UIJ UroToday International Journal®

www.urotodayinternationaljournal.com
Volume 5 - June 2012

Adult Wilms Tumor with Spinal Metastases

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Submitted October 24, 2011 - Accepted for Publication November 7, 2011

ABSTRACT

Wilms tumor is rare in adulthood. The prognosis is poor when compared with children. Adults more frequently present with advanced disease stages, and chemotherapy has a moderate effect. The various protocols of chemotherapy and indications for surgery and radiotherapy are not yet precisely defined, mainly due to the rarity of the disease. Here we report a case of adult Wilms tumor with spinal metastases who remains disease free for 3 years after undergoing multimodality treatment.

INTRODUCTION

Wilms tumor is the most common pediatric renal neoplasm, but its occurrence in adults is very rare. There is no morphological difference between adult Wilms tumor (AWT) and pediatric Wilms tumor (PWT), but the pathogenesis may be different [1]. The prognosis of AWT is poorer when compared to PWT, in part because adults frequently present with advanced disease stages. Clinical presentation of AWT is often indistinguishable from other more common adult renal tumors. Staging and treatment guidelines for adults are still lacking. Currently, the staging of both AWT and PWT is done in the same way, according to the National Wilms Tumor Stage Group (NWTSG) or the Society of Pediatric Oncology (SIOP) [2,3].

CASE REPORT

A 21-year-old female presented with complaints of severe low backache and radiating pain to her right lower limb over a 3-month duration. Ultrasound of the abdomen showed a 10.2 cm x 8.1 cm heterogeneous mass arising from the lower pole of her right kidney. Magnetic resonance imaging (MRI) of the abdomen and spine showed a mass lesion arising from the lower pole of the right kidney (Figure 1, Figure 2) with metastases in the lumbar spinal cord (Figure 3). CT-guided, fine-needle aspiration cytology (FNAC) showed renal cell carcinoma metastasizing to the spinal cord. A right cytoreductive nephrectomy was done, and the postoperative period was uneventful. The gross specimen showed a gray-white tumor that was 15 cm x 10 cm x 6 cm with areas of hemorrhage and necrosis. Histopathological examination of the kidney showed adult Wilms tumor (monophasic blastemal type) with neuronal differentiation (Figure 4). Immunohistochemical (IHC) markers cytokeratin, CD 117, S100, and neuron-specific enolase (NSE) were positive, and the vimentin, epithelial membrane antigen (EMA), leucocyte common antigen, synaptophysin, chromogranin, and PAS with diastase were negative. Postoperatively, the patient received chemotherapy and radiotherapy, and there was no recurrence of disease. The patient has been disease free for 3 years.

KEYWORDS: Adult Wilms Tumor, spinal metastases

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CITATION: *UroToday Int J*. 2012 June;5(3):art 24. http://dx.doi.org/10.3834/uij.1944-5784.2012.06.11

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case report

Figure 1. MRI abdomen showing a mass arising from the lower pole of the right kidney.

http://dx.doi.org/10.3834/uij.1944-5784.2012.06.11f1

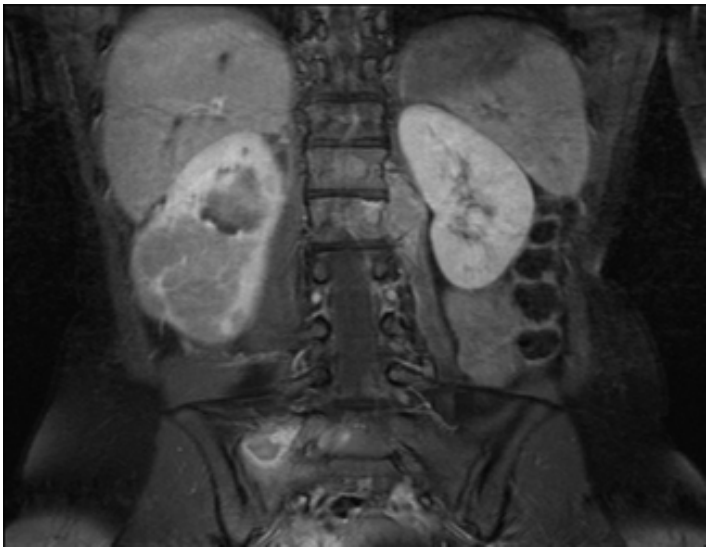

Figure 2. MRI abdomen showing a mass arising from the lower pole of the right kidney.

