Renal Mass and Nonmetastatic Hepatic Dysfunction: Stauffer Syndrome or Autoimmune Disorder

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Introduction

Renal tumors largely fall into two classes:

-Renal Cell Carcinoma (RCC).

-Renal Epithelial Stromal Tumors (REST).

Multilocular Cystic Renal Neoplasm of Low Malignant Potential (MCRNLMP) is a subtype of RCC.

-Shares most imaging/histological features of RESTs.

-Indistinguishable by most histological markers.

Adult Cystic Nephroma (ACN) is a subtype of REST

-Benign/ incidental lesion in middle-aged women.

-Presents with bloating and hematuria, mimicking RCC.

-Stromal component seen on pathology is ovarian.

Stauffer's Syndrome (SS): A rare paraneoplastic cholestatic hepatitis without metastatic disease that resolves after tumor resection.

-Associated with RCC and ovarian cancers

Autoimmune Hepatitis (AIH) and Primary Biliary Cirrhosis (PBC) also share similar symptoms and biochemical markers as SS but are differentiated by autoantibodies and histopathology.

Renal Tumor Pathology Report

DIAGNOSIS:

Left kidney, nephrectomy:
CYSTIC KIDNEY, CONSISTENT WITH ADULT CYSTIC NEPHROMA
UNREMARKABLE ADRENAL GLAND

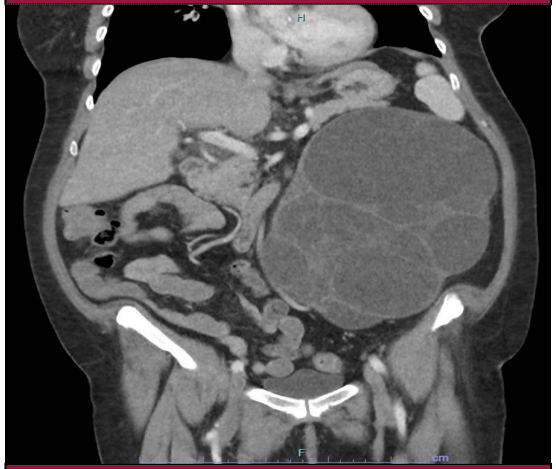
Note: Cysts are lined by flattened to hobnail epithelium, with intervening fibrotic and ovarian appearing type stroma.
Differential diagnosis includes adult cystic nephroma versus multilocular cystic renal neoplasm of low malignant potential. Adult cystic nephroma is favored.
Remainder of kidney parenchyma is unremarkable.

DIAGNOSIS:

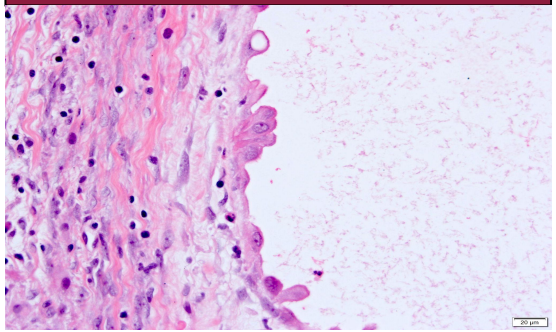
Liver, CT guided core biopsies:
MODERATE ACTIVE HEPATITIS AND HEPATOCANALICULAR CHOLESTASIS.
NEGATIVE FOR MALIGNANCY.

Note: Case reviewed at the Mayo Clinic in Rochester Minnesota (Mayo case number CR20-52198). They observed moderate degree of portal inflammation comprised of lymphocytes, plasma cells and eosinophils. There is prominent lobular inflammation in zone 3 with rare acidophil bodies are identified. There is minimal steatosis. Trichrome stain shows portal fibrosis, mild. CK-7 highlights bile duct proliferation/regenerative type. Immunostains for IgG and IgM show rare plasma cells. Iron stain is negative. They believe the findings most likely represent adverse drug/toxin reaction. Differential includes autoimmune hepatitis although this less likely. Features of primary biliary cirrhosis are not seen in this biopsy. Complete report shared with [redacted]

CT Scan



Cystic Nephroma Pathology



Case Presentation

A 62 year old woman with a 12-year history of a multicystic left-sided renal mass presented with 10 days of worsening jaundice.

CT Abdomen: Imaging revealed that the renal mass had doubled in size and new portal lymphadenopathy.

Labs: Consistent with cholestasis, positive AMA titer of 1:80.

Liver biopsy: Nonspecific inflammation with associated cholestasis.

Mgmt: Left radical nephrectomy was performed

Kidney pathology: Consistent with ACN vs MCRNLMP

-ACN was favored.

The patient's liver function tests improved post-nephrectomy and continued to improving after discharge.

6 months later she developed jaundice, hypergammaglobulinemia, and ASMA and AMA titers of 1:160. Repeat imaging revealed stable portal lymphadenopathy but no masses. She was diagnosed with AIH/PBC overlap syndrome and is improving on treatment.

Conclusion

REST and RCC are often difficult to distinguish.

-Particularly ACN and MCRNLMP.

SS and AIH/PBC share most symptoms, biochemical markers, and the immune dysregulation is even caused by similar mechanisms.

In this case, the patient's initial improvement post-nephrectomy supports the theory that her hepatic dysfunction was partly due to the tumor, although the recurrence was due to an occult autoimmune disease.

-Did a 12-year history of ACN cause a seronegative immune response to become a seropositive immune response?

This case highlights clinical ambiguity in the diagnosis of renal tumors, the need for prompt tumor resection if paraneoplastic syndromes are suspected, and the need to differentiate paraneoplastic disorders from occult autoimmune disorders.