Case report

Botryoid Wilms' tumor: report of two cases

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Background: **Botryoid Wilms' tumor is a rare kind of Wilms' tumor. We report two cases of this tumor.**

Methods: **Case 1, a 2-year-old boy, was admitted with macrohematuria for 5 months. Case 2, a 19-month-old boy, was referred for a palpable abdominal mass. The two cases were checked by 64-row multi-slice spiral CT (MSCT) and scanned with the dynamic contrast enhancement. The masses were excised and pathologically confirmed.**

Results: **In case 1, the mass occurred in the renal pelvis and calyx bilaterally, with heterogeneous density and prominent calcification. By contrast enhanced CT scan, the mass was mildly enhanced. In case 2, the left renal pelvis and ureter were filled with the tumor. Unenhanced scan revealed that the mass was homogeneous and non-calcified. In contrast, the mass was slightly and heterogeneously enhanced. Macroscopically, the mass filled in the pelvicalyceal system and had a botryoid appearance. Microscopically, the typical features of Wilms' tumor with blastemal, epithelial, and stromal components were evident.**

Conclusion: **Botryoid Wilms' tumor should be included in the differential diagnosis of tumors in the pelvicalyceal system no matter it is unilateral or bilateral.**

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Key words: **botryoid Wilms' tumor; computed tomography; enhancement**

Introduction

Wilms' tumor is the most common renal malignancy in childhood. It usually occurs as a mass in the renal parenchyma and expands into the surrounding tissues.[1-3] It rarely arises from the renal pelvicaliceal wall, occupying the collecting system with minimal parenchymal involvement. Its appearance is similar to that of botryoid sarcoma, so called botryoid Wilms' tumor.[2-5] Herein we report two cases of this tumor.

Case report

Case 1: A 2-year-old boy presented with macrohematuria for 5 months. Blood analysis showed white blood cell count of 11.6×10^9/L, red blood cell count 3.61×10^9/L, and hemoglobin level 93 g/L. Urinalysis showed numerous red blood cells per high-power field. Ultrasonography revealed two mixed echogenicity masses with calcification occupying the bilateral renal pelvis respectively, and the kidneys were obviously enlarged. Abdominal plane film showed multiple calcification in both kidneys (Fig. 1). Pre-contrast computed tomography (CT) showed enlarged kidneys bilaterally with heterogeneous masses, which filled the pelvicalyceal system. A lot of calcification was seen within the masses (Fig. 2A). Contrast enhanced CT scan revealed that the masses were enhanced slightly and inhomogeneously. The renal parenchyma was thin and enhanced moderately (Fig. 2B). The ureters and bladder were not involved, and no lymph metastasis was found. Nephron-sparing surgery was performed. The mass in the right kidney was first enucleated after opening the kidney via a transabdominal technique. The left mass was enucleated about one month later. During the operation, two polyploidy (botryoid) and pinkish masses with capsule were found, occupying the bilateral renal calyx and pelvis respectively. The pedicle of the tumor was attached to the renal parenchyma. No mass was seen in the renal parenchyma macroscopically. Microscopically, the mass consisted of stromal, epithelial and blastemal components as Wilms' tumor (Fig. 3). The intralobar nephrogenic rests were not found in the renal parenchyma of the removed kidneys. WT1 mutations were negative. After the operation, the boy received radiotherapy and chemotherapy but died from renal failure half a year later.

Case 2: A 19-month-old boy experienced abdominal distension for one month. Physical examination found a palpable mass, which was firm

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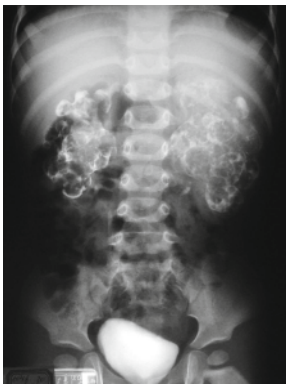

Fig. 1. Case 1: a 2-year-old boy with macrohematuria. Plain X-ray film showing multiple calcifications in the bilateral renal area. The shadow of the kidneys was enlarged.

