Radiology Case Reports 16 (2021) 166–170

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journal homepage: www.elsevier.com/locate/radcr

Case report

Bilateral Botryoid Wilms Tumor presented with acute renal failure

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ARTICLE INFO

Article history:
Received 21 September 2020
Revised 25 October 2020
Accepted 26 October 2020
Available online 13 November 2020

Keywords:
Wilms tumor
Botryoid Wilms tumor
Acute renal failure
Nephron-sparing surgery

ABSTRACT

Botryoid Wilms tumors are intrapelvic polypoid masses that are uncommon feature of Wilms tumor.

The authors reported a rare case of bilateral botryoid Wilms tumor in a 19-month-old boy who admitted to hospital with acute renal failure secondary to hydronephrosis caused by bilateral botryoid Wilms tumor with ureteral extension. The patient was successfully treated with chemotherapy and bilateral nephron sparing surgery in 2 separate operations. During 15 months follow-up, no evidence of recurrence or metastasis was found.

Introduction

Wilms tumor is the most common pediatric renal tumor accounting for about 90 % of all renal neoplasm up to 15 years of age [1]. Wilms tumor typically originates from renal parenchyma and usually is localized to parenchyma with sparing the pyelocalyceal system. However, uncommonly, it arises from renal pyelocalyceal wall and filling the collecting system with no or minimal parenchymal involvement [1–3].

Since this unusual growth feature has a similar gross appearance to botryoid sarcoma, the Wilms tumor showing this growing pattern called botryoid Wilms tumor [1–2]. Wilms tumor initially presented with Acute renal failure is extremely rare and in the literatures, there is only 2 cases are reported [4]. Herein, we described a rare case of bilateral botryoid Wilms tumor with acute renal failure presentation that successfully treated with bilateral nephron sparing surgery and chemotherapy.

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https://doi.org/10.1016/j.radcr.2020.10.055

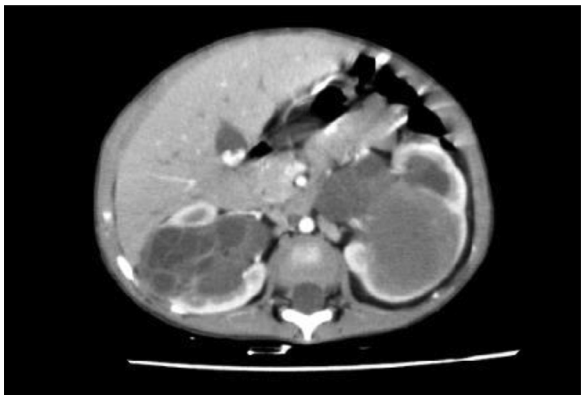


Fig. 1 – After neoadjuvant chemotherapy, Contrast enhanced CT scan (CECT scan) showed bilateral heterogeneous enhancing masses occupying pyelocalyceal systems with extension into proximal ureters and secondary hydronephrosis.

Case presentation

A previously healthy 19-month- old boy was admitted to our hospital due to vomiting, oliguria, edema, and severe weakness. The patient's initial laboratory tests demonstrated renal failure (Cr: 6/1 mg/dl, BUN: 150 mg/dl, and K: 8.2 mEq/dL) and anemia (Hb: 8/5 g/dl). Urinalysis showed culture negative Pyuria and hematuria. Ultrasound examination revealed bilateral renal enlargement (length of right kidney: 116 mm and left kidney: 98 mm). In both side, heterogeneous hypoechoic masses occupying nearly entire of pyelocalyceal systems with extension into proximal of ureters and caused moderate hydronephrosis were seen. Despite of parenchymal thinning, there was no evidence of obvious intraparenchymal mass in Right kidney but in left side, a 57*41 mm homogeneous intraparenchymal solid mass with intrapelvic extension was also found. Color Doppler ultrasound showed mild vascularity of intrapelvic and intraparenchymal masses, IVC, and both

