An Uncommon Variant of Wilms Tumor: Botryoid Intrapelvic Renal Mass

Telli et al. Botryoid Wilms Tumor

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11 March 2020
24 June 2020

DOI: 10.4274/balkanmedj.galenos.2020.2020.3.59

Infrequently, wilms tumor (WT) can be detected as a polypoid structure in renal pelvis without any parenchymal component and may extend to the bladder. This uncommon type of WT named as Botryoid Wilms tumor (BWT).1 We report a rare case of a 5-year-old girl, who admitted to our clinic with a chief complaint of gross hematuria and weight loss. Urinary system ultrasonography (USG) revealed grade 2 hydronephrosis in the right kidney and echogenicity (like renal stones or infected material) was observed in the renal pelvis and proximal ureter. Non-contrast computed tomography (CT) showed right enlarged inhomogeneous renal mass extending to collecting system with dilated renal pelvis and ureter. (Figure 1) Punch biopsy and pelvic cytology demonstrated malignant neoplasia after ureterorenoscopy. Right laparoscopic nephroureterectomy was performed and pathology reported macroscopically a 6 x 4 x 2 cm grayish polypoid mass growing into renal pelvis extending into the proximal ureter (Figure 2). The tumor had triphasic pattern of blastema, epithelial and stromal components with ratios of 40%, 30% and 30% verifying as Wilms’ tumor typical features. The blastema was composed of diffuse and packed primitive small blue cells with scanty cytoplasm and overlapping nuclei. The epithelial component was at the tubular formation and observed as glomeruli-like structures. The stromal component was composed of fibroblast-like stroma with the myxoid background. The tumor cell was not defined as anaplastic histology. There was triphasic tumoral tissue under the urothelial epithelium. (Figure 3A) Immunohistochemically, the epithelial tumor cells were positive for WT-1 (Figure 3B) The ureter was wide and its lumen was filled with coagulation necrosis. The tumor was separated from other kidney tumors with its triphasic appearance. Stage II WT was diagnosed after clinical, imaging, and histologic findings. Wilms tumor growing in a botryoid fashion is usually not infiltrating the pelvic wall, so intrapelvic botryoid growth should not be regarded as renal sinus/pelvic invasion and a reason for up staging a tumor.2 This kind of growth pattern may cause incorrect staging of the tumor. Therefore, it should be evaluated in detail by the clinicians to prevent incorrectly upgrading the stage of the tumor.

Moreover, wilms tumor staging tells biopsy may cause tumor spread intraperitoneally should be staged as Stage III. We performed an endoscopic biopsy (urolaroscopy) procedure that is not related with any tumor spread or any collecting system perforation that may think pathologists or oncologists as a peritoneal spread. There could be a concern however this is not like a biopsy that will cause perforation of tumor or any peritoneal implant. All procedure was performed in the lumen of collecting system.

WT is usually presented as asymptomatic abdominal mass, however the most common clinical finding in BWT is macroscopic hematuria. In addition to hematuria, dysuria, urinary tract infection, hydroureteronephrosis (secondary to obstruction) and loss of renal function (secondary to chronic obstruction) will be clinical findings.2,3 The diagnosis of BWT is quite difficult because it is rarely described in the literature and is therefore
ignored in the differential diagnosis. The malignant rhabdoid tumor of the kidney, rhabdomyosarcoma, hemangioma or xanthogranulomatous pyelonephritis should be considered in the differential diagnosis.4-5 In conclusion, clinicians should be aware of the existence of this unusual form of Wilms tumor, which originates in the pelvis and extends into the ureter or bladder with macrohematuria.

REFERENCES

FIG. 1 (A) Non-contrast CT shows right kidney consisting of inhomogeneous mass (Arrow) which filled the pelvicalyceal system. (B) The mass extends into the right renal pelvis (Arrow) and proximal ureter.
FIG. 2. Nephroureterectomy Specimen (A) Grayish polypoid mass with coagulation necrosis. (B) Botryoid tumor invades pelvicaliceal system, renal pelvis and ureter (Blue arrow= tumor, red arrow= normal kidney tissue)

FIG. 3. (A) Microscopical image of botryoid wilms tumor tumoral tissue composed of blastema, epithelial and stromal components -HEX100) (B) Immunohistochemical staining for WT1 (epithelial tumor cells were positive)