CASE REPORTS

Wilms Tumor: An Uncommon Entity in the Adult Patient

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ABSTRACT

Wilms tumor, the most common kidney tumor in children, is rarely seen in adults, making it a challenge for the adult oncologist to diagnose and treat. Unlike with renal cell carcinoma, patients with Wilms tumor should receive adjuvant chemotherapy with or without radiation therapy. Adult oncologists may not be familiar with pediatric oncology protocols, so it is important to consult with pediatric oncologists who have more experience in this disease. Multimodal therapy based on pediatric protocols improved the outcomes of adults with Wilms tumor worldwide. We report a rare case of a 24-year-old woman with a slow-growing mass of the left kidney during a 4-year period. The mass was surgically removed and final diagnosis confirmed by pathology to be Wilms tumor. The patient received adjuvant chemotherapy and has been free of disease since 2014.

CASE PRESENTATION

We report a case of a 24-year-old woman who was incidentally found to have a 2-cm left kidney mass during an evaluation after a motor vehicle accident in 2010. Fine-needle aspiration performed at a local tertiary hospital revealed metanephric adenofibroma, which is one of the rarest benign renal tumors. She had had recurrent urinary tract infections since 2013. A routine ultrasound during her pregnancy in 2012 showed a slight progression in the left kidney mass. In April 2014, a follow-up contrast computed tomography scan of the abdomen and pelvis revealed a 6.4 × 4.8 cm left upper pole kidney mass. She underwent left laparoscopic radical nephrectomy and adrenalectomy with para-aortic lymph node resection.

Sections of tumor show a multinodular neoplasm (Figure 1). Histologic patterns range from sheets of small round blue cells to areas with tubule formation (Figure 2). The tumor is predominantly composed of epithelial and blastemal elements, with scant stromal elements present (Figure 3). Areas of necrosis and frequent mitotic figures are present, but no areas of anaplasia are seen (favorable histology). Immunohistochemical stains show patchy nuclear staining for Wilms tumor 1 (WT1) protein, patchy areas positive for cytokeratin 7, and diffusely strong positive nuclear staining for PAX-8 and CD56. Staining for CD57 and CD99 are negative. Cytogenetic testing revealed normal female chromosome analysis 46, XX [20]. The immunohistochemical profile and the morphology support the diagnosis of Wilms tumor.

We presented this case at a multidisciplinary pediatric oncology tumor board. Adjuvant chemotherapy is the standard of care for patients with favorable histology in the early stage of Wilms disease. The patient completed a full course of adjuvant chemotherapy per the National Wilms Tumor Study Roadmaps pediatric protocols to treat Wilms disease.1 Her chemotherapy regimen included dactinomycin, vincristine, and doxorubicin per protocol.1 A follow-up computed tomography scan of the chest, abdomen, and pelvis every three months starting at the end of adjuvant chemotherapy did not show evidence of recurrence. The patient is free of disease two years later.

DISCUSSION

Wilms tumor is the most common kidney tumor in children, whereas renal cell carcinoma is most common in adults.2 Only 3% of Wilms tumors are reported in adults.
Wilms Tumor: An Uncommon Entity in the Adult Patient

Wilms tumor is primarily a sporadic disease. Only 1% to 2% of patients have a family history of Wilms tumor. Loss-of-function mutations of a number of tumor suppressor genes, including the WT1 gene located on chromosome 11p13, p53, familial WT1 and 2 (FWT1 and FWT2) genes, and at the 11p15.5 locus, are detected in patients with Wilms tumor.

More often than children, adults present with pain, weight loss, decrease in performance status, or fever; but sometimes, as with most children, they present with indolent growing renal mass. Staging is based upon the anatomic extent of the tumor and currently there are two staging systems: the National Wilms Tumor Study and the International Society of Pediatric Oncology. Wilms tumor in adults is a curable disease if managed with the multimodal therapy according to pediatric protocols, including surgery and chemotherapy with or without radiation therapy.

