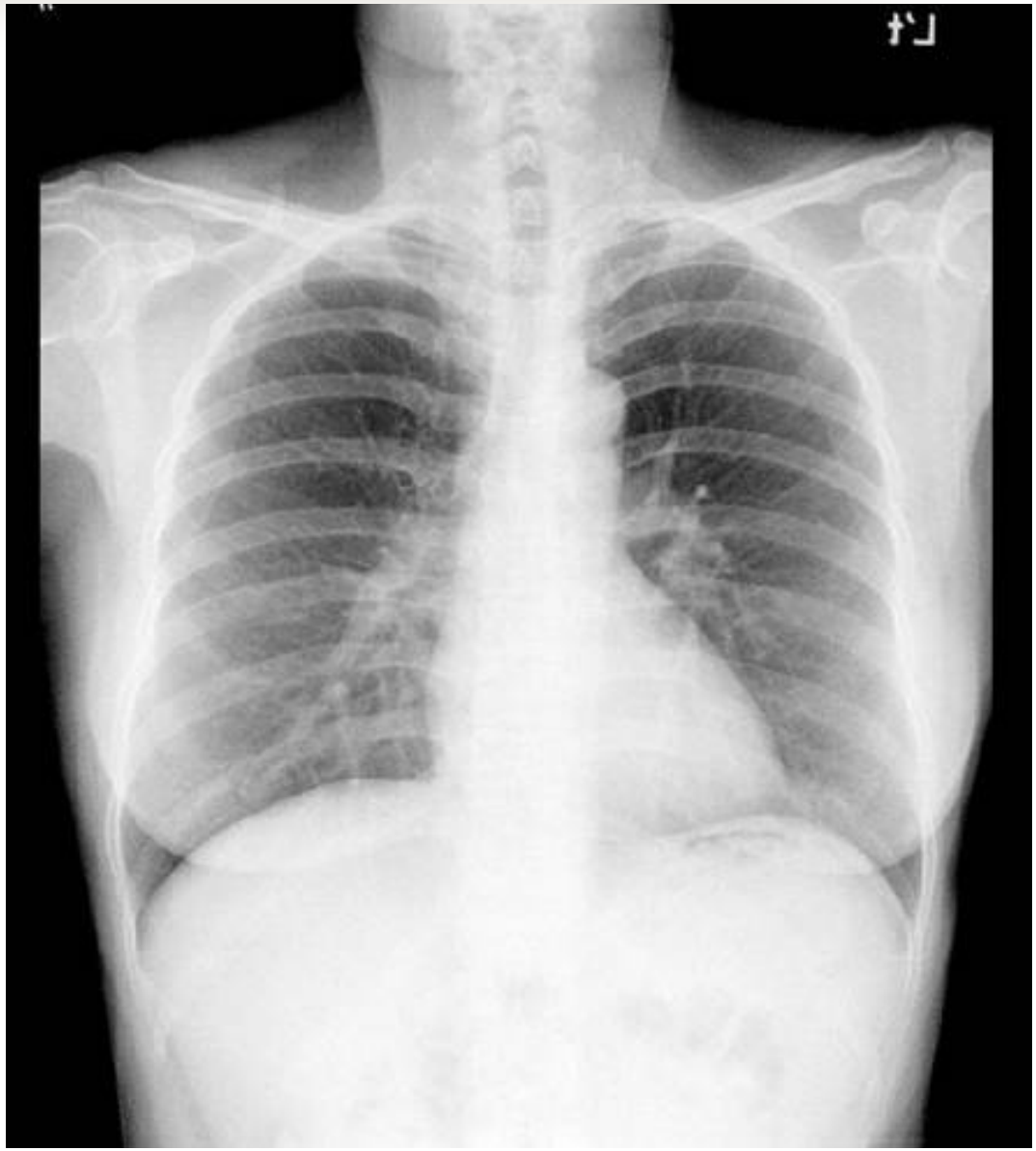
General History

- This 53-year-old female suffered from uterine myoma s/p open myomectomy 12 years ago at ChangGung hospital.
- Left peri-renal mass was noted during health examination at Ren-Ai hospital. There was no HTN, glucose intolerance, or electrolyte imbalance.
- She visited our urology department.