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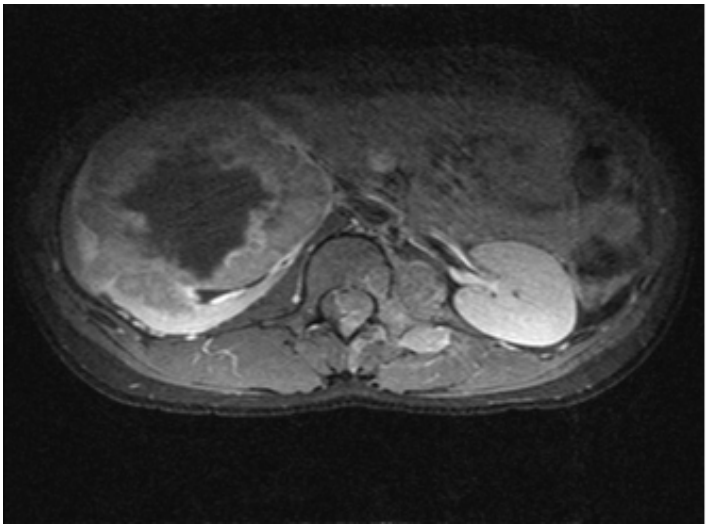

DISCUSSION

Wilms tumor is an embryonal malignancy that arises from remnants of an immature kidney. Only 3% of Wilms tumors are diagnosed in adult patients [4]. Results from the California Cancer Registry [5] showed that AWT represents 30% of primary renal cancer in the second decade of life, and that patients have a mean age of 13.9 years. Because AWT is rare, its diagnosis is infrequently suspected in adult patients. Tumor symptoms are unspecific (e.g., flank pain, abdominal mass, hematuria), and it is not possible to achieve a safe diagnosis with imaging studies alone. Imaging only confirms the presence of a renal mass. Usually, the diagnosis is established after primary nephrectomy [4]. A fine-needle or true-cut biopsy may be a plausible approach in cases of primarily inoperable tumors or metastatic disease.

Histologically, AWT does not differ from PWT. The tumors are composed of blastemal, stromal, and epithelial cells that recapitulate normal kidney development. The proportions of the 3 cell components vary greatly [2,3]. In some cases diagnosis is difficult by morphology alone, especially in adults. Diagnosis of nephroblastoma is retained after the exclusion of other disorders that are more common to this age, such as renal cell carcinoma (RCC), renal sarcoma, and primitive neuroendocrine tumors (PNET) [4,6].

Figure 3. MRI of the spine showing lumbar spinal cord metastases.

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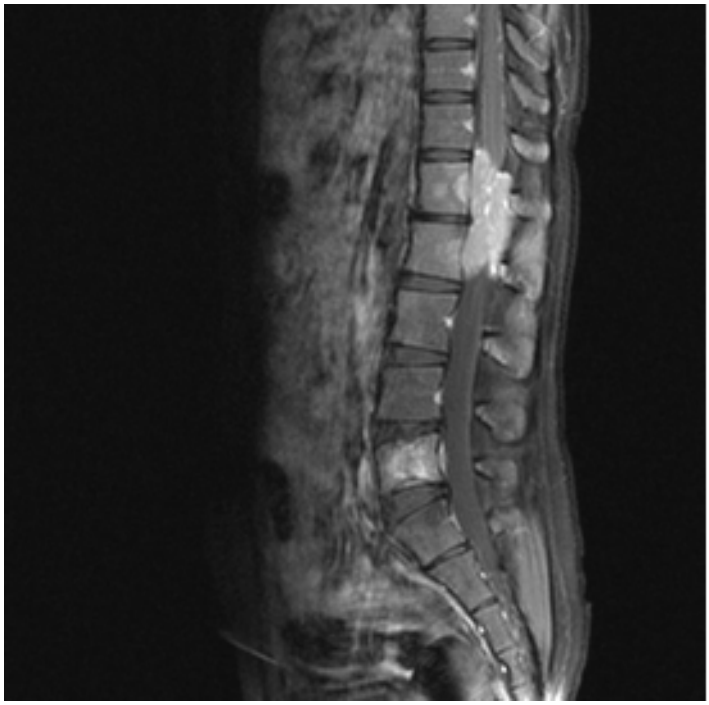

