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Lysozyme-Induced Nephropathy Secondary to Chronic Myelomonocytic Leukemia

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Primary Renal Squamous Cell Carcinoma: Demographic Features and Survival Analysis

(Poster No. 80)

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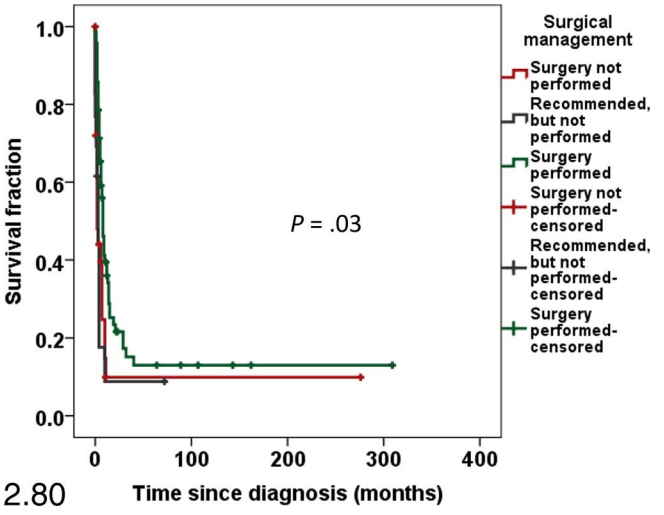
Context: Primary renal squamous cell carcinoma (SCC) is a sporadic tumor, accounting for less than 1% of all renal malignancies. Squamous metaplasia induced by long-standing nephrolithiasis is considered the most important etiologic factor. Because of its rarity, the epidemiologic characteristics have not been systematically investigated. Moreover, the factors affecting patients' outcomes of this neoplasm are unknown.

Design: We queried the National Cancer Institute's Surveillance, Epidemiology, and End Results database for all histologically confirmed SCC with kidney designated as the primary site. All data were transferred to SPSSv.25, and analysis was performed to demonstrate descriptive statistics. We used Kaplan-Meier curves, log-rank test, and multivariate regression analysis to test the effect of each variable on the patients' overall survival.

Results: One hundred forty patients of primary renal SCC were found in the database. Our analysis shows that most of the patients were diagnosed after 70 years, with a predominantly white population (85%) and a slight male predominance (56%). Surgical treatment of the primary site was the choice of management for the majority of the patients. Our survival analysis shows that age at diagnosis, sex, or race of the patients did not affect their disease-specific survival. However, patients who received surgical management of the primary site showed significantly better outcomes than patients who received other management options (Figure 2.80).



2.80

Conclusions: Our data suggest that age at diagnosis, sex, or race do not affect the outcome of primary renal SCC patients. However, the patients with this malignancy managed with surgery of the primary site do significantly better.

Adrenal Endothelial (Vascular) Cysts: A Rare Entity

(Poster No. 81)

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Context: Adrenal endothelial cyst, also known as vascular adrenal cyst, is a rare entity with female predilection, usually presenting with abdominal pain. Smaller cysts can be detected as incidental findings during surgery or imaging.

Design: We retrospectively reviewed our archives for adrenal cysts and identified cases with the diagnosis of "endothelial" or "vascular" cyst. For each case, we recorded the patient's age, gender, race, size of the lesion, clinical presentation, and disease associations.

Results: Six cases were identified over the course of 30 years. Five of the patients were female (83%). The patients' ages ranged from 29 to 46 years (average, 37.8), and the majority of the patients were white (83%). Four cases (67%) were on the left side. The size of the cysts ranged from 5.2 to 11.2 cm (average, 8.2 cm) (Table). Two patients presented with left-sided abdominal pressure/pain. None of the patients had prior abdominal surgery, and none of the cases recurred. No other vascular lesions or tumors were present.

Conclusions: Adrenal endothelial cysts are rare, isolated lesions affecting adult white women in our population. The cysts can be large and symptomatic, and they do not recur after surgical excision. Awareness of this entity is important in patients with adrenal masses or cysts.