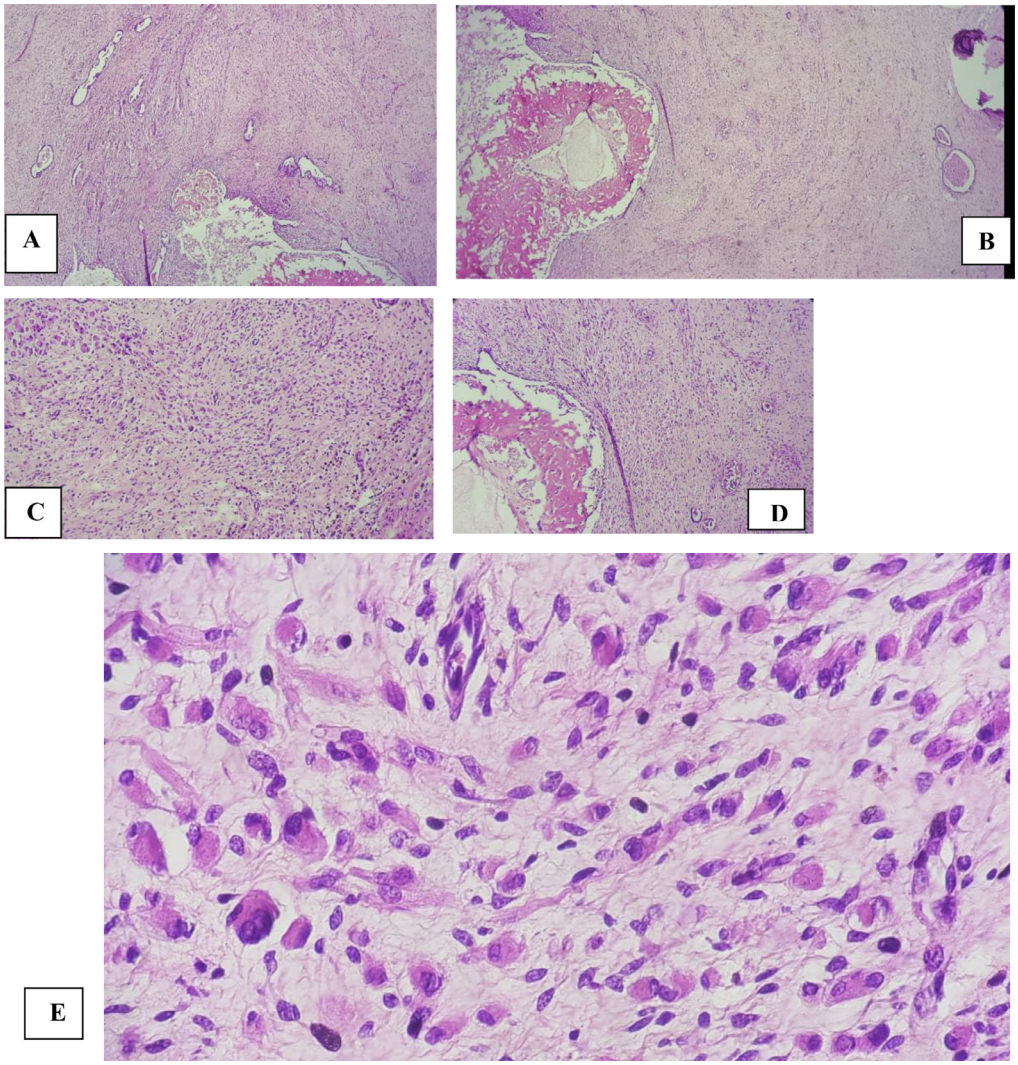
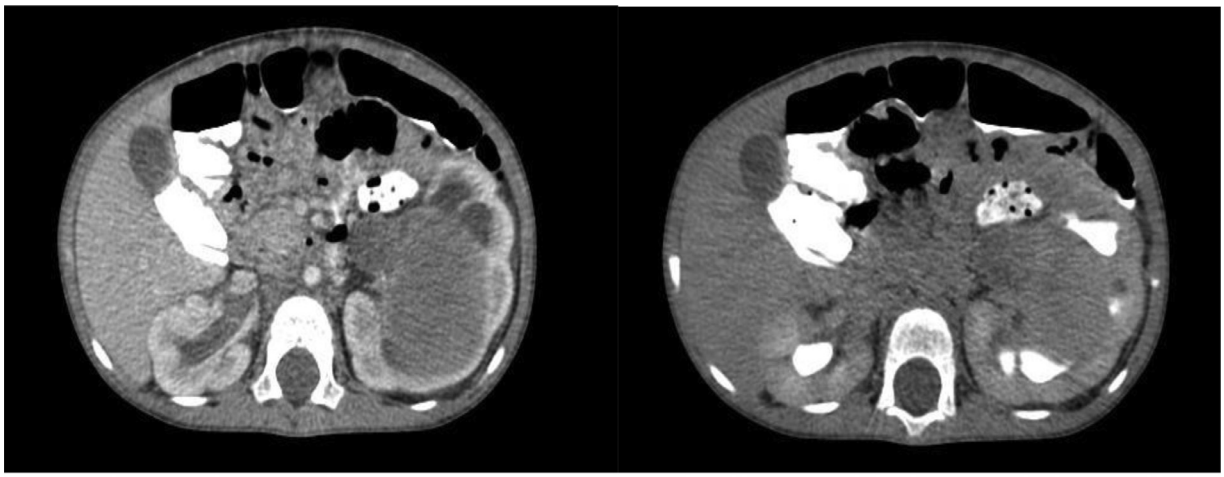