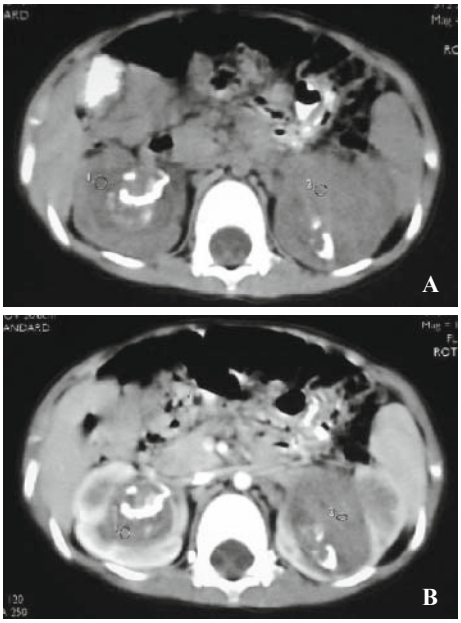

Fig. 2. Case 1: a 2-year-old boy. **A:** Non-contrasted CT showing enlarged kidneys with heterogeneous masses, which filled the pelvicalyceal system bilaterally. Calcifications were seen within the masses. **B:** Contrasted enhanced CT at the same level revealed that the masses were enhanced slightly and inhomogeneously.

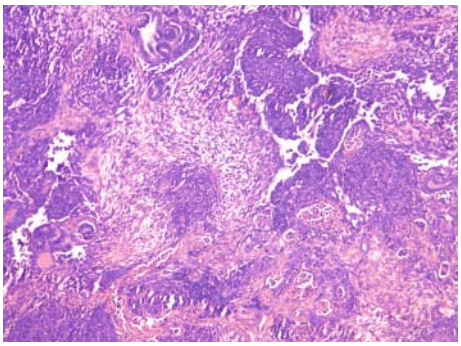

Fig. 3. Case 1: a 2-year-old boy. Microscopically, the typical features of Wilms' tumor of favorable histology with evident blastemal, stromal, and epithelial components.

and unmovable; however, there was no associated tenderness. Urinalysis showed numerous red blood cells per high-power field. The hemoglobin level was 104 g/L. Unenhanced CT scan revealed a left renal mass occupying the left renal pelvis (Fig. 4A). Its density was homogeneous and lower than that of the renal parenchyma. No calcification was observed. Enhanced CT scan revealed that the mass was slightly and heterogeneously enhanced. The left renal parenchyma was enhanced not as prominent as the right one (Fig. 4B). The interface of the mass with the renal parenchyma was obvious but no cut-off line was seen. Multi-planar reformation reconstruction manifested the mass extending to the left ureter, which was obviously outstretched and descended along the ureter close to the bladder. Radical nephroureterectomy was performed via a transabdominal technique. During the operation, the mass with a complete capsule was found to be originated from the left kidney. Macroscopically, it arose from the edge of the left renal parenchyma close to the pelvis and no mass was seen in the parenchyma. However, its major portion was located in the renal collecting system. The mass protruded into the left ureter and descended to the lower ureter, close to the bladder. Microscopically, the tumor was composed of epithelial, stromal, and blastemal components (Fig. 5). There was no evidence of intralobar nephrogenic rests in the renal parenchyma of the removed kidneys. WT1 mutations were also negative. The boy received radiotherapy and chemotherapy following the operation and is still alive.

Discussion

To our knowledge, about 17 cases of botryoid Wilms' tumor have been reported in the English literature, but very few focusing on the CT manifestations of the tumor.[5] Previous studies found that botryoid Wilms' tumor occurred in unilateral kidney.[1-8] In our study, the masses occurred in the renal pelvis and calyx bilaterally in case 1. Inoue et al[1] reported that multiple calcifications can be seen in the tumor as in our case 1. But no calcification was observed in the tumor in case 2 of our study.