Lab Studies

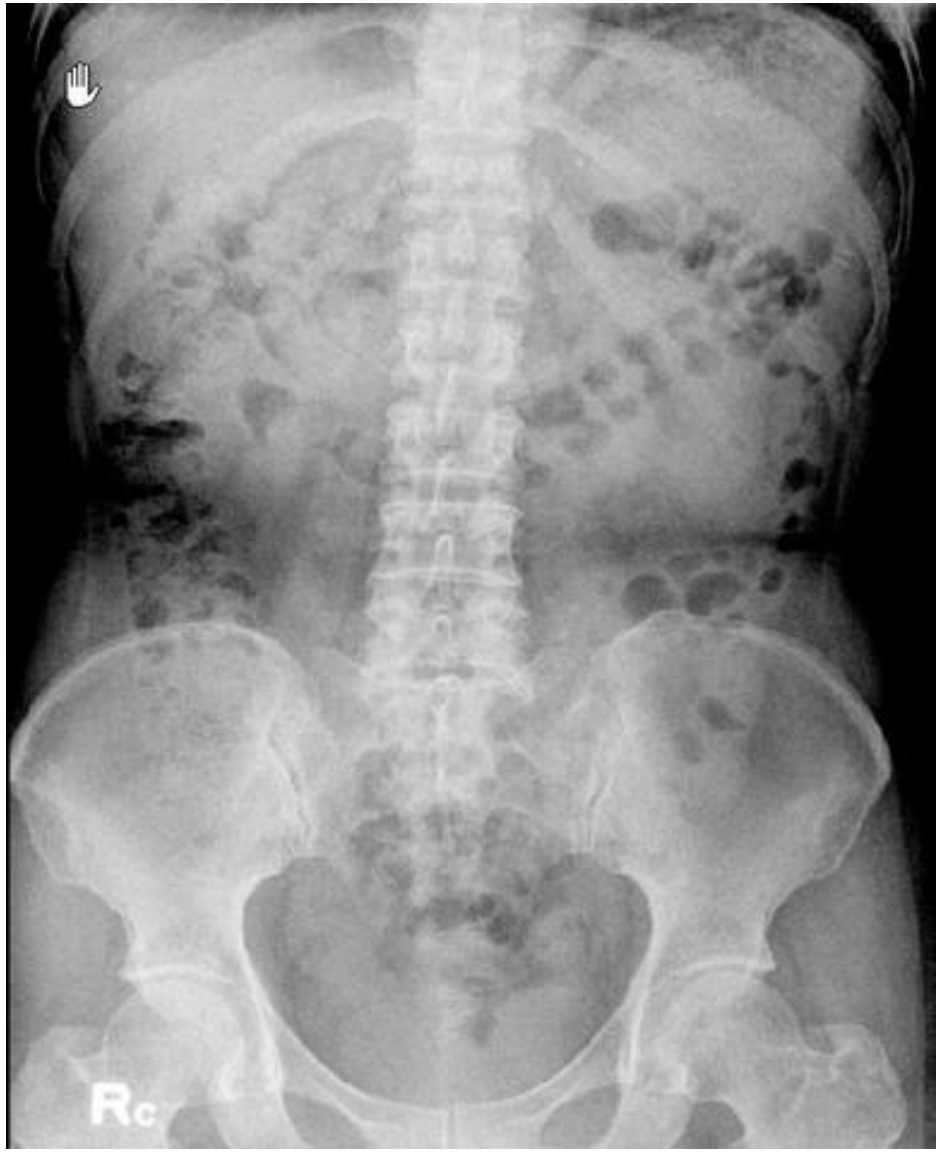
- Levels of ACTH, renin, aldosteron, and cortisol did not show significant abnormality.
- WBC [5.2-12.4 x10.e3/uL]: 3.54
- Cl (blood)[98-108 meq/L]: 109.0

Imaging Studies



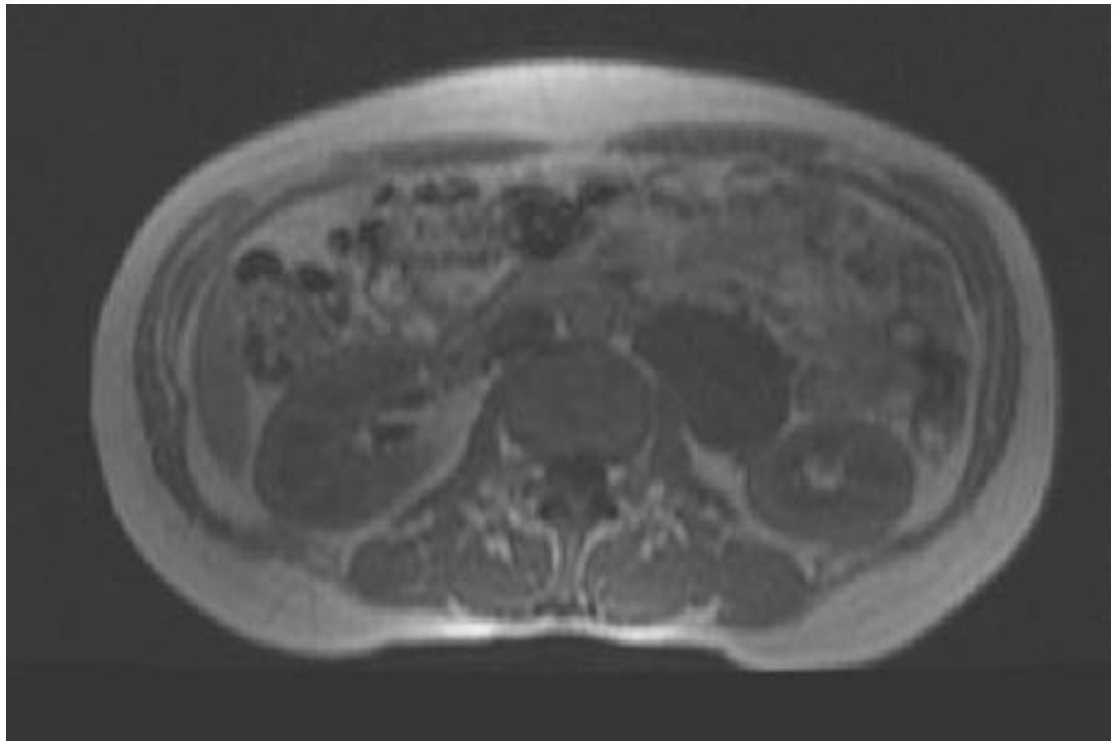
- **CXR**: No positive finding.

- **KUB**: No positive finding.