Fig. 2 – A- Wilms tumor two component seen here(epithelial and stromal), B- Myxoid stromal component contain heterologous elements such as rhabdomyblast and bony tissue, C- Rhabdomyoblast of stromal component (10x magnification), D- Bony element (10x magnification), E- Rhabdomyoblast of stromal component(40x magnification).



a b

Fig. 3 – CECT scan showed mild hydronephrosis in right kidney without evidence of tumor residue and heterogeneous intrapyelocalyceal system mass in left kidney with secondary hydronephrosis in favor of Tumor residue.

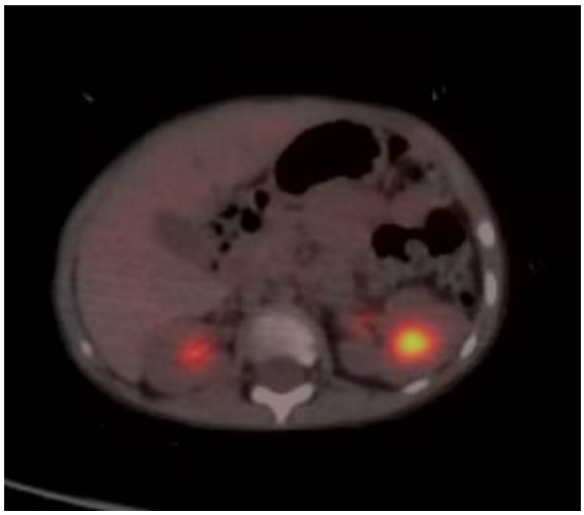


Fig. 4 – 18-F FDG PET-CT scan showed hypermetabolic mass in midpole of left kidney.

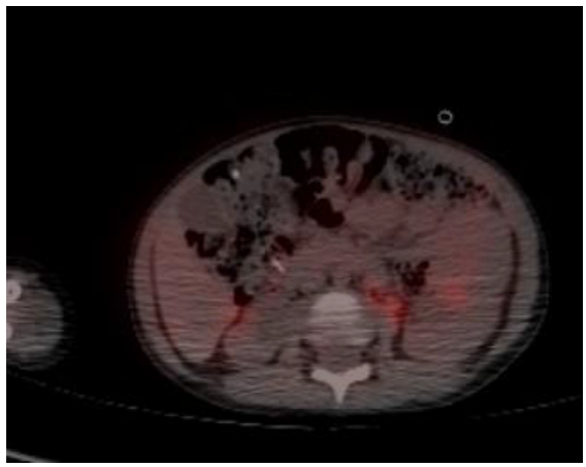


Fig. 5 – Follow up 18-F FDG PET-CT scan revealed no hypermetabolic lesion in left kidney.

renal veins had normal appearance. No evidence of liver metastasis and pararaortic lymphadenopathies was seen. Mild echo-free ascites was also present.

