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Noninvasive botryoid extension of Wilms' tumor into the bladder

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Abstract We present the previously unreported CT appearance of a Wilms' tumor which extended down the ureter and protruded into the bladder as a botryoid mass. The tumor apparently arose from an intralobar nephrogenic rest and demonstrated local invasion into renal sinus vessels and papillae. There was no tumor invasion into the wall of the ureter or bladder, and therefore, the extension into the ureter and bladder did not upstage the tumor. This report adds to the list of differential diagnoses of a botryoid bladder mass in a child and demonstrates yet another unusual manifestation of Wilms' tumor.

Case report

A 23-month-old African-American girl was referred for evaluation of a palpable right upper-quadrant mass. Physical examination revealed a thin infant with a right upper-quadrant abdominal mass estimated to be 6 cm in size. The results of a urinalysis revealed greater than 100 RBCs and WBCs per high power field, and 3+ protein. The peripheral WBC and creatinine were normal.

Sonographic evaluation of the abdomen revealed a large heterogeneous right renal mass with associated pelvicaliectasis (not shown). Subsequent computed tomography of the abdomen revealed a heterogeneous right renal mass measuring 9 cm × 7 cm × 7 cm, and obstructing the upper pole collecting system (Fig. 1). The renal capsule appeared intact. The mass extended down the entire course of the distended right ureter and protruded into the bladder lumen in a botryoid fashion (Figs. 2–4). No lymph node enlargement or hematogenous metastases were noted.

A radical nephroureterectomy and cystotomy were performed. During surgery, the portion of the mass protruding into the bladder was easily retracted proximally into the ureter. In light of this apparent lack of invasion into the bladder wall, a cystectomy was considered to be unnecessary. The results of a pathologic evaluation by the National Wilms' Tumor Study Pathology Center revealed a Wilms' tumor of favorable histology with a predominantly blastemal composition. The tumor exhibited an invasive nature with probable invasion of vessels within the renal sinus. The tumor was almost completely confined to the pelvicalyceal system both grossly and microscopically and had apparently arisen from an intralobar nephrogenic rest in the pelvicalyceal wall. The distal 1.5 cm of botryoid tumor was firm, hemorrhagic and necrotic with extensive bacterial growth. Microscopically, the tumor extended under the epithelium of the renal papillae but was well confined within the renal capsule. There were changes of chronic submucosal inflammation in the ureter extending into the muscular wall, but the epithelium was intact without evidence of tumor invasion. Tumor protrusion into the ureter and bladder without invasion did not upstage the tumor, which was considered to be stage 2 on the basis of possible invasion of vessels in the renal sinus.

Postoperatively the patient did well and received actinomycin D and vincristine therapy. Presently she is tumor-free nine months after surgery.

Discussion

Wilms' tumor typically presents as an intrarenal mass which invades and distorts the renal parenchyma. The tumor may extend beyond the renal capsule, invade the renal sinus, metastasize to lymph nodes or spread hematogeneously to distant sites. Occasionally the tumor arises predominantly within the renal collecting system.