MRI T1W

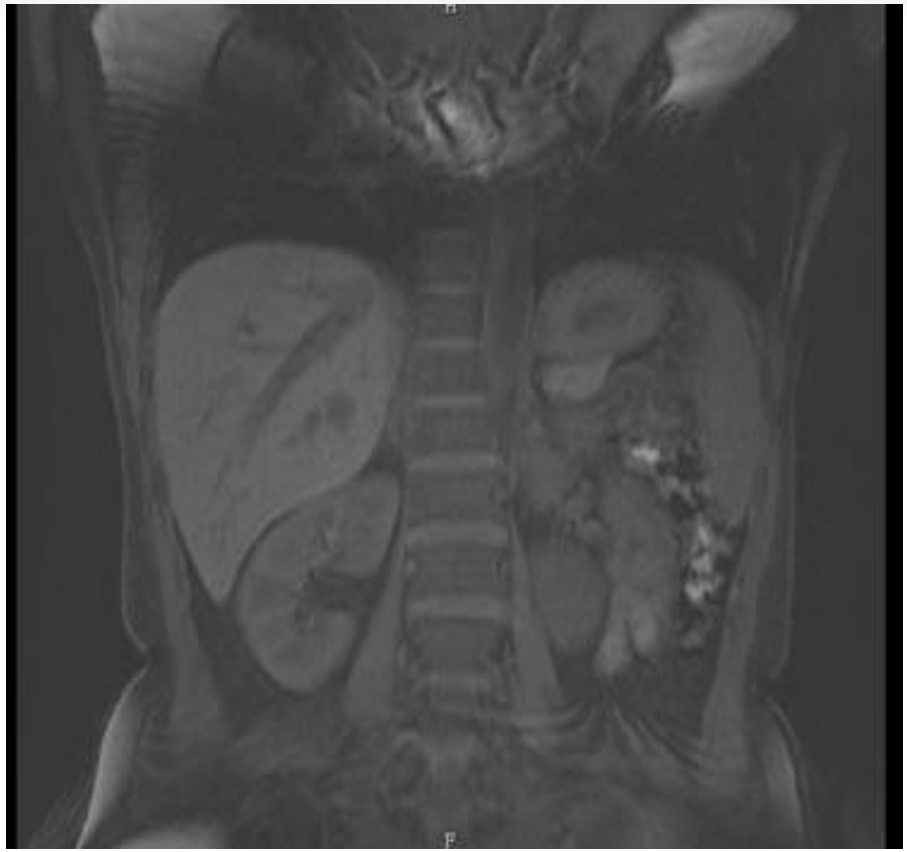
transverse view



- A large well-defined T1W low signal mass is noted at the left para-aortic region just below the left renal hilar level and anterior medial to the left kidney.The mass about 5x5x3 cm in size.

MRI T1W

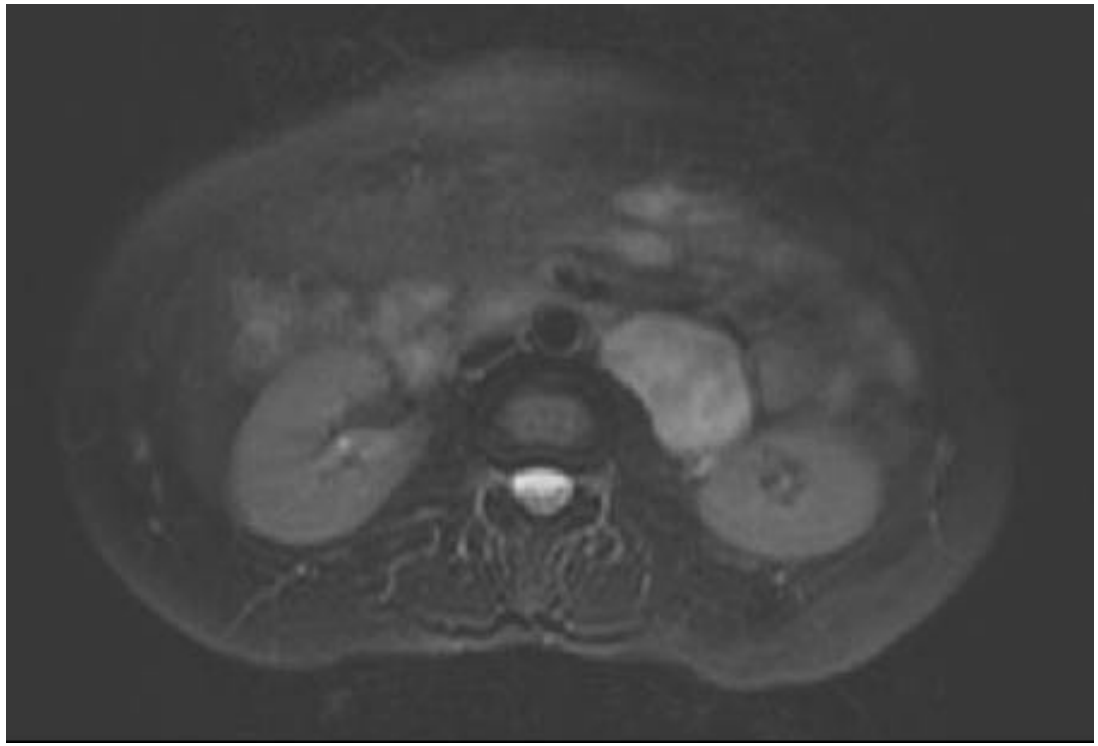
coronal view



- A large well-defined T1W low signal mass is noted at the left para-aortic region just below the left renal hilar level and anterior medial to the left kidney.The mass about 5x5x3 cm in size.