Regarding to ultrasound finding, a diagnosis of bilateral botryoid wilms tumor was suspected and after peritoneal dialysis catheter placement for treatment of acute renal failure, the child was put on neoadjuvant chemotherapy consistent of National Wilms Tumor Study 5 for previously untreated stage II-IV diffuse anaplastic type.

After 6 cycles of chemotherapy, partial reduction in the size of both kidneys and renal masses respect to initial imaging was seen on ultrasound and abdominal CT scan (Fig. 1). The patient underwent right kidney nephron-sparing surgery and then was continued the mentioned adjuvant chemotherapy. During surgery, polypoild masses were found filling nearly entire of right pyelocalyceal system that extended to proximal of Right ureter without obvious parenchymal mass or ureteral wall infiltration.

The masses of right kidney were successfully enucleated. The histopathologic examination showed triphasic Wilms tumor with favorable histology (Fig. 2). In follow up imaging, right kidney had mild hydronephrosis without evidence of tumoral mass and left kidney showed mildly heterogeneous intrapelvic and intraparenchymal masses with moderate hydronephrosis (Fig. 3). Three months after first operation, the child underwent left kidney nephron sparing surgery. The histopathologic examination demonstrated biphasic wilms tumor with involvement renal cortex and the renal capsule was intact. Ultrasound and after that CT scan revealed a well-defined heterogeneous mass approximately measuring 25*15

