CASE REPORT

Atypical Presentation of Renal Cell Carcinoma in a Child

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Abstract

Renal cell carcinoma is a rare malignancy in the pediatric age group whereas it is a more common lesion in older adults. Wilm's tumors out number renal cell carcinomas by a ratio of 30:1 in children. There have been less than 175 cases of pediatric renal cell carcinoma reported in the English literature to date. Besides being an uncommon lesion, an unusual mode of presentation at the time of the diagnosis, and diagnostic imaging appearance of this case makes it even more interesting. The usual signs and symptoms of painless gross hematuria, flank pain, insidious fever, and infections, as well as heterogenous and ill-defined margins, were completely lacking.

Key words: Renal cell carcinoma, adenocarcinoma, pediatric renal cell carcinoma, renal malignancy.

A 9-year-old male woke up one morning with severe acute abdominal pain. He had enjoyed good health until two days before. For the preceding two days he had received antibiotics and decongestants for a common cold. His temperature was found to be 104°F. During the next few hours his pain became worse and he was taken to the emergency room. At the ER, the physician found no unusual symptoms except the fever. The abdomen was soft and without guarding but the child complained of excruciating pain in the right upper quadrant. Laboratory tests including complete blood work, urine analysis, and abdominal radiographs were unremarkable. The physician suspected acute cholecystitis and requested an ultrasound examination of the gall bladder. This showed the gall bladder to be normal, but there was a dense, round, well-defined, smooth bordered mass in the lower pole of the right kidney. The mass was surrounded by an echoless halo. (Figure 1) To confirm the nature of the vascularity and to rule out involvement of surrounding structures computed tomography (CT) was performed. This again revealed the solid, hypovascular, homogenous, regular shaped mass. (Figure 2) No vascular pedicle or lymph nodes were involved. The child underwent surgery and frozen sections showed an undetermined malignancy. A radical right nephrectomy and exploration of the surrounding area were performed. No other structures were involved. The child tolerated the procedure well and had an uneventful recovery. His pain subsided. The tumor was classified as stage I. On permanent section, it was found to be adenocarcinoma.

Discussion

Data compiled in the third National Cancer Survey indicated that renal malignancy ranks 6th in frequency among all childhood cancers, with Wilm's tumor taking the major proportion.1-4 Renal cell carcinoma comprises about 7% of all primary intrarenal cancers in persons under the age of 21 years. Raney reported on the largest series of cases (20) in 1983, collected from four oncology centers.4 Lack et al reported on 17 cases during 29 years in the Children's Hospital and Dana Farber Cancer Institute (Boston).1

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Figure 1. Longitudinal sector scan of the right kidney shows a dense, circumscribed mass, well situated within the lower pole of the right kidney. A faint halo surrounds the mass. (white arrow).

Figure 2. Transverse CT scan shows a hypodense, hypovascular, well-defined mass in the lower pole of the right kidney (black arrow). There is no extension outside the renal capsule.
The childhood renal cell carcinoma looks similar to adult types but their growth pattern, rate of growth, and prognosis are unpredictable. There is a natural tendency to metastasize to lungs, bones, and para-aortic lymph nodes.

Most of the children reported in the literature had been symptomatic for some time and had received treatments for symptoms related to the urinary tract. Also in the literature there is emphasis on poorly defined borders and heterogeneity of the lesion. At times, these cancers are confused with renal cysts. Our case was unique due to the fact that it was well-defined, smooth, and had all criteria of a benign tumor. At the time, it was considered to be a hematoma. The child had no symptoms pertaining to the urinary tract. Violent upper quadrant pain without previous illness is considered a very unusual presentation.

The treatment of choice for stage I renal cell carcinoma in children is radical nephrectomy followed by radiation therapy. Due to the rarity of the lesion and lack of prospective randomized studies, recommendation of adjuvant therapy for uncomplicated cases remains to be proven. For some advanced stages, post operative radiation therapy has been utilized; however, there is no proven curative regimen.

Post operative local radiation was suggested, but due to complete removal of the well defined mass and the absence of any sign of vascular pedicle or capsular invasion, it was decided not to initiated either radiation or chemotherapy, and to follow him closely with time interval check ups, ultrasound, and CT scans. The child completely recovered from the surgery and returned to school. At a six month follow-up visit, he was found to be in good condition with no sign of recurrence.

References