MRI T2W

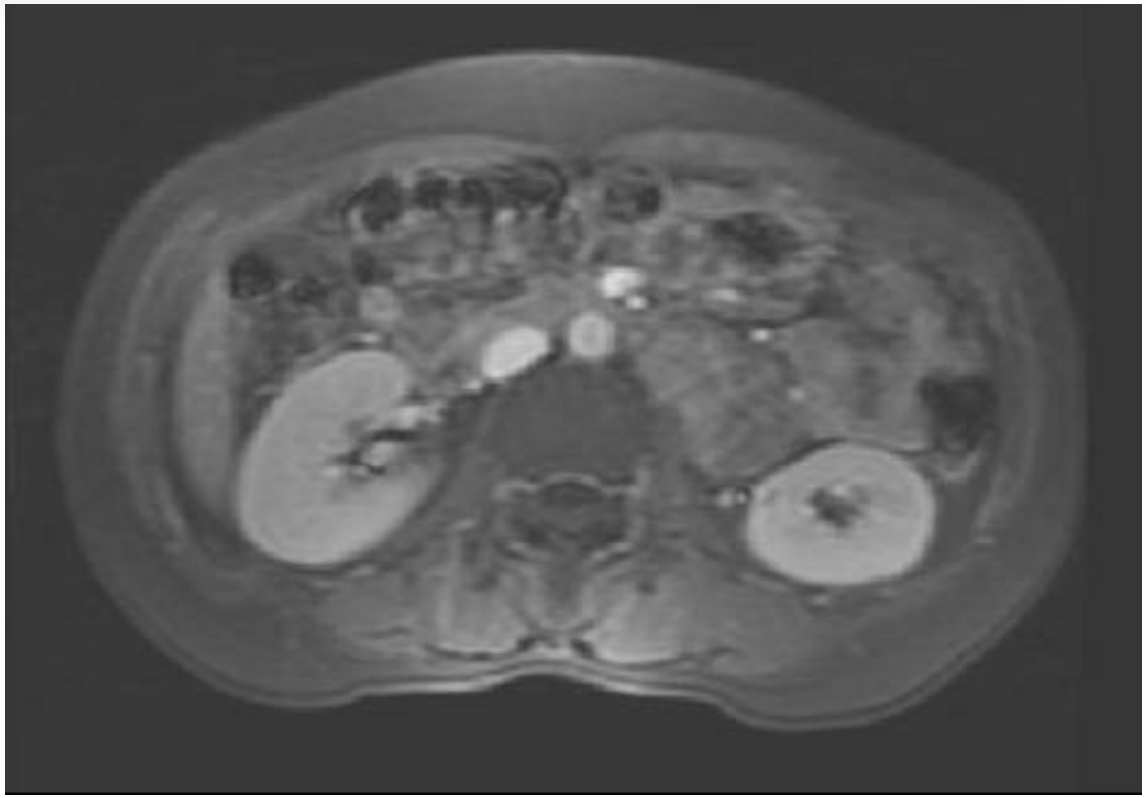
transverse view



- A large well-defined T2W high signal mass is noted at the left para-aortic region just below the left renal hilar level and anterior medial to the left kidney. The mass about 5x5x3 cm in size.

MRI T1W+C

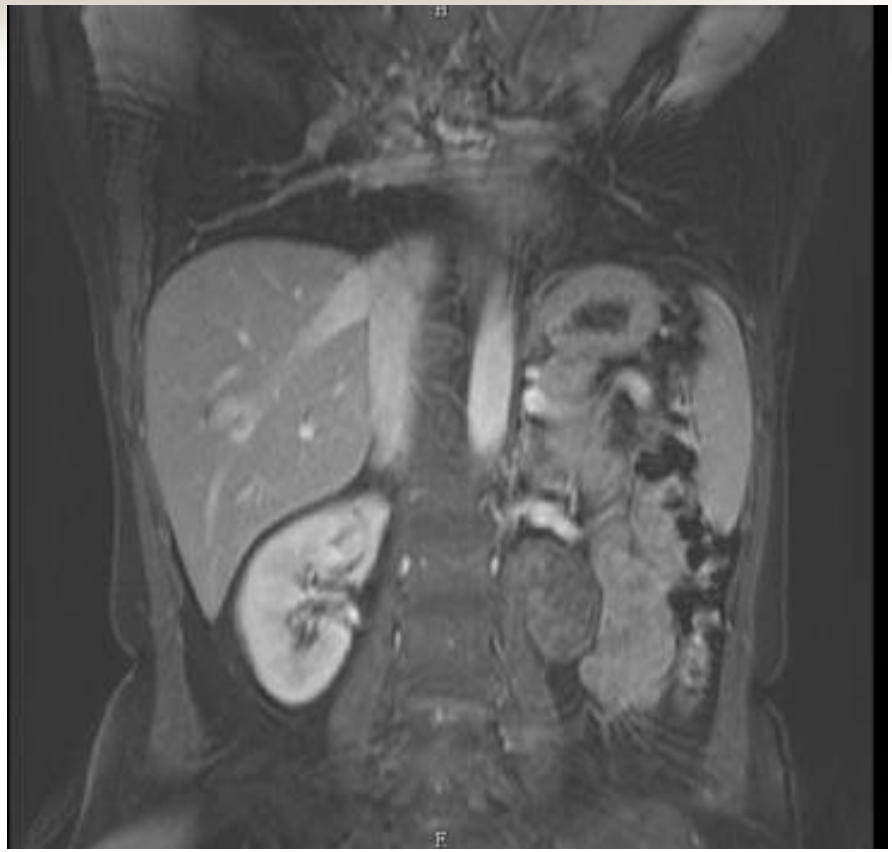
transverse view



- A large well-defined T1W low signal mass is noted at the left para-aortic region just below the left renal hilar level and anterior medial to the left kidney, whereas heterogeneous enhancement on the post-Gd images. The mass about 5x5x3 cm in size.