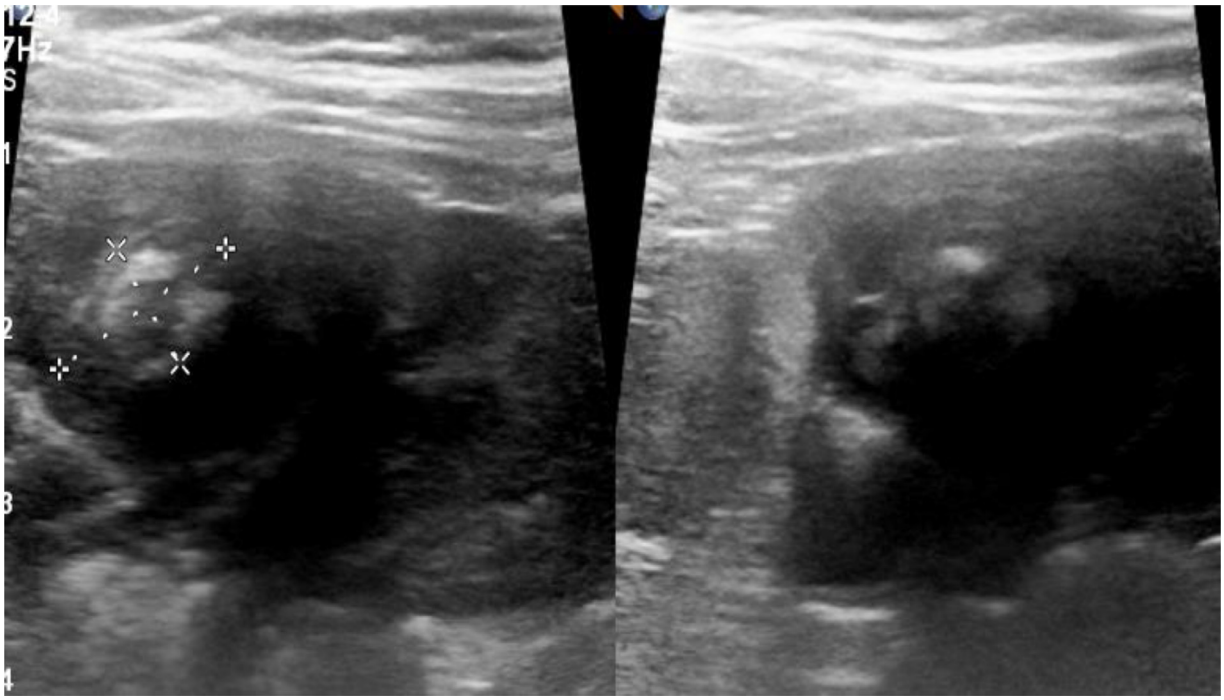


Fig. 6 – Follow up ultrasound showed predominantly calcified solid mass in midpole of left kidney.

mm with vascularity on color Doppler and mild enhancement on CT scan in midpole of left kidney.

With diagnosis of residual tumor, 2 courses postoperative chemotherapy was done with using ICE regimen. However, thereafter, a residual anatomic mass was still seen. For differentiation, residual tumor mass from nonfunctional anatomic mass, 18-F FDG PET-CT scan was done that showed hypermetabolic mass in midpole of left kidney (Fig. 4). After that, chemotherapy was continued and four months later, despite no obvious change in size of renal mass on ultrasound, the 18-F FDG PET-CT scan revealed no hypermetabolic lesion in renal kidney (Fig. 5). During the 15 month follow up, the left kidney mass is calcified and its size is stable on ultrasound (Fig. 6), the patient is healthy and no evidence of tumor residue or metastasis are found.

Discussion

The botryoid is a descriptive term used for grape-like lesions with polypoid growth pattern into the lumen [1].

Botryoid Wilms tumor used for Wilms tumor with exclusively or predominantly intrapelvic polypoid growth pattern and is not a separate type of Wilms tumor for pathological view [1–2].

For the first time in 1968, Harberg et al. [6] used the term Botryoid for the intrapelvic growing renal tumor and in 1981 Mahoney et al. [7] introduced a term of botryoid Wilms tumor for Wilms tumor showing this growth feature.

Yani et al. and Ordonez et al. [8,9] assumed that botryoid Wilms tumor originates from intralobar nephrogenic rests situated in the wall of pyelocalyceal system that extends into collecting system.

Due to the intrapelvic location of botryoid Wilms tumor, the clinical manifestation of botryoid Wilms tumor are different from typical Wilms tumor. In contrast to typical Wilms tumor, that an asymptomatic abdominal mass is the most common initial presentation with other symptoms happened in < 25% of cases, in most cases of botryoid Wilms tumors, gross hematuria is the most common presenting symptom and due to tendency of botryoid Wilms tumor for ureteral extension, hydronephrosis with or without urinary tract infection is an important manifestation of these tumors [10–11].

In review of literatures, only 5 cases of bilateral botryoid Wilms tumor have been reported and only one of them presenting with acute renal failure [1,4,5,12,13].

Indeed, renal insufficiency is a considerable complication of Wilms tumor treatment in the 5 % with bilateral Wilms tumor, but renal failure as an initial presentation of Wilms tumor is extremely rare [14].

Management of bilateral Wilms tumor is challenging because eradication of neoplasm is critical and on the other hand, preservation of maximal functional renal tissue to avoiding renal insufficiency is important. To achievement these goals, Chemotherapy and nephron sparing surgery provide a safe and effective mean [14].

Therefore, our report is extremely unique case since it is bilateral botyroid Wilms tumor that is presented with ARF and despite of obvious extension of tumor, successfully treated with bilateral tumorectomy associated with chemotherapy.

Conclusion

Despite of rarity of botyroid Wilms tumor, it must be considered in differential diagnosis of pediatric intrapelvic mass and we suggested that nephron sparing surgery could be considered even in cases with extensive Wilms tumor.

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