THE KIDNEY CONUNDRUM
Decoding Perirenal Infiltrative Processes

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Learning Objectives

1. Review and illustrate relevant anatomy of the perirenal space to understand pathways of disease spread.

2. Emphasize key clinical or radiologic differentiating features of infiltrative perirenal disease on imaging.

3. Present diverse pathology including neoplastic and non-neoplastic entities of the perinephric space.
The retroperitoneum is generally divided into 3 compartments:

1.) **Lateral compartment**  
(form formed by the renal fascia)

2.) **Great vessel compartment**

3.) **Posterior compartment**  
(containing musculature posterior to transversalis fascia)
The anterior and posterior renal fascia variably blend with fascia and connective tissue around the great vessels and posterior body wall. They are also known as Gerota and Zuckerkandl respectively.

The lateral compartment is divided into 3 spaces:

1) Anterior Pararenal Space (APS)
2) Perirenal Space (PRS)
3) Posterior Pararenal Space (PPS)

Drawing by: Jordan Mackner
Lesions of the Perirenal Space

**Neoplastic**
- Renal cell carcinoma
- Lymphoma
- Metastasis
- Retroperitoneal sarcoma
- Erdheim-Chester
- Rosai-Dorfman

**Non-neoplastic**
- Abscess
- Urinoma
- Retroperitoneal hematoma
- Lymphangiomatosis
- Extramedullary hematopoiesis
- Retroperitoneal fibrosis
Xanthogranulomatous pyelonephritis with perinephric abscess

**Clinical History**
- Fever, flank or abdominal pain, chills, hematuria, dysuria

**Interpretation pearls**
- On CT → Well-defined, low attenuation lesion w/rim enhancement & no central enhancement ± perinephric stranding ± gas within collection

**Etiology**
- UTI ascends (80%) → Pyelonephritis → liquefaction/sequestration → renal abscess
- Usually seen 1-2 weeks following pyelonephritis
- Hematogenous spread (20%)

**Prognosis/Treatment**
- Prognosis is good if caught early
- Abx therapy for small lesion + drainage for large lesions
- Surgery is rare if complicated

**Risk Factors**
- DM II, UTI, obstruction, immunosuppression

**Demographics**
- Any Age; M=F

**Top Differential**
- RCC
- Mets/Lymphoma
- Hemorrhagic or proteinaceous cyst

**Abscess**
- Urinoma
- Retroperitoneal hematoma
- Lymphangiomatosis
- Extramedullary hematopoiesis
- Retroperitoneal fibrosis
- Hypoechoic
- Perinephric Fat

**Case:** Frank Gaillard
**rID:** 16883

**staghorn calculus**
Urinoma

Clinical History
- Most common signs/symptoms
  - Pain due to mass effect; fever if infected
- Other signs/symptoms
  - Ileus, electrolyte imbalance

Etiology
- Disruption of collecting system or ureter
- Forniceal rupture (e.g., acute obstruction from stone)
- Intra- or extraperitoneal bladder rupture

Prognosis/Treatment
- Small urinomas typically do not require treatment
- Large urinomas require percutaneous or surgical drainage

Top Differential
- Hematoma
- Abscess
- Lymphocele

Interpretation pearls
- Excretory-phase with delays

Terminology
- Loculated collection of urine from urinary leak

Demographics
- Any Age; M=F

Case: Momen Mahmoud rID: 92323

Adapted from Harrigan et al. 2017
**Clinical History**
- May present with severe flank pain, palpable mass, or shock

**Treatment**
- Varies with etiology
- May manage conservatively w/o significant pathology
- Superselective embolization if unstable patient

**Etiology:**
- Trauma, tumor or cyst rupture, renal biopsy, AAA rupture, h/o anticoagulants

**Top Differential**
- Lymphoma Infiltration
- Cystic Lymphangioma
- Perinephric Abscess

**Pearls**
- CT is best diagnostic method of choice
- US ideal adjunct → Guide aspiration & assess fluid resolution over time
- Imaging follow-up is needed in spontaneous hematoma to exclude underlying mass

**Terminology**
- Hemorrhage collection in any of the perirenal spaces: Anterior pararenal, posterior pararenal, perirenal, or subcapsular

