Tumors & Pseudotumors of the Perinephric Space
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Perinephric Space: Introduction

- Inverted cone shaped space between 2 renal fasciae
- Formed during embryonic ascent of the kidneys
- Intercommunicating space that opens into the pelvis
- Abuts the bare area of the liver & subphrenic space
- Contains the kidneys, adrenals, & proximal ureters
Solitary soft tissue mass in the perirenal space
Perirenal mass: Renal Cell Carcinoma

- Most lethal urological Ca; 30% of patients manifest metastases; 50% of patients die within 5 years
- 200,000 annual new cases worldwide; 13,000 annual deaths in the US
- >125% increase in incidence in the US (1950)
- Heterogeneous disease with variable histo-biology
- RCC: >90% of kidney masses; most common perirenal mass

* Jemal A et al. CA Cancer J Clin 2004
<table>
<thead>
<tr>
<th>Histology</th>
<th>(%) of RCCs</th>
<th>Putative Cell of Origin</th>
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</thead>
<tbody>
<tr>
<td>Clear Cell RCC</td>
<td>70</td>
<td>Proximal convoluted tubular epithelium</td>
</tr>
<tr>
<td>Papillary RCC</td>
<td>15</td>
<td>Proximal convoluted tubular epithelium</td>
</tr>
<tr>
<td>Chromophobe RCC</td>
<td>5</td>
<td>Cortical collecting duct; Type B intercalated cell</td>
</tr>
<tr>
<td>Hereditary Cancer Syndrome</td>
<td>5</td>
<td></td>
</tr>
</tbody>
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* Eble JN et al. WHO Handbook 2004
Renal Leiomyoma

- Rare, benign renal mesenchymal neoplasm
- Mostly capsular origin; less commonly renal pelvis
- Uniform solid tumor; exophytic/peripheral location
- Hypervascular tumor supplied by capsular vessels
Malignant fibrous histiocytoma

- Most common retroperitoneal soft tissue sarcoma
- 15% of MFHs occur in the retroperitoneum
- Large heterogeneous mass that displaces the kidney
Hemangioma

- Rare mesenchymal retroperitoneal neoplasm
- Renal hemangioma may extend to perirenal space
  - Arise from medulla or renal sinus
- T2-bright mass with persistent enhancement on MR
- Soft tissue mass with characteristic phleboliths
Multiple perirenal space soft tissue masses
Perirenal Metastases

- Lung/breast Ca, melanoma may rarely involve perirenal space
- Lung Ca: Proclivity due to connections between perirenal & mediastinal lymphatics
- Multiple soft tissue nodules around the kidneys
Rind-like perirenal space mass
Perirenal Lymphoma

• Usually contiguous spread from retroperitoneal dz
• B-cell NHL > common in retroperitoneum/kidney
• Isolated perirenal dz in <10% of pts; Hodgkin dz
• Multiple masses; diffuse-infiltrative; rind-like mass
• Compress kidney without affecting function
Retroperitoneal fibrosis

- Isolated or multisystem idiopathic fibrosing disorder
- Autoimmune disorders such as AIP, scleroderma
- Usually encases aorta, IVC & ureters; perirenal rare
- Stage-specific imaging findings: inflammatory tissue or fibrosis; MR more specific than CT
Erdheim Chester disease

- Idiopathic systemic non–Langerhans cell histiocytosis
- Skeletal & systemic manifestations more common
- Rind-like perirenal/periureteral soft-tissue masses
- Severe kidney compression may lead to CRF
Perirenal space mass with macroscopic fat
Angiomyolipoma

• Now classified as PEComas; sporadic or inherited
• Tuberous sclerosis complex (B/L, multiple AMLs)
• Life threatening hemorrhage (AMLs > 4cm), CRF
• Uncommon triphasic renal mesenchymal neoplasm
  – Dysmorphic vessels, fat & smooth muscle (soft tissue)
  – Lipid-poor in <5% patients; indistinguishable from RCC
Perirenal liposarcoma

- Liposarcoma: most common retroperitoneal sarcoma
- 6\textsuperscript{th}-7\textsuperscript{th} decades; frequently large at presentation
- Variable biology & rates of recurrence/metastases
- Variable fat, soft tissue & cystic components
  - Well diff: >fat; myxoid: cystic; round cell: soft tissue
Extramedullary hematopoiesis

- EMH occurs in hemoglobinopathies/MP disorders
- Liver, spleen, paravertebral areas: Embryonic hematopoietic regions
- Perirenal soft tissue masses with macroscopic fat
Perirenal myelolipoma

- Rare, benign monoclonal tumors with fat/marrow
- Commonly adrenal; extra-adrenal: perirenal/presacral
- Predominant fatty mass; variable soft tissue content
Perinephric space masses: Summary

- Wide spectrum of masses; may be renal or RP in origin
- Isolated disease or part of systemic syndrome
- Classified based on origin, distribution & morphology
- Some tumors have characteristic imaging findings
- Histopath exam after biopsy: definitive diagnosis
- Accurate diagnosis: optimal patient management
Perinephric space masses: References

• Gore RM, et al. The great escape: interfascial decompression planes of the retroperitoneum. AJR 2000
• Israel GM, et al. CT differentiation of large exophytic renal angiomyolipomas and perirenal liposarcomas. AJR 2002