MRI T1W+C

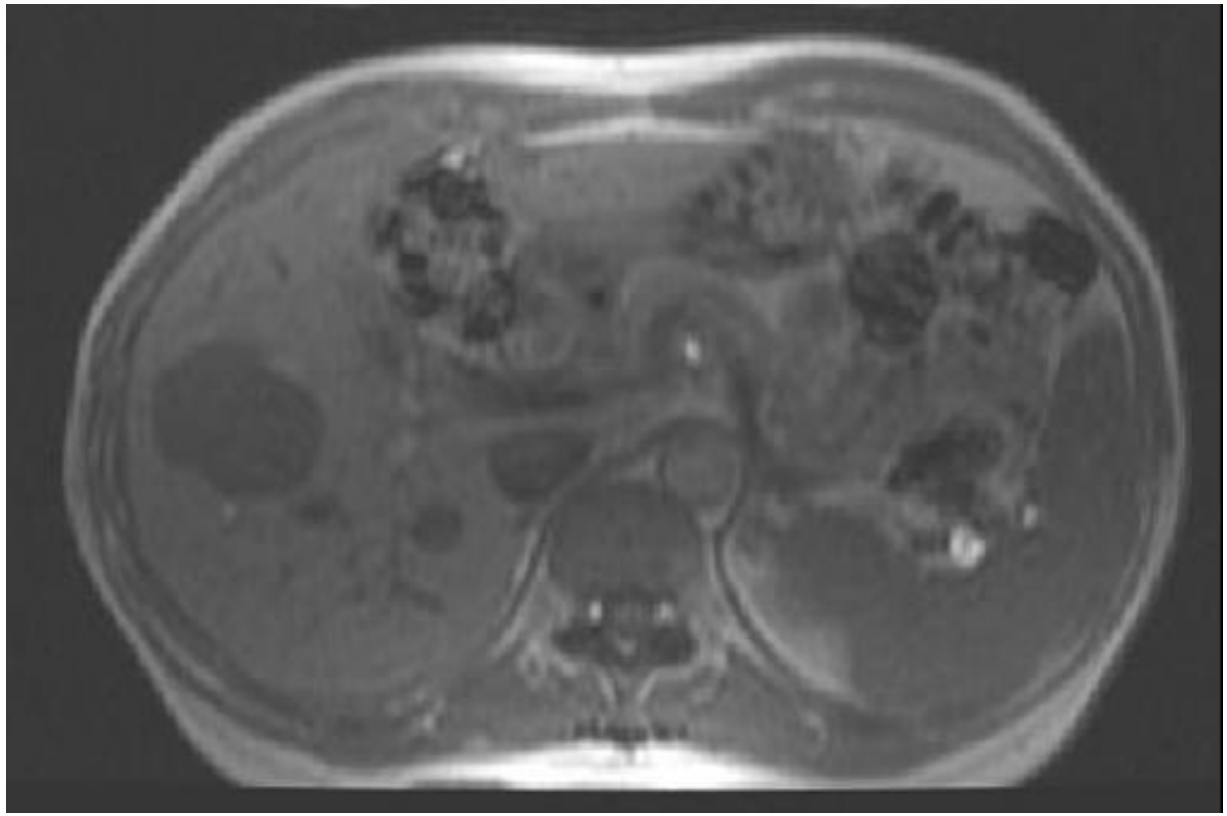
coronal view



- A large well-defined T1W low signal mass is noted at the left para-aortic region just below the left renal hilar level and anterior medial to the left kidney, whereas heterogeneous enhancement on the post-Gd images. The mass about 5x5x3 cm in size.

MRI T1W

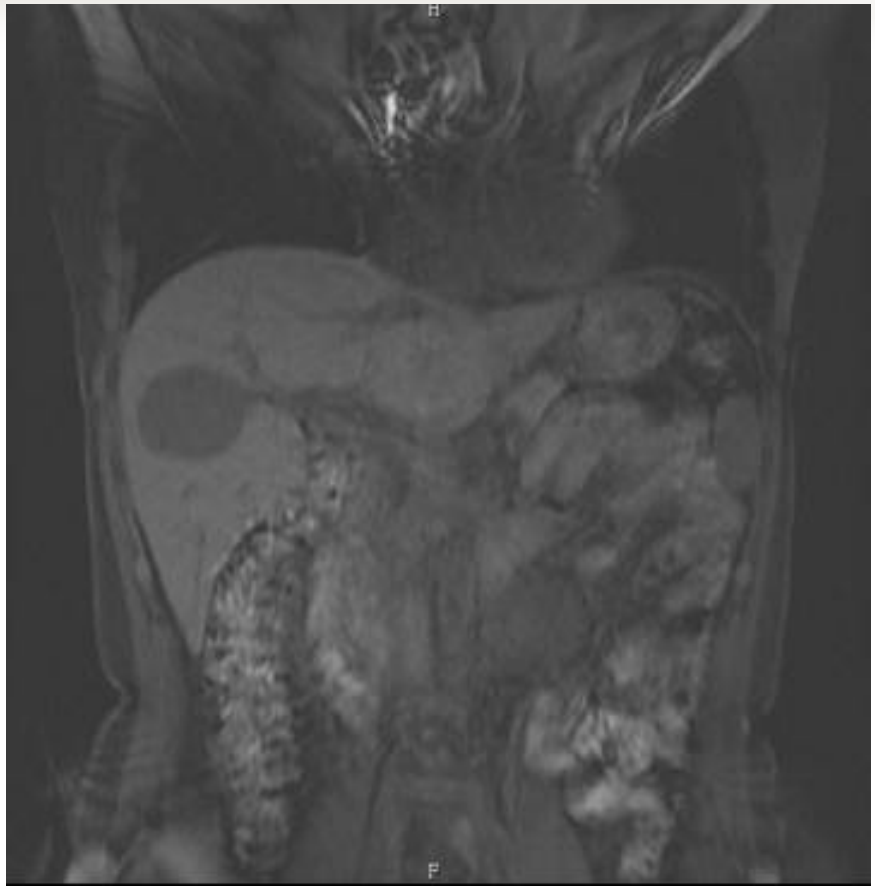
transverse view



- There is an T1W low signal irregular mass with 4.5 cm in largest diameter found in the S5 of the liver.