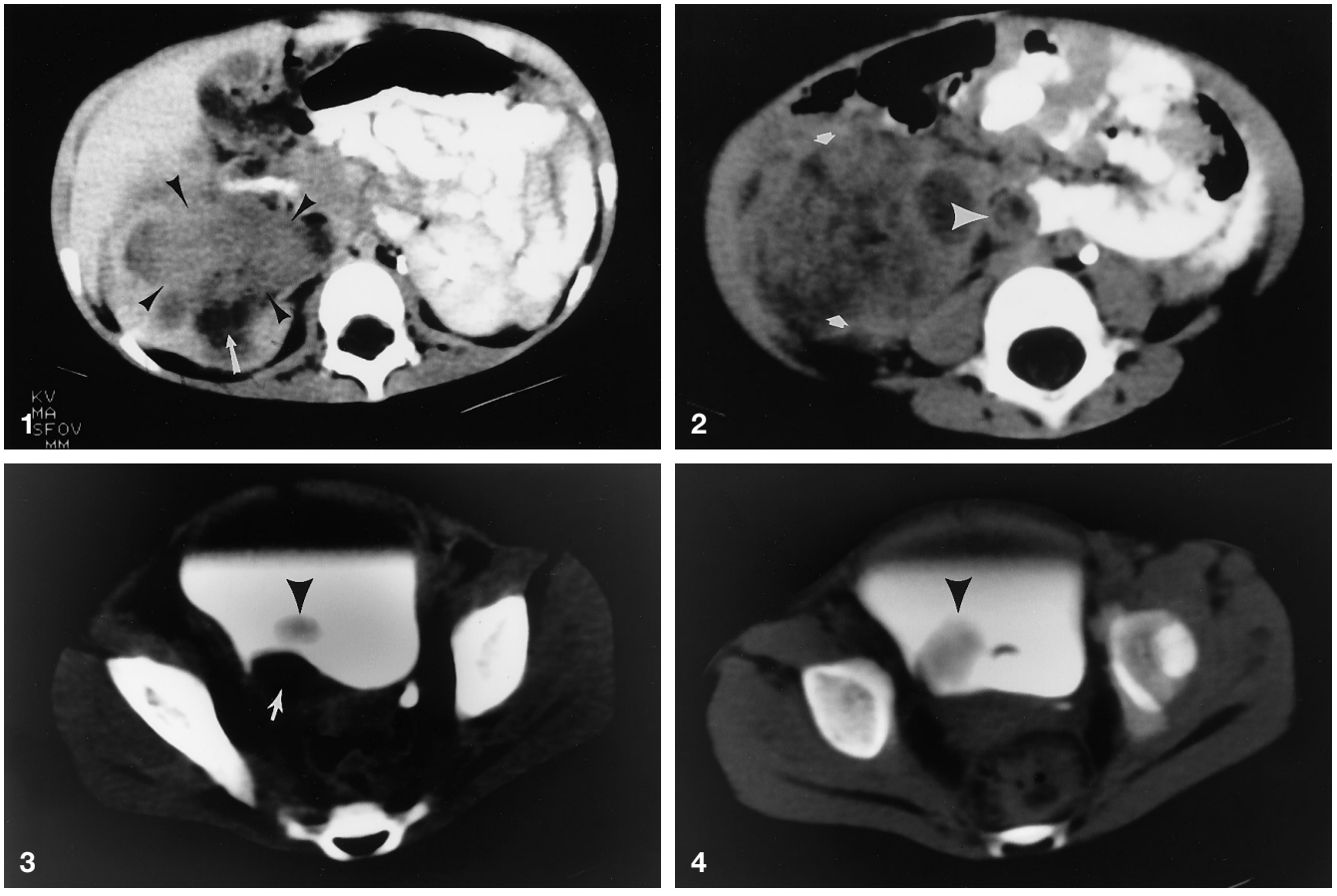

Fig. 1 Axial CT with oral and intravenous contrast. The intrapelvic Wilms' tumor *(arrowheads)* is causing dilatation of upper pole calyces *(arrow)*

Fig. 2 Axial CT with oral and intravenous contrast. The right ureter is distended with Wilms' tumor *(arrowhead)*. The inferior aspect of the intrarenal tumor mass is also seen *(arrows)*

Fig. 3 Axial CT with intravenous contrast reveals an impression on the posterior bladder made by the distended distal ureter *(arrow)*. Extension of the Wilms' tumor into the bladder is also seen *(arrowhead)*

Fig. 4 Axial CT with intravenous contrast shows botryoid extension of the Wilms' tumor into the bladder lumen *(arrowhead)*

In 1993, Niu et al. [1] described two new cases of intrapelvic Wilms' tumor. The authors also reviewed six additional cases of documented intrapelvic Wilms' tumor from the medical literature. In none of these cases did the tumor extend into the bladder. The incidence of hematuria in these cases was 88 % versus approximately 25 % in typical Wilms' tumors. There was variability in the presence of tumor invasion into the epithelium of the renal pelvis or ureter. Groeneveld et al. [2] reported a case of intrapelvic Wilms' tumor not associated with hematuria, either grossly or microscopically. These data suggest that the presence of hematuria in a child with a Wilms' tumor strongly suggests extension into the renal pelvis, but is not pathognomonic. Niu et al. [1] also reported an incidence of palpable abdominal mass in only 38 % of the cases reviewed versus an incidence of up to 94 % in the typical Wilms' tumor. Navoy et al. [3] reported a number of unusual manifestations of Wilms' tumor including multiple cases of tumor extending into the renal pelvis and proximal ureter. No mention was made concerning the presence or absence of epithelial invasion by the tumors. No cases of tumor extension into the bladder were reported.

Neither of the two new cases of intrapelvic Wilms' tumor reported by Niu et al. [1] exhibited positive urine cytology. This suggests that urine cytologic analysis is not a sensitive indicator of intrapelvic tumor extension. In our case, the extension of the tumor into the ureter and bladder were not initially appreciated on the CT scan. This was due to high attenuation contrast in the bladder obscuring the botryoid tumor. The ureteral involvement was erroneously interpreted initially as tumor invasion of the inferior vena cava. Had the extension of the tumor into the bladder been detected preoperatively, retrograde pyelography and cystoscopy may have been indicated to rule out invasion of the ure-

ter or bladder [1]. We believe the necrosis of the distal tumor in our case can be explained by the unusually long distance between the tip of the tumor in the bladder and its blood supply arising within the kidney.

There are two types of Wilms' tumor variants which have been reported to exhibit a tendency toward extension into the renal pelvis. Wigger [4] described a review of 20 fetal rhabdomyomatous nephroblastomas, three of which (15%) formed polypoid extensions into the renal pelvis. These tumors all exhibited skeletal muscle with interspersed fibrous tissue as the predominant element. Fernandes et al. [5] described three cases of teratoid Wilms' tumor, two of which (66%) extended into the renal pelvis. Teratoid Wilms' tumors exhibit a clear predominance of teratoid elements comprising greater than 50% of the tumor. The tumor which we have described contained no rhabdomyomatous or teratoid elements but rather was determined to be of predominantly blastemal composition.

It has been suggested by others [1] that Wilms' tumors which are predominantly intrapelvic arise from nephrogenic rests in the renal pelvis or perhaps from the ureteral bud itself. Beckwith [6] has classified nephrogenic rests into perilobar, intralobar, combined and panlobar. The tumor we describe likely arose from an intralobar nephrogenic rest in the pelvicalyceal wall.

As far as we know, this is the first reported case of a Wilms' tumor extending transureterally into the bladder. Interestingly, there was no invasion of the tumor into the epithelium of either the ureter or the bladder. This was in spite of the fact that the tumor was pathologically considered to exhibit an invasive nature with apparent invasion into renal sinus vessels. This report adds to the differential diagnosis of a botryoid bladder mass in a child which also includes rhabdomyosarcoma, blood clot, calculus, ureterocele and tumoral cystitis.

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