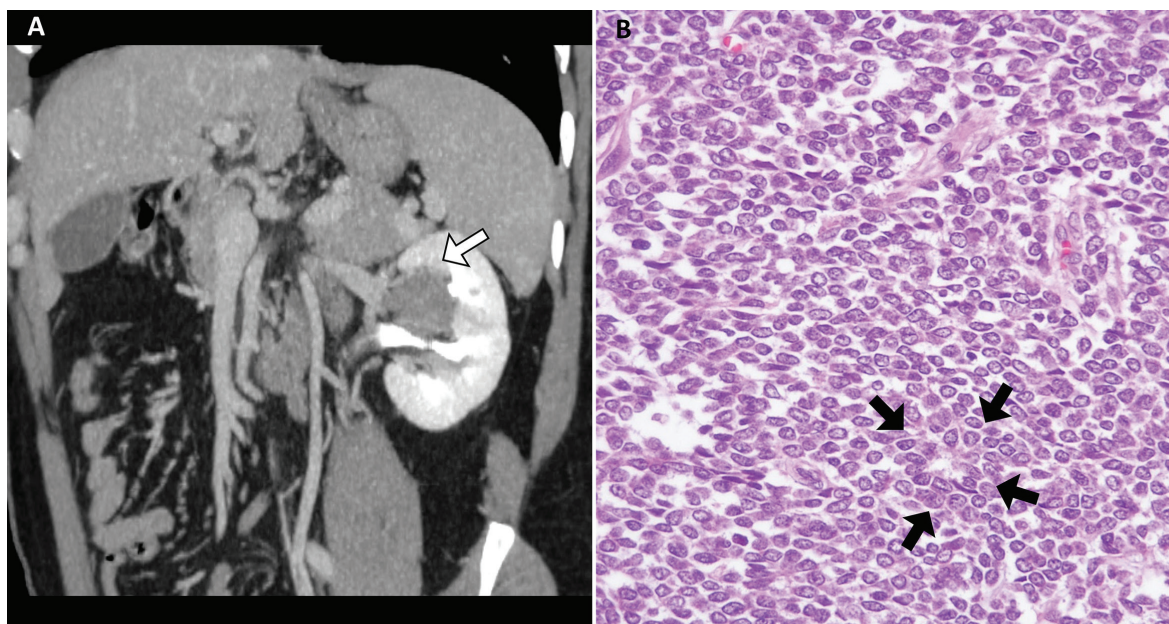


Renal Ewing Sarcoma: A Challenging Diagnosis

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(A) Coronal images of abdominal CT of the patient showing a large multilobulated heterogeneous enhancement lesion occupying the central region of the left kidney with an extension of tumor thrombus (arrows) in the left renal vein. (B) Histopathology of the resected specimen showing sheets of small, blue, round uniform cells with scant clear cytoplasm divided into irregular lobules by fibrous strands. Tumor cells are uniformly small and exhibit a high nuclear-cytoplasmic ratio, and might present as rosette arrays with central lumens, known as the Flexner-Wintersteiner rosette (arrow).

A 33-year-old man with a history of psoriasis presented with left flank pain developed insidiously over several months. The patient had no hematuria, dysuria, fever, or bodyweight loss. He had no medical history of renal calculi, trauma, or illnesses related to the symptom. Physical examination revealed left flank tenderness with otherwise normal back and abdominal examinations. Laboratory evaluations and urinalysis are normal. Abdominal computed tomography (Figure A) showed a large multilobulated heterogenous enhancement lesion in left kidney with an extension of tumor thrombus in the left renal vein. Then, radical nephrectomy was performed, and pathology revealed a renal Ewing sarcoma. At the 4-week interval after surgery, the patient was treated with 12 cycles of adjuvant chemotherapy every 3 weeks with vincristine, doxorubicin, cyclophosphamide, ifosfamide, etoposide, and dactinomycin. No signs of recurrence were observed 9 years after the diagnosis.

Primary Ewing sarcoma in the kidney is extremely rare. Histologically, Ewing sarcoma presents as small blue round cells and several neuroblastic Homer Wright rosettes, a Flexner-Wintersteiner rosette (Figure B, arrows), or pseudorosettes. Diagnosis of renal Ewing sarcoma can be challenging, but it can be confirmed using the positive immunohistochemical staining for CD99.^{1,2} Aggressive treatment with radical nephrectomy, and adjuvant combination chemotherapy is recommended as the principal management strategy.

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