CONCLUSION

Wilms tumor, the most common kidney tumor in children, is rarely seen in adults. More often than children, adults present with pain, weight loss, drop in performance status, or fever; but sometimes, as with most children, they present with indolent growing renal mass. Staging is based upon the anatomic extent of the tumor and currently there are two staging systems: the National Wilms Tumor Study and the International Society of Pediatric Oncology. Wilms tumor in adults is a curable disease if managed with the multimodal therapy according to pediatric protocols, including surgery and chemotherapy with or without radiation therapy.

Figure 2. Epithelial elements within the tumor show cells with hyperchromatic nuclei and eosinophilic cytoplasm arranged in a tubular architecture (haematoxylin and eosin stain, 10x magnification).

Figure 3. Blastemal elements within the neoplasm show sheets of monomorphic, primitive-appearing cells with small, hyperchromatic nuclei and scant clear to lightly eosinophilic cytoplasm (haematoxylin and eosin stain, 10x magnification).

(> 16 years old), making it a challenging entity for diagnosis and treatment. Wilms tumor is primarily a sporadic disease. Only 1% to 2% of patients have a family history of Wilms tumor. Loss-of-function mutations of a number of tumor suppressor genes, including the WT1 gene located on chromosome 11p13, p53, familial WT1 and 2 (FWT1 and FWT2) genes, and at the 11p15.5 locus, are detected in patients with Wilms tumor. More often than children, adults present with pain, weight loss, decrease in performance status, or fever; but sometimes, as with most children, they present with indolent growing renal mass, as in this case.

Wilms tumor is frequently misdiagnosed, as was our patient 4 years earlier, as metanephric adenofibroma, one of the rarest benign renal tumors. Pathologic features of metanephric adenofibroma (6 cases) and Wilms tumor (7 cases) were reported in a case series. Six cases of metanephric adenofibroma were strongly and diffusely positive with antibodies to Wilms tumor (WT1) protein and CD57 and focally positive with antibodies to cytokeratin 7. Seven cases of Wilms tumor were strongly and diffusely positive with WT1 in the blastema and epithelium but showed only weak focal positivity in stromal cells. Moreover, 6 cases of Wilms tumor were diffusely positive and 1 case showed focal positivity for CD56.

There are 2 major systems in use to stage Wilms tumor, namely the National Wilms Tumor Study adopted in Canada and the US and the International Society of Pediatric Oncology adopted in Europe. The National Wilms Tumor Study was established in 1969 and was 1 of the first multidisciplinary cooperative groups that included oncologists, surgeons, pathologists, radiation oncologists, radiologists, epidemiologists, and statisticians with the ultimate goal of finding a cure for children with kidney cancer, with emphasis on Wilms tumor. During the course of 5 clinical trials, with the last patient enrolled in 2002, tumor mortality rates were cut in half, so that today nearly 90% of children with Wilms and other kidney tumors can expect to survive at least until their teenage years, with excellent prospects thereafter.

Dramatic improvement in overall survival has occurred during the past decade because of improved surgical techniques, effective chemotherapeutic agents, advances in radiation oncology, and improved supportive care. Adults with Wilms tumor are treated with the same risk-based protocols used in children. These risk-based protocols incorporate multimodal therapy including surgery, chemotherapy, and radiation. This approach resulted in a dramatic improvement in outcomes with 5-year overall survival approaching 90%. The histopathology of Wilms tumor in adults seems to be identical to that in children and tends to respond to the same protocols used in children. However, the rate of treatment-related toxicity, such as fatigue, nausea, vomiting, fever, pancytopenia, neuropathy, liver function test abnormalities, skin rash, allergic reaction, pneumonitis, and congestive heart failure, appears to be higher in adult patients.

CONCLUSION

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Wilms Tumor: An Uncommon Entity in the Adult Patient

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