Figure 4. Histopathological examination of the kidney showing an adult Wilms tumor (monophasic blastimal type) with neuronal differentiation.
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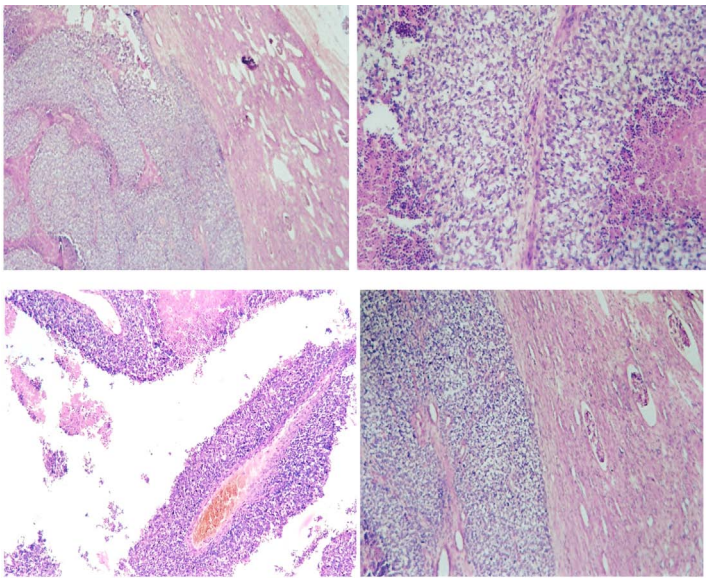

AWT often shows anaplasia, which is associated with a more aggressive clinical behavior. Patients with anaplasia have a 5-year overall survival (OS) rate of 37%; patients without anaplasia have a corresponding OS rate of 65% [7]. The prognosis of AWT is poor when compared with children. Approximately 33 to 50% of adult patients have metastatic diseases at the time of diagnosis, and adults often have weight loss or a decline in their performance status that is not typically seen in children [2,4]. Adult Wilms tumor is diagnosed based on the criteria given by Kilton, Mathews, and Cohen [8]. These include: 1) the tumor under consideration should be a primary renal neoplasm, 2) the presence of a primitive blastemal spindle or round cell component, 3) the formation of abortive or embryonal tubules or glomerular structures, 4) no area of tumor diagnostic of renal carcinoma, 5) pictorial confirmation of histology, and 6) the patient's age is greater than 15 years.

Recently, several markers have been identified as poor prognostic indicators, such as the loss of heterozygosity (LOH) at chromosomes 1p and 16q and telomerase expression level [2]. These factors should be investigated in patients with AWT. Molecular studies and gene expression profiling should be carried out to identify new prognostic factors and incorporate them into treatment decisions.

In AWT, surgical treatment has the highest priority, and even the discovery of metastatic disease should not prevent exploration or the attempted removal of the primary tumor. If the primary tumor is initially inoperable, a second-look laparotomy is worth consideration following chemotherapy [9]. Tumor debulking and removal of the primary tumor might carry an advantage over initial systemic therapy for patients with AWT, because standard chemotherapy used for patients with PWT is expected to be less effective in adults.

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