**Case:** Yaïr Glick rID: 86400
- Perirenal hematoma post lithotripsy
Non-Neoplastic

Urineoma
Lymphangiomatosis
Extramedullary hematopoiesis
Retroperitoneal fibrosis
Hypoechoic
Perinephric Fat

Abscess

Post Biopsy Hematoma

Case: Matt A. Morgan rID: 39891

Other signs/symptoms:
- Diminished hematocrit may prompt evaluation; subcapsular hematoma may cause new onset or worsening HTN

Page Phenomenon: Systemic hypertension secondary to compression by subcapsular collection (such as urinoma or hematoma)

Special Considerations
- Must identify underlying etiology in spontaneous perinephric hematoma to exclude mass (most commonly RCC or AML)
**Lymphangiomatosis**

**Clinical History**
- Most commonly asymptomatic but can present with hematuria, flank and abdominal pain
- Other signs/symptoms: HTN, ascites, infection, renal mass or insufficiency

**Treatment**
- If symptomatic (rare) conservative treatment with antihypertensives usually suffices
- Significant Sx --> percutaneous drainage, injection of sclerosing agents, resection of mass, or nephrectomy (if unilateral)

**Demographics**
- Any Age; M=F

**Synonyms**
- Renal lymphangioma, hygroma renalis, renal lymphangiectasia

**Top Differential**
- Renal/perirenal abscess
- Urinoma
- Perinephric hematoma
- PKD (ARPKD/ADPKD)
- RCC
- Perinephric lymphoma

**Best Dx Clues**
- Multilocular cystic mass in perirenal area with thin septa
- Usually bilateral
- Uni- or multilocular, cystic mass
- Medulla sparing

**Case:** Chris O'Donnell rID: 16908
Clinical History
- Primary bone marrow sites fail -> extramedullary sites take the role of blood formation
- Sickle Cell
- Myelofibrosis
- Thalassemia

Top Differential
- Mimicks lymphoma or Erdheim-Chester

Best Dx Clues
- Hepatomegaly
- Splenomegaly
- Perirenal soft tissue with NORMAL renal contour
Non-Neoplastic

Extramedullary Hematopoiesis

Thorax: paraspinal masses, rib expansion

Can include liver, pre-sacral masses, peritoneal nodules

Less Common Sites
- Lymph nodes, Skin, Breast, Brain, Adrenal Glands, Lungs, Pluera, Epidural Space
**Retroperitoneal Fibrosis**

**Non-Neoplastic**
- Abscess
- Urinoma
- Retroperitoneal hematoma
- Lymphangiomatosis
- Extramedullary hematopoiesis

**Clinical History**
- Insidious, nonspecific symptoms common → e.g., fever, malaise, weight loss, flank pain, back pain

**Treatment**
- Withdrawal/change medication
- Corticosteroids and immunosuppressants
- Surgery: Ureteral stent &/or ureterolysis/transposition

**Epidemiology/Etiology:**
- Rare (1 per 200K-500K), 40–60 years, Sex: ♂ > ♀ (2:1)
- **Primary/idiopathic RPF:** most common (70% of the cases)
  - Systemic autoimmune disease
  - IgG4-related disease
- **Secondary RPF**
  - Drugs, malignancy, infection, iatrogenic/trauma, tobacco use, asbestos exposure

**Image Pearls**
- Homogeneous soft tissue density encasing great vessels and ureters → medial deviation of ureters
- Suspect if b/l hydronephrosis of unknown etiology
- Consider bx to confirm dx and exclude malignancy

**Top Differential**
- Retroperitoneal metastases & lymphoma
- Retroperitoneal hemorrhage

**Case:** Carmen Bodlak rID: 78339

**Fibrosis:** progressive delayed enhancing soft tissue
**Clinical History**

- Exclusively identified in prediabetic and diabetic patients
- Presence may help to identify those with chronic kidney disease among prediabetic/diabetic patients


**Hypoechoic Perinephric Fat**

- **DID YOU KNOW?**
  
  Hypoechoic perirenal fat (HPF) is the sub-layer of perirenal fat immediately adjacent to the renal capsule. It is very hypoechoic on ultrasound. DO not mistake with perinephric sweat or other fluid.