The mass is located in the renal collecting system or the underlying parenchyma.[1,2] In our two cases, the renal parenchyma was shown around the masses by CT. The border between the lesion and the renal parenchyma was not clear on the unenhanced scan, but it was obvious on the enhanced scan. The density of renal parenchyma was evidently increased following enhancement, which was homogenous but lower than that of the normal kidney. The outer margin of renal parenchyma was velvety, and its edge near the tumor in the pelvis could be observed

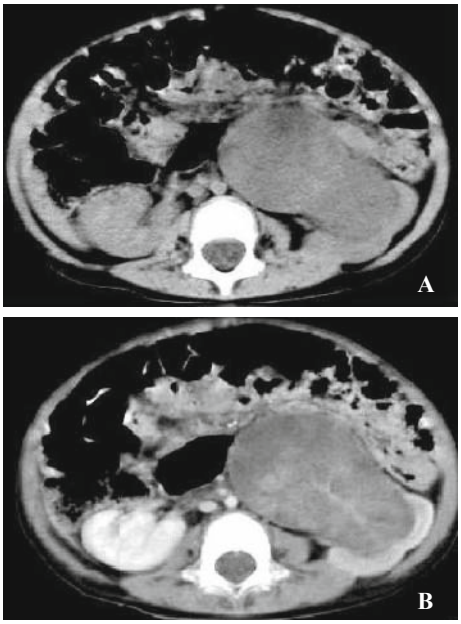

Fig. 4. Case 2: a 19-month-old boy with a palpable abdominal mass. **A:** Non-contrasted CT showing a homogeneous, hypodensity mass, whose margin appeared to be distinguishable but not clear cut; **B:** Contrasted enhanced CT showing that the mass was enhanced mildly and inhomogeneously with a clear margin.

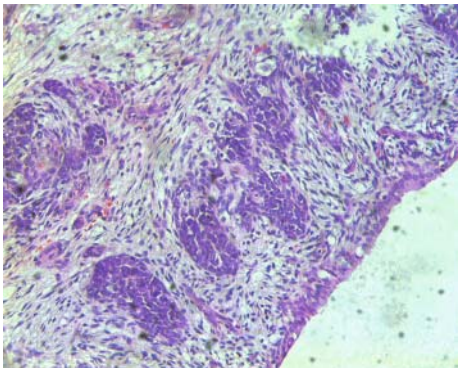

Fig. 5. Case 2: a 19-month-old boy. Microscopically, the typical features of Wilms' tumor of favorable histology with evident blastemal, stromal, and epithelial components.

but not separable from the tumor.

The mass shows no or slight enhancement on the contrasted scan.[1-4] The density of the lesion was similar to that of the renal parenchyma (CT value approximately 16-29 HU). Enhanced CT revealed that the lesion presented with mild or moderate enhancement (41-67 HU). The enhancement was frequently observed in the central region of the lesion. Previous reports found that botryoid Wilms' tumor can extend down the ureter into the bladder.[3,5] In our case 2, the mass extended into the left dilated ureter adjacent to the bladder. Before contrast, the density of the mass in the left ureter was approximately 30 HU. After contrast, however, the density of its upper part was enhanced for approximately 61 HU. But there was no enhancement in the lower part of the lesion in the ureter.

The clinical presentations of the two cases included the occurrence of a palpable mass in the right or left flank, and grossly observed blood in the urine. When a palpable mass is detected in the flank with gross or microscopic occurrence of blood in the urine, and CT or magnetic resonance imaging reveals that the major portion of the lesion is located in the renal pelvis or the collecting system, botryoid Wilms' tumor should be included in the differential diagnosis no matter it is unilateral or bilateral.

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