MRI T1W

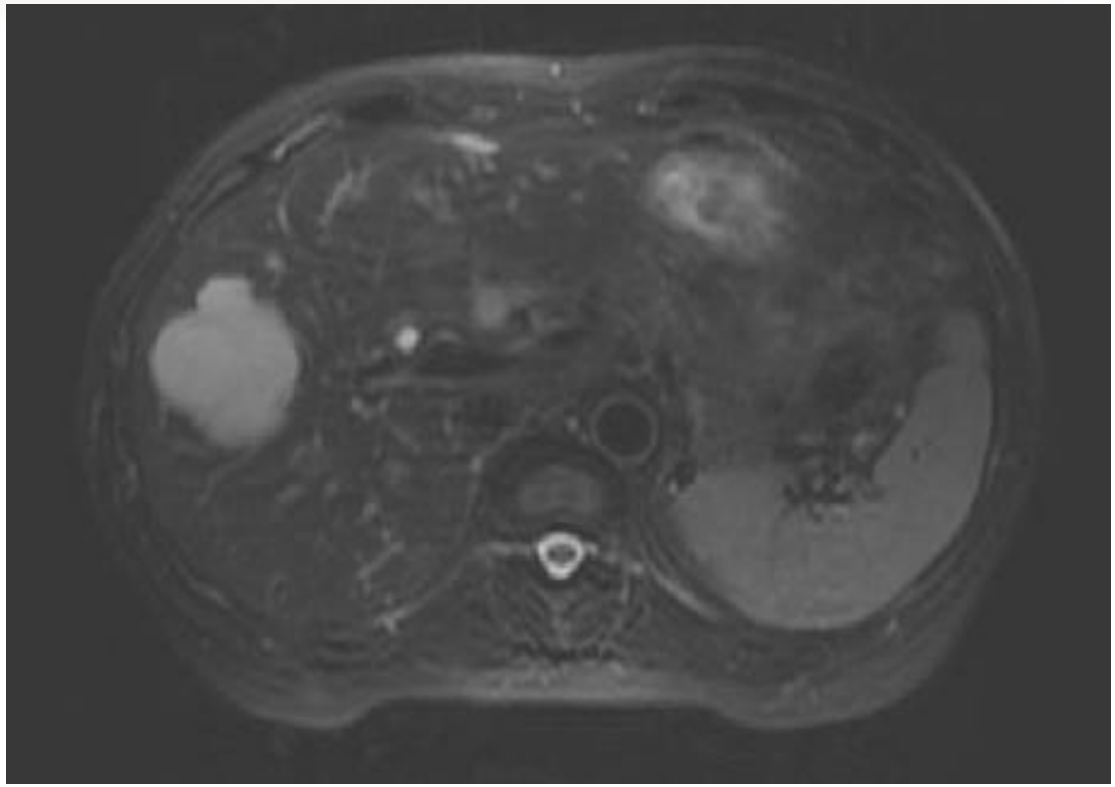
coronal view



- There is an T1W low signal irregular mass with 4.5 cm in largest diameter found in the S5 of the liver.

MRI T2W

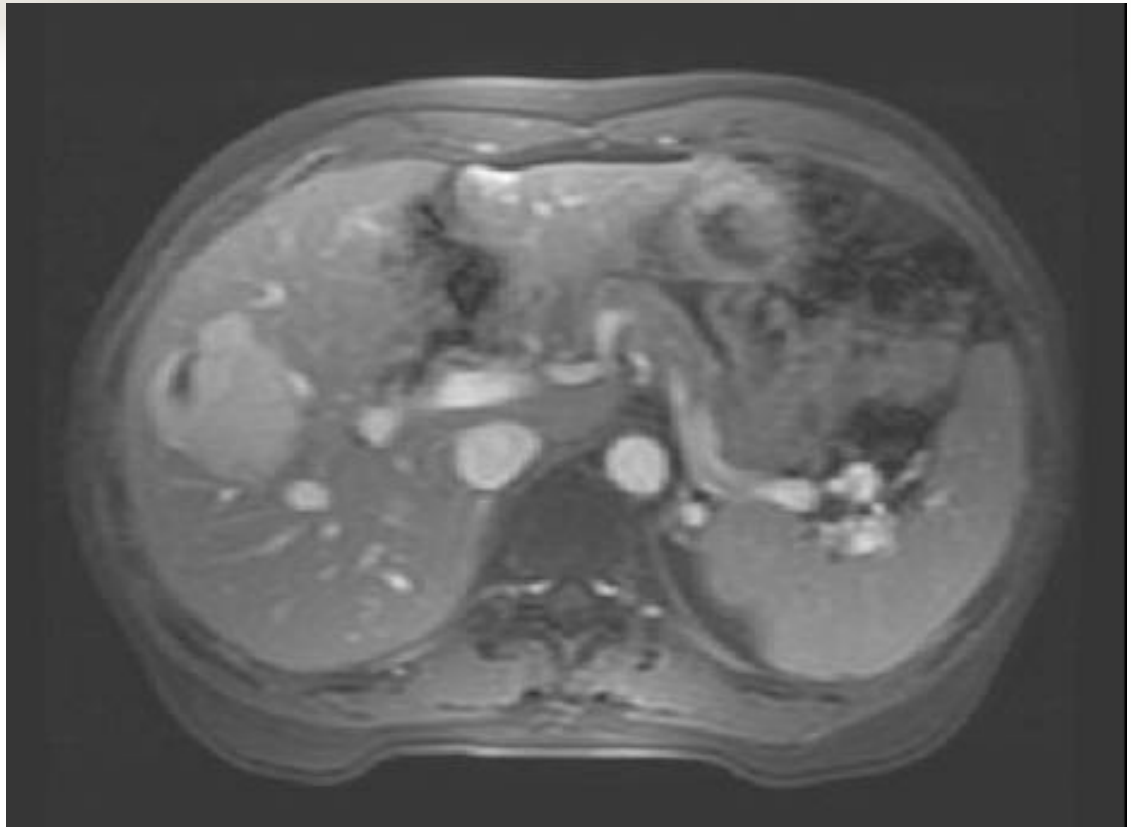
transverse view



- There is an T2W high signal irregular mass with 4.5 cm in largest diameter found in the S5 of the liver.

