

Rare Diagnosis for a Renal Mass: Adult Variant Wilms' Tumor

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History and Physical Examination

-28 year old, 30 week gestation, para 1-0-1-1 female presented with sudden/severe left flank plain

-she denied history of UTI, pyelonephritis, or nephrolithiasis, hematuria, and constitutional symptoms

-Physical examination

- -severe tenderness to palpation of the in the left flank
- -prominent abdomen secondary to 30 weeks gestational age

-Initial imaging (in office)

-office ultrasound examination demonstrated a large left flank mass; patient then referred to radiology

Imaging CT Axial Plane



Imaging CT Coronal Plane

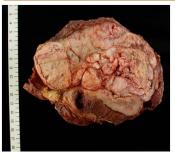


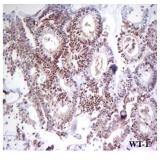
-Findings

-heterogeneous left renal mass (14 x 12 cm), which engulfs the left kidney

-mass effect on aorta; merges with left psoas muscle; regional left retroperitoneal lymphadenopathy; no renal vein invasion; normal contralateral kidney; numerous parasitic collateral vessels in left retroperitoneum; no metastatic disease to lungs -differential diagnosis: renal cell carcinoma, lymphoma, angiomyolipoma with hemorrhage, retroperitoneal sarcoma, or other malignancy of retroperitoneum or kidney

Pathology





-Ultrasound guided biopsy

-18 gauge sharp Hawkins advanced into pass; 16 gauge biopsy gun passed through Hawkins cannula; 5 core biopsies obtained

-Pathology

- -Gross specimen
- -Positive WT1 (Wilms' tumor protein 1), Pancytokeratin AE1/3, and CD56
- -Tubular epithelial cells and small blue cells concordonat with triphasic pathology of Wilms' tumor

Treatment

-Treatment Plan

- -staging: Stage III Wilms' Tumor
- -delivery of fetus
- -nephrectomy, lymphadenectomy
- -total abdominal radiation (unique to stage III/IV therapy)
- -chemotherapy

Discussion

-Incidence

- -250 cases of adult Wilms tumor have been documented in the literature
- -< 2% of Wilms tumors occur in individuals over the age of 15 years
- -Adult Wilms tumors are rare, with an annual incidence of less than 0.2 cases per million (5). However, Wilms tumor is one of the most common renal tumors in childhood, with an annual incidence in children younger than 15 years of 10 cases per million

-Diagnostic Criteria for Nephroblastoma in Adults

-primary renal neoplasm, primitive blastomatous spindle or round cell component, formation of abortive or embryonal tubular or glomeruloid structures, no areas of hypernephroma, pictorial confirmation of histologic findings,

Prognosis

age >15 years

-5-year survival rate: 73.7% (localized tumor), 47.5% (regional extension), and 14.7% (metastatic tumor)