

Cystic Variant of Wilms' Tumor Presenting with Ureteral Extension

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SUMMARY

Wilms' tumor is a neoplasm of the kidney and the incidence is approximately 8 cases/million children younger than 15 year of age. The tumor is usually solid but rarely predominantly cystic and cystic variant of tumor is extremely rare. Therefore, the cystic variant can be confused with the other cystic renal diseases especially in older children. Herein, we presented an 17 month-old patient with cystic Wilms' tumor associated with ureteral extension. This case is important in terms of both the rare variant of Wilms' tumor and the presence of ureteral extension.

Key words: Kidney Diseases, Cystic, Wilms Tumor, Ureter

ÖZET

Wilms tümörü böbreğin malign bir tümördür ve insidansı 15 yaş altında milyonda 8 olgu şeklindedir. Tümör genellikle soliddir fakat nadir de olsa kistik tarzda olabilir ve tümörün bu şekli son derece nadirdir. Bu nedenle, tümörün kistik tipi özellikle büyük çocuklarda böbreğin diğer kistik hastalıkları ile karışabilir. Biz burada üreter yayılımının da olduğu 17 aylık bir hastada saptadığımız kistik Wilms tümörü olgusunu sunduk. Bu olgu, tümörün hem Wilms tümörünün nadir bir varyantı olması hem de üreteral yayılım göstermesi nedeniyle önemlidir.

Anahtar Kelimeler: Böbrek hastalıkları, kistik; Wilms Tümörü; Üreter

Introduction

Wilms' tumor, also known as nephroblastoma, is a complex mixed embryonal neoplasm of the kidney composed of three elements: blastema, epithelia and stroma (1). The tumor is usually solid but rarely predominantly cystic (2). Although invasion of renal collecting system is common, extension down into the ureter is not (3). Herein, we presented an 17 month-old patient with cystic Wilms' tumor. Initially the tumor presented itself like cystic kidney disease.

Patient

A 17-month-old boy was referred to our hospital because of intermittent clotted mass in his urine and then gross hematuria that persists 2-3 days. Ultrasound and MRI showed left cystic renal tumor measuring 9 x 7 x 10 cm, which had thick irregular septas with solid components. The mass was extending to superior exophytically. But cystic nephroma (CN) and cystic partially differentiated nephroblastoma (CPDN) distinction could not be made with MRI studies. Renal trucut biopsy was performed for diagnostic purpose but limited amount of tumor tissue has been obtained, probably due to cystic component, so distinction between NB, CN and CPDN has not been made properly. For this reason, chemotherapy was not implemented preoperatively. Radical nephroureterectomy was performed.

Macroscopic examination of specimen showed a rounded and sharply demarcated tumoral mass with 11 x 10 x 9 cm in size. It was mostly cystic and contains limited solid areas. Lumen was filled with tumoral tissue in the proximal part of the ureter. Microscopically, the solid areas of the tumor showed triphasic pattern, composed of blastemal, epithelial (tubular structures) and differentiated stromal elements (Figure 1),(Figure 2) . Renal calyces were infiltrated by tumor. Ureteral opening filled with necrotic tumoral extension but ureteral invasion has not been demonstrated (Figure 3) . Tumor was diagnosed as cystic Wilms' tumor histopathologically.

Thorax computerized tomography studies were normal. The diagnosis was stage 2 cystic Wilms' tumor with favorable histology. Postoperative chemotherapy was started with vincristine and dactinomisin for stage 2 diseases.

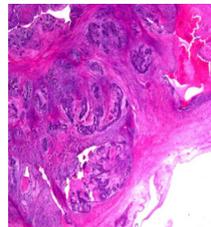


Figure 1: Nephroblastoma with cystic areas containing blastemal, epithelial and stromal elements (H&E, X40)

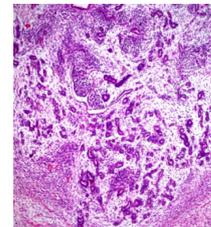


Figure 2: Epithelial elements in tubular forms and packing of blastemal cell groups (H&E, X100)

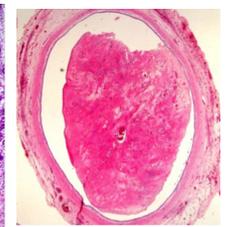


Figure 3: Necrotic tumor extension into the ureteral opening (H&E, X40)

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Discussion

Cystic renal tumors of childhood are relatively rare and are believed to be part of spectrum with CN at the benign end, CPDN in the intermediate region and cystic Wilms' tumor (CWT) at the malignant end (4). The discrimination of these three entities has to be based on histology, as radiological findings are often inconclusive. In addition, all three entities are very similar in their gross appearance and do not have any discernible difference in clinical presentation. The only discriminating clinical factor is age at presentation as most patients with CN and CPDN present before one year of age, whereas patients of Wilms' tumor are often older (5). CN and CPDN are thought as benign lesions, manageable with surgery alone, not requiring preoperative chemotherapy but CWT requires a multidisciplinary approach.

Pathologically, CN consist of cysts and septa with cysts lined by flattened, cuboidal or hobnail epithelium and septa that may be composed of fibrous tissue containing mature tubules but no immature elements. Immature mesenchymal / epithelial tissue or blastemal cells are found in the septa of the lesions classified as CPDN. CWT contains more solid structures between the cysts which bulge into the cystic spaces and also contain blastemal / mesenchymal / epithelial elements (6).

Tumor extension into the ureter is an unusual finding in Wilms tumor (3). Reports of intrapelvic extension are more frequent but progress down the ureter or into the lower urinary tract is uncommon (7). Clues to the presence of ureteral involvement include presentation with gross hematuria like our patient and the finding of hydronephrosis or nonfunction of the kidney on preoperative studies (3). But we could not detect hydronephrosis on ultrasound and did not make renal function studies such as nuclear imaging studies.

Some reports in the literature have suggested that tumors with ureteral extension are more resistant to therapy (8-10). But the rightness of this view could not be proven (3).

Conclusion

Ureteral extension of Wilms' tumor is rare. None of Wilms tumor patients that reported ureteral extension, ureteral invasion has not been found like our patient (3,7,11). The case reported by Stevens and Eckstein may be an exception (8). In addition, Wilms tumor should be considered in the differential diagnosis of multilocular cystic lesions of the kidney especially in older children.

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