**GREAT MIMIKER**

- Scalloping with subtle mass effect. Color Doppler would show internal flow.
Spontaneous Rupture w/ Renal Cell Carcinoma

Clinical History
- Most patients are asymptomatic → majority of RCC is diagnosed following an incidental finding
- Triad: hematuria, flank pain, palpable mass found in < 10%
- Can have paraneoplastic manifestations such as hypercalcemia (PTH) and hypertension (renin)

Etiology
Most cases are sporadic, but some hereditary disorders
- Smoking, acquired cystic disease of the kidney, nephrolithiasis, and long-term acetaminophen use

Erdheim-Chester Rosai-Dorfman

Renal Cell Carcinoma

Nephrectomy is first-line treatment as most RCCs are radiotherapy and chemo resistant
- Targeted and immunotherapy in metastatic disease

Top Differential
- Oncocytoma
- Angiomyolipoma (AML)
- Urothelial carcinoma
- Renal mets and lymphoma
- Renal abscess
- Hemorrhagic renal cyst

Best Imaging
- Multiphasic CT: Diagnosis and staging (cortico-medullary, nephrographic, and excretory phases)
- MR: Equal to or better than CT for staging
- Protocol advice
  - Multiphase CT

Prognosis/Treatment
- Nephrectomy is first-line treatment as most RCCs are radiotherapy and chemo resistant
- Targeted and immunotherapy in metastatic disease

Case: Mohamed Saber rID: 85034

Active bleed

Large Hematoma

Spontaneous Rupture w/ Renal Cell Carcinoma
Clinical History
- Usually asymptomatic ± flank pain, fever, weight loss

Etiology
- Most renal lymphoma is 2nd to systemic dx → Hematogenous (90%) vs. direct nodal extension
- Additionally, immunosuppression via iatrogenic (transplant) or acquired (AIDS/Autoimmune)

Demographics
- Any age or sex, < 10% lymphoma involves kidneys

Treatment
- Chemotherapy ± radiation therapy ± debulking or ureteral stenting if obstruction

Top Differential
- RCC
- Transitional cell carcinoma
- Infection

Image Pearls
- Consider lymphoma in cases of systemic adenopathy with multiple renal masses
- CECT or MR is best for eval of suspected renal lymphoma
- Nephrographic phase ideal for renal lymphoma
- Bx required for dx
**Demographics**

Peak: 50-70 years

♂ > ♀ (3:2)

**Terminology**

- Solitary plasmacytoma of bone (SPB)
- Extramedullary plasmacytoma (EMP)
- Multiple myeloma (MM)

**Image Pearls**

- When evaluating MM, be vigilant for the presence of soft tissue masses (plasmacytomas) as they have the potential to impact any organ system. Broaden your examination for signs of disease as these areas could be considered for palliative radiation therapy.

**Clinical History**

- Often asymptomatic, most common clinical feature is bone pain (especially back pain).
- Constitutional sx, hypercalcemia, spontaneous fx.
**Perirenal/Perihilar Follicular Lymphoma**

**Patient Data**
- Age: 75 years
- Gender: Male

**Presentation**
- Incidental bilateral renal/perirenal masses

**Differential Dx**
- Lymphoma
- Plasmacytoma
- Erdheim-Chester
- RPF/IgG4
- Sarcoid
- Transitional Cell Carcinoma

**Rare Considerations**
- Bilateral TCC
- Extramedullary hematopoiesis
- Amyloid

Differential provided in case by Michael Hartung rID: 81082
Melanoma Metastases: Pancreatic and Perirenal space

**Special Considerations**
- Kidney and perirenal space is the 3rd most common metastasis (after lung/breast)
- 1/3 of cases will involve kidney
- Presentation and biopsy to distinguish from lymphoma
- PET/CT should be performed prior to surgery

**History**
- Presentation: Left flank pain
- Known history of treated melanoma one year prior

**Top Differential**
- Lymphoma and Leukemia
- Metastases from other primary tumors
- Primary GI malignancies
- Kaposi sarcoma

