A rare case of adult Wilms' tumor

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Although Wilms' tumor is the most common primary renal malignancy in children, it is exceedingly rare in adults and has an estimated incidence of less than 0.2 cases per million. Little is known about the biology of this tumor in adults and clinicians have had to rely on pediatric treatment protocols. Overall, prognosis is worse in adults, though like in children, unfavorable histology and higher stage at presentation confer a worse prognosis.

Key Words: Wilms' tumor, nephroblastoma, adult

Case report

A previously healthy 31-year-old man sought medical attention for abdominal pain after suffering a fall during a soccer game. As part of his initial evaluation, he underwent an enhanced CT scan of the abdomen which revealed a large, complex 9 cm x 13 cm x 13 cm exophytic mass with heterotopic calcifications effacing the upper and middle poles of the right kidney, Figure 1. There were several prominent interaortocaval lymph nodes in close proximity. Other than vague abdominal pain associated with his recent blunt trauma, he denied any recent history of flank pain, hematuria, fever, weight loss, or any other symptoms. Given its large size and atypical appearance, the tumor was subsequently staged with a CT scan of the chest and a bone scan, neither of which showed any evidence of distant metastases.

At our recommendation, the patient then underwent an open right radical nephrectomy, adrenalectomy, and regional lymphadenectomy. The surgery itself was uncomplicated. Grossly, the tumor did not involve any adjacent viscera. When examined, the specimen had a thick capsule and, when bisected, had areas of focal necrosis and hemorrhage. Pathologic examination revealed a Wilms' tumor with favorable histology, i.e., the presence of all three classic elements – blastema, epithelium, and stroma – and the absence of anaplasia, Figure 2. On immunohistochemistry staining, there was intense uptake for the gene product of *WT1*. The surgical margins were negative. The adrenal gland and perirenal and retroperitoneal lymph nodes were uninvolved. After an uneventful recovery from his operation, the patient underwent an 18 week course of adjuvant chemotherapy with dactinomycin and vincristine.

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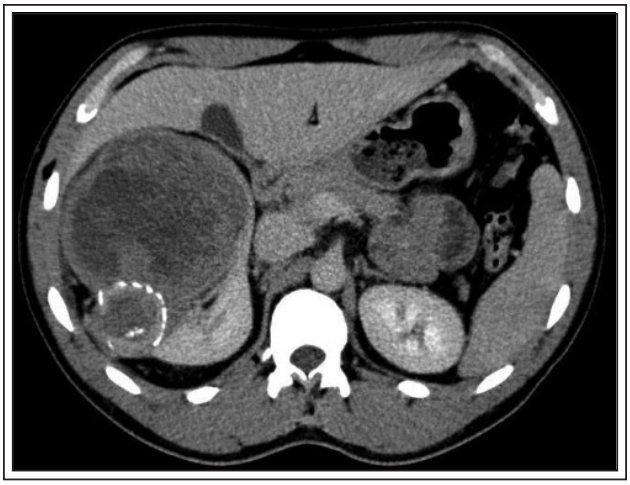

Figure 1. CT of the abdomen showing a large, complex mass effacing the upper and midpoles of the right kidney.

Literature review

Wilms' tumor, or nephroblastoma, arises from remnants of immature kidney tissue and, as a consequence, occurs primarily in childhood. It is the most common primary renal malignancy in children and has a median age of presentation of 3.5 years.[1] It is exceeding rare in adults and has an estimated incidence of less than 0.2 cases per million in the United States and Europe.[2] Because it occurs so infrequently in adults, the collective understanding of this tumor and its biology in this subset of patients is limited. Clinicians have had to rely on data from the pediatric literature to stage, treat, and prognosticate these lesions in the adult population. Most of the experience with Wilms' tumors in adults has been published in case reports and retrospective series.[3-6] Despite these limitations, some generalizations can still be made with the data at hand. In a retrospective review of 143 adults with Wilms' tumor diagnosed between 1983-1994, Mitry and associates showed that the overall prognosis is worse in adults. The overall survival in their series was 69.9% (95% CI 61.8%-78.0%) at 1 year and 47.3% (38.2%-56.4%) at 5 years. In more contemporary series, overall survival was 83% at 4 years and 62.4% at 5 years.[5,7] This is a stark contrast to the pediatric population where overall survival is over 90% at 4 years in patients with favorable histology. To identify potential clinical variables that pre, Izawa and associates performed univariate and multivariate analysis on survival data on 128 patients and showed that unfavorable histology and advanced stage predicted worse overall survival and disease specific survival.[8] Clearly, more study is needed to better characterize this tumor as the entire body of medical literature is limited to several hundred cases.[4] □

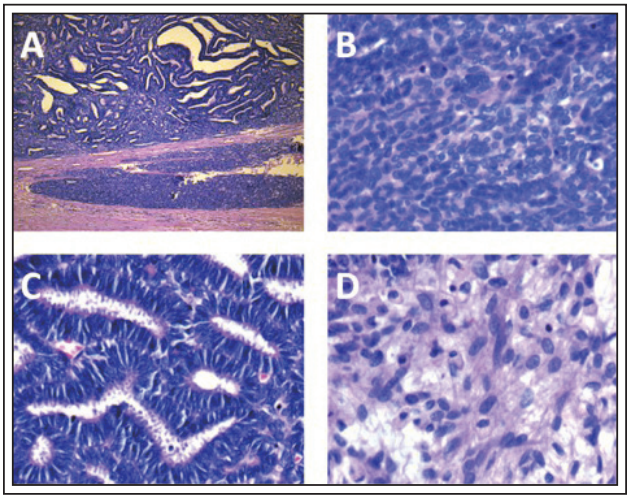

Figure 2. Pathology from the specimen showing (A) all three elements of classical Wilms' tumor, (B) nests of blastema, (C) epithelial elements, and (D) stroma.

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