MRI T1W+C

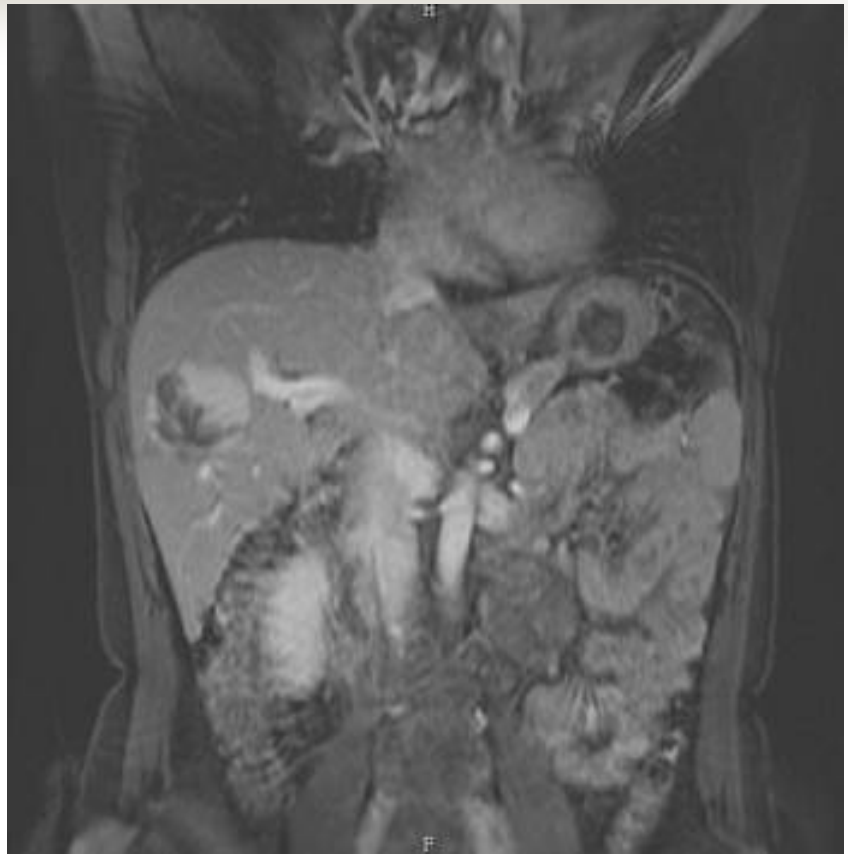
transverse view



- There is an T1W low signal irregular mass with 4.5 cm in largest diameter found in the S5 of the liver with homogeneous enhancement on the delayed venous phase.

MRI T1W+C

coronal view



- There is an T1W low signal irregular mass with 4.5 cm in largest diameter found in the S5 of the liver with peripheral contrast enhancement on the early arterial phase.

MRI

- Impression：
  Left retroperitoneal mass, just below the left renal hilar level and anterior medial to the left kidney, adrenal mass is not likely, R/o neurogenic tumor.
  S5 liver hemangioma.

Differential Diagnosis

- Neurilemmoma
- Malignant Fibrous Histiocytoma
- Fibrosarcoma
- Liposarcoma

Liposarcoma (1)

- Liposarcoma is a malignant tumor of mesenchymal origin in which the bulk of the tumor differentiates into adipose tissue.
- Liposarcoma is a common neoplasm of the soft tissues, and it affects middle-aged patients.

Liposarcoma (2)

- Commonly affected sites include the thigh, gluteal region, retroperitoneum, leg, and shoulder area.
- Liposarcomas rarely arise from a preexisting lipoma.
- Liposarcoma tumors are the most radiosensitive soft tissue tumors.

Liposarcoma (3)

- 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.

Fibrosarcoma (1)

- Fibrosarcoma is a tumor of mesenchymal cell origin that is composed of malignant fibroblasts in a collagen background.
- It can occur as a soft tissue mass or as a primary or secondary bone tumor.

Fibrosarcoma (2)

- 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.

Malignant Fibrous Histiocytoma (1)

- MFH occurs most commonly in the extremities (70-75%, with lower extremities accounting for 59% of cases), followed by the retroperitoneum.
- Tumors typically arise in deep fascia or skeletal muscle.

Malignant Fibrous Histiocytoma (2)

- Retroperitoneal MFH usually presents with constitutional symptoms, including fever, malaise, and weight loss.
- The tumor is often larger than 10 cm in diameter at presentation and may cause displacement of the bowel, kidney, ureter, and/or bladder.

Malignant Fibrous Histiocytoma (3)

- 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.

Malignant Fibrous Histiocytoma (4)

- Regions of prominent fibrous tissue (high collagen content) may demonstrate low signal on both T1-weighted and T2-weighted images.
- Calcification may present as foci of low signal on both T1-weighted and T2-weighted sequences.

Malignant Fibrous Histiocytoma (5)

- 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.
- Solid components of MFH typically reveal nodular and peripheral enhancement.
- Tumor margins appear relatively well defined on MRI. A low signal intensity margin may be observed, representing a pseudocapsule.

Neurilemmoma (1)

- 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.

Neurilemmoma (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.

Operation Note

- Pre-OP diagnosis: Left adrenal tumor
- Post-OP diagnosis: Left para-renal tumor
- OP method: Laparoscopic excision of retroperitoneal tumor
- OP findings: A para-renal tumor measured 5cm in diameter was resected

Pathology Report (1)

- The specimen submitted consists of one mass measuring 5.0x5.0x4.0 cm in size and 45 gm. in weight, in fresh state.
- Grossly, it is a well defined mass with whitish elastic capsule.
- On cut, the tumor is yellow in color and soft to myxoid in consistency.
- Focal hemorrhage and calcification is noted.

Pathology Report (2)

- Microscopically, it shows a picture of ancient neurilemoma with marked hyaline and myxoid degeneration.
- Focal nuclear palisading arrangement is noted in the tumor.
- The immunohistochemical study reveals the tumor cells are positive for vimentin, S-100, and negative for actin and desmin.
- The result also supports the diagnosis of neurilemoma.

Neurilemmoma

- Neurilemmoma is the most common neurogenic tumor.
- 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.
- No racial or sex predilection exists. Neurilemmomas affect persons aged 20-50 years.

Clinical (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.

Clinical (2)

- The head and flexor surface of the upper and lower extremities and the trunk are common locations in decreasing order.
- Tenderness to palpation is often present; secondary neurologic symptoms may be present if the tumor is large.

Labs Studies

- Laboratory studies generally are not beneficial.

Imaging Studies (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 Studies (2)

- Special studies to consider are CT scan or MRI.
- 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.

Histologic Fingdings (1)

- The correct diagnosis of NL is established by microscopic examination of tumor biopsy tissue.
- Lesions of the spinal cord often have a dumbbell shape; otherwise, they are fusiform in shape.
- They have an epineurium encapsulation, often with overlying vessels. The cut surface is pink or white.