Case: Bruno Muzio rID: 59879
Breast Carcinoma Metastases

**Special Considerations**
- Perirenal spaces are uncommon sites of metastasis → bone is most common (+65%) with lungs, liver, and brain to follow
- Many treatment options depending on type and stage
- PET/CT for staging and monitoring response to treatment

**History**
- Presentation: Patient with a diffuse metastatic disease.
- Known history of breast carcinoma → presumed to be mets based upon prior history and previous CT scans

**Differential Dx**
- Other metastases (e.g. melanoma, lung)
- Lymphoma/Leukemia
- Biopsy may be required

**Case:** Bruno Muzio rID: 20101

**Metastasis**
- Renal Cell Carcinoma
- Lymphoma
- Erdheim-Chester
- Rosai-Dorfman

**Neoplastic**
Erdheim-Chester Disease (ECD)

**History**
- Presentation: **Pulmonary** involvement most common (90%)
  - Insidious onset cough and dyspnea
- PFTs → restrictive disease, reduced DLCO

**Special Dx Considerations**
- ECD is **NEVER** unilateral → symmetric involvement of long bones, lungs, and perirenal
- **Triad:** osteosclerotic bone lesions, perirenal soft tissue, and pleural septal thickening (w/aortic wall thickening considered pathognomonic for ECD)

**Terminology**
Non-Langerhans cell histiocytosis of unknown origin

**Top Differential**
- Paget of Bone
- Lymphoma
- Rosai-Dorfman
- IgG4-Related Dx
- Lysosomal Storage Dx
Rosai-Dorfman Disease (RDD)

**ECD vs. RDD Dx Pearl**
- Clinical Differentiation
  - **ECD**: Presents in 4th-6th decades with diabetes
  - **RDD**: Children > Adults + painless, palpable lymphadenopathy

**Clinical Differentiation**

**Emperiopolesis:**
The hallmark of Rosai-Dorfman Disease.
Lymphocytes and other inflammatory cells reside undisturbed within the cytopasm of the histocytes. Histiocytes show positive staining for CD68 and S-100.

**Special Considerations**
- Rare GU involvement → most common is nodal masses (cervical)
- Common extranodal sites → nasal cavity, bone, orbit, soft tissue
- Bx required to distinguish from top differential

**Top Differential**
- Lymphoma
- IgG4-Related Dx
- Retroperitoneal fibrosis
Wunderlich Syndrome (WS)

**Etiology**
- Neoplasm (e.g. angiomyolipoma, RCC) in +60% of cases
- Vascular disorders (e.g. polyarteritis nodosa, renal artery aneurysms, AV malformation, fistulas) 20%–30% of cases of WS
- Rare cases: renal infections, cystic diseases, calculi, kidney failure, coagulation disorders, idiopathic

**Imaging Features**
- Echogenic lesion (blue arrow) representing AML amidst complex perinephric collection (yellow arrow)

**Terminology**
- A rare condition characterized by spontaneous, acute-onset renal hemorrhage into the subcapsular and perirenal spaces
- Classic manifestations → **Lenk triad**: acute flank pain, the presence of a flank mass, and hypovolemic shock

**Treatment**
- Depending on size of hemorrhage may be self-limiting
- Life-threatening bleed → Renal arterial embolization increasingly used to avoid radical surgery

**Case: Hani Makky Al Salam rID: 10090**
Understanding anatomy of the perirenal spaces is crucial for recognizing and interpreting disease processes that involve these areas.

Familiarity with common and uncommon pathologies involving the perirenal spaces can help in generating a comprehensive list of differential diagnoses, facilitating timely and effective treatment decisions.

Recognizing the distinctive clinical presentations and radiological features affecting the perirenal spaces can aid in accurate diagnosis and appropriate management.

**Take Home Points**

1. Understanding anatomy of the perirenal spaces is crucial for recognizing and interpreting disease processes that involve these areas.

2. Recognizing the distinctive clinical presentations and radiological features affecting the perirenal spaces can aid in accurate diagnosis and appropriate management.

3. Familiarity with common and uncommon pathologies involving the perirenal spaces can help in generating a comprehensive list of differential diagnoses, facilitating timely and effective treatment decisions.
References