Summary of Study Cases

Case	Age, y/Sex	Race	Side	Size, cm
1	29/M	W	Left	10.5
2	46/F	W	Left	7.1
3	29/F	W	Left	10.1
4	45/F	W	Left	5.2
5	33/F	B	Right	5.2
6	45/F	W	Unknown	11.2

Lysozyme-Induced Nephropathy Secondary to Chronic Myelomonocytic Leukemia

(Poster No. 82)

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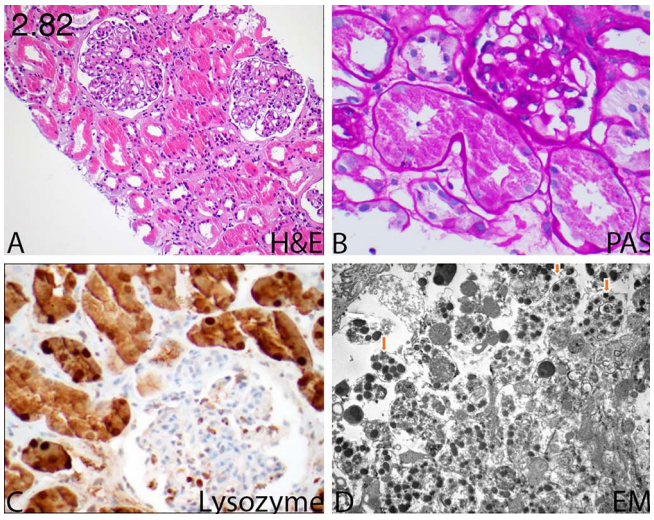
Lysozyme-induced nephropathy is a rare cause of acute kidney injury (AKI) in patients with chronic myelomonocytic leukemia (CMML). We report the case of an 85-year-old man with type II diabetes and hypertension who was being evaluated for an elevated serum creatinine of 2.27 mg/dL. Additional studies showed proteinuria (1 g), predominantly nonalbumin, along with an IgA λ monoclonal protein found on serum electrophoresis. His serum κ:λ ratio, however, was elevated and urinalysis showed free κ monoclonal protein. Further hematopathologic workup revealed CMML, and he also had progression of his AKI, for which a renal biopsy was performed. Light microscopy showed unremarkable glomeruli with dilated proximal tubules containing granular and strongly eosinophilic cytoplasm (Figure 2.82, A and B). Immunohistochemical staining for lysozymes showed strong positivity



in the cytoplasm and nuclei of the proximal tubule cells (Figure 2.82, C). Electron microscopy showed expanded lysosomes with rounded, dense dots arranged in an annular "chocolate chip cookie" pattern ranging from 1.0 to 1.5 μm in diameter (Figure 2.82, D). No monoclonal proteins were detected by immunofluorescent stains in the renal biopsy. Plasma lysozyme level was >10.8 μg/mL (reference range, 2.6–6.0 μg/mL). Taken together, these data support the diagnosis of lysozyme-induced nephropathy likely secondary to overproduction of lysozymes from his CMML. He began treatment with azacitidine, and following 1 cycle of therapy, he had a reduction of his serum creatinine to 1.67 mg/dL after 1 month follow-up. Awareness and recognition of lysozyme-induced nephropathy as a cause of AKI in patients with CMML is important as it may have potential diagnostic and therapeutic considerations.

Renal Cell Carcinoma With (Angio)Leiomyomatous Stroma Associated With Recurrence

(Poster No. 83)

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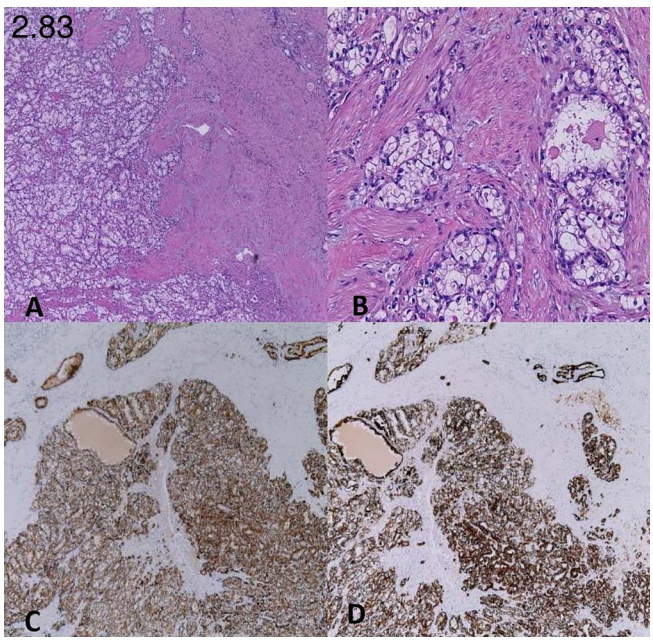
Renal cell carcinoma with (angio)leiomyomatous stroma (RCCLMS) is a poorly described entity that is known to be a benign lesion with an indolent course. This case of RCCLMS is the first with an associated recurrence. We report a case of RCCLMS in a 73-year-old woman with a past medical history significant for right renal cell carcinoma associated with prominent smooth muscle stroma status post right partial nephrectomy in 2009. She presented in 2021 with a 2-cm lesion involving the anterior cortex lateral to the hilar area and just lateral to the likely site of her previous resection. She underwent right partial nephrectomy. Gross examination revealed a 1.4 × 1.2 × 1.1-cm tan-white well-circumscribed lesion. Microscopic analysis revealed a well-circumscribed neoplasm composed of clear cells arranged in nests and tubules within a stroma rich in smooth muscle (Figure 2.83, A). The epithelial cells had abundant clear to light eosinophilic cytoplasm and nuclei with variably conspicuous nucleoli. A rich vascular network was present surrounding the epithelial nests (Figure 2.83, B). No tumor necrosis or angiolymphatic invasion was present. Immunohistochemical stains show the epithelial cells were positive for AE1/AE3, carbonic anhydrase IX (Figure 2.83, C), CK7 (Figure 2.83, D), and CD10, and negative for racemase (p504S), HMB-45, and Mel-A. The smooth muscle stroma is positive for smooth muscle actin and negative for pankeratin, HMB-45, and Mel-A. The prior report for the renal cell carcinoma associated with prominent smooth muscle stroma from 2009 strongly suggests that this is a recurrence of the same entity.