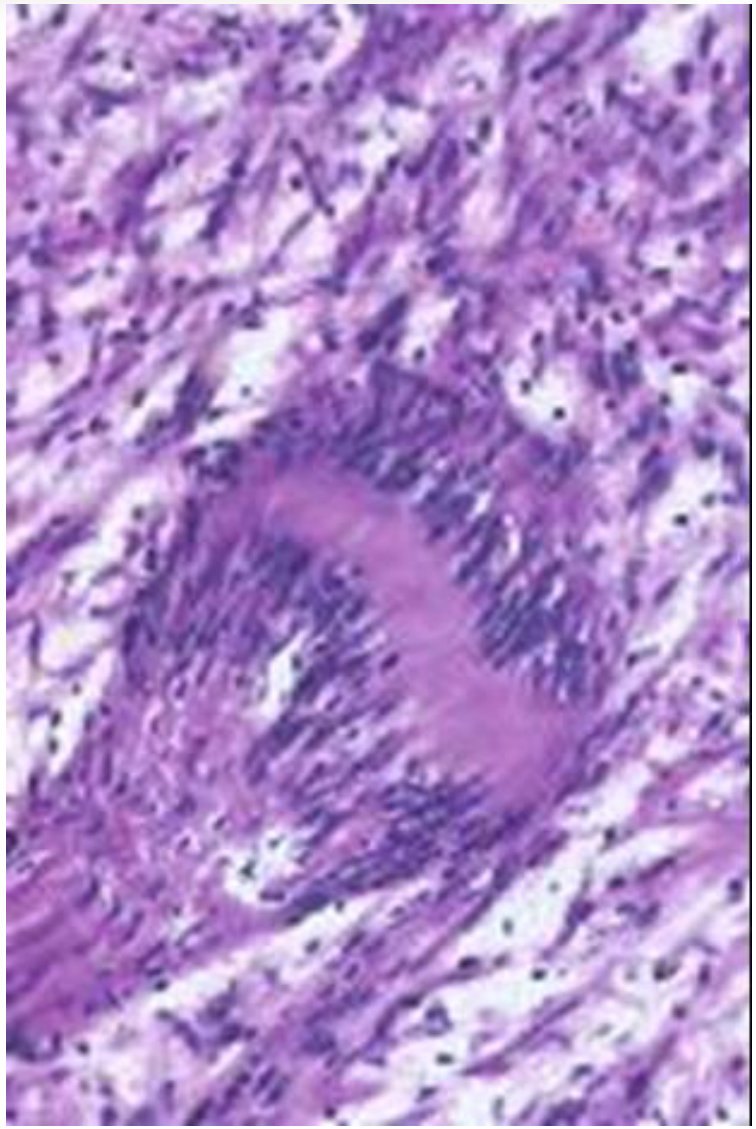
Histologic Fingdings (2)

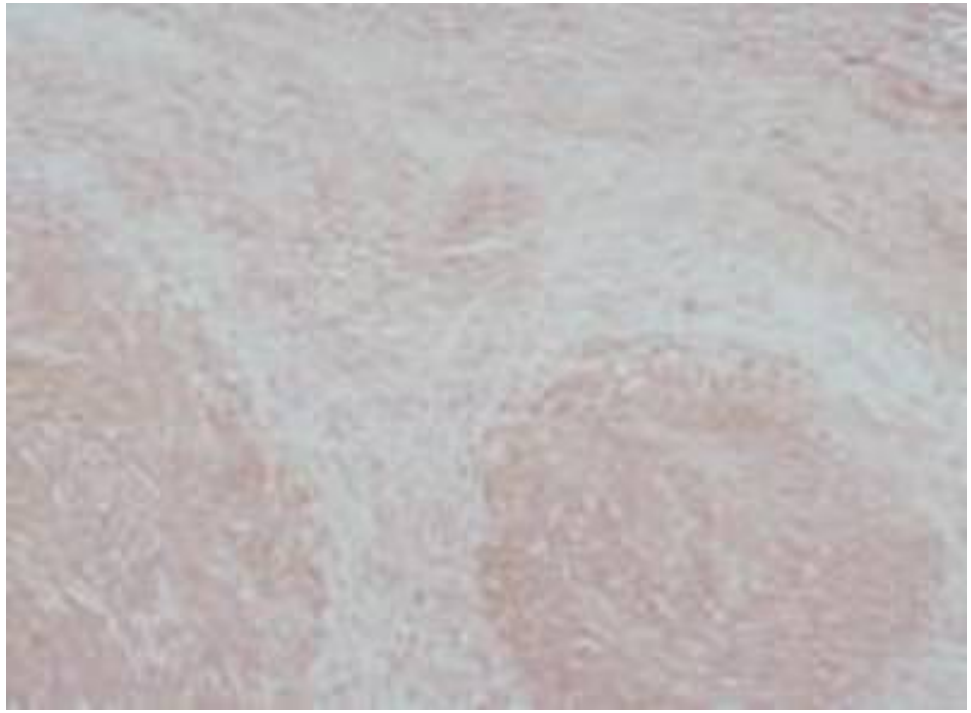
- In very large masses, degenerative cysts, hemorrhage, or dystrophic calcification may be present.
- The lesion has a well-defined fibrous capsule. Histologically, 2 distinct regions exist and are referred to as Antoni A and B regions.

Histologic Fingdings (3)

- Antoni A areas are cellular regions with predominately benign spindle cells in many intersecting bundles.
- They may palisade around eosinophilic regions that are called Verocay bodies.
- These cells are positive for S100 staining.

- A photomicrograph showing a characteristic Verocay body of a neurilemoma, consisting of tight, discrete aggregates of spindle-shaped, palisaded nuclei with a central fibrillary area, representing collections of cytoplasmic processes of tumorous Schwann cells.





- A photomicrograph of a dermal neurilemoma with anti–S-100 protein immunostaining. The tumorous Schwann cells exhibit uniformly positive staining.

Histologic Fingdings (4)

- Antoni B areas are much less cellular and have a background of loose connective tissue that is myxomatous in appearance.

Treatment & Complication

- As with most benign tumors, neurilemmomas respond well to local resection.
- The most common complication is initial neuropraxia; however, this neurologic deficit can be permanent depending on the resection of neural tissue.

Outcome & 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.


ThankYou