Aberrant Expression of CDX-2 in Metastatic Adenocarcinoma

(Poster No. 84)

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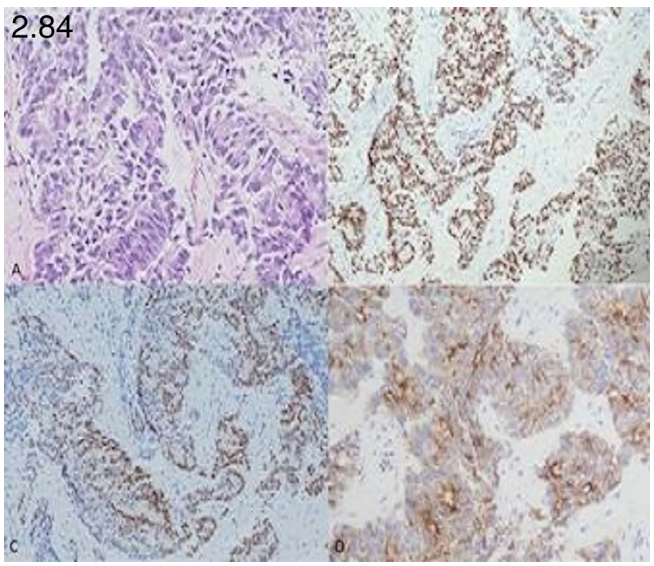
Context: CDX-2 is a marker of intestinal differentiation. Recently it has been reported to be positive in few primary prostatic adenocarcinomas (PCAs); however, expression in metastatic prostatic adenocarcinomas (MPAs) is not well documented. We present the analysis of CDX2 expression in MPA.

Design: After a computer search, 9 cases of MPA were included in the study. Review of clinical history, imaging, and pathologic findings was performed.

Results: Of 9 cases studied, the metastatic sites included 5 regional LNs, 3 bone, and 1 adrenal. All cases showed expression of prostate-specific antigen (PSA) or NKX3.1. Only 1 case showed CDX2 expression. The patient was a 63-year-old man with no prior history of malignancy, who presented with urinary frequency and weight loss. Imaging studies revealed enlarged prostate, lymphadenopathy, and sclerotic bone lesions. The PSA level was 104 ng/mL. Biopsy of retroperitoneal LN showed infiltration by tumor cells with cribriform architecture and prominent nucleoli (Figure 2.84, A). The tumor cells were positive for NKX3.1 (diffuse) (Figure 2.84, B), p504S (Figure 2.84, C), and CDX-2 (moderate, 40% cells) (Figure 2.84, D), and negative for PSA. The morphologic features and immunophenotype were consistent with metastatic prostatic adenocarcinoma. Next-generation sequencing analysis showed no significant alterations for therapy.



Conclusions: CDX2 can show moderate expression in rare cases of MPA. Awareness of such an unusual occurrence will help to avoid misdiagnosis as metastatic gastrointestinal carcinoma, especially during the histologic assessment of a metastasis from an unknown primary. PSA and/or NKX3.1 must always be included in the immunohistochemical panel for workup of metastasis from unknown primary.

An Uncommon Cause of Arterial Hypertension in Children: Juxtaglomerular Cell Tumor

(Poster No. 85)

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