Urology Case Reports 25 (2019) 100898

Contents lists available at ScienceDirect

Urology Case Reports

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

Pediatrics

Rare case of primary bladder Wilm's tumor in a 1-year old boy

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A B S T R A C T

Extrarenal Wilms' tumor (EWRT) is a rare entity, but primary bladder Wilm's tumor is even rarer with only 1 case reported. A 1-year old boy came with chronic urinary retention. Abdominal pelvic CT scan revealed intravesical mass arising from anterior bladder wall extending to the prostate and bladder neck. Initial cystoscopic diagnosis revealed chronic granuloma. We decided to perform partial cystectomy with final pathologic result of bladder Wilms' tumor. EWRT may occur in various organs, but primary bladder Wilms' tumor is extremely rare case.

Introduction

Nephroblastoma or Wilms' tumor is one of the most common kidney malignancy in pediatric population. Wilms' tumor could arise not only in the kidneys, but also in different site in the body, even though it is a rare entity. Extrarenal Wilms' tumor most commonly occur in retroperitoneum, pelvic or inguinal region. In previous reports, bladder wilm's tumor were commonly an extension from renal pelvis or ureter to the bladder, while primary bladder Wilms' tumor is extremely rare, with only 1 case reported previously by Zhang et al.[1] This case provides new possibility of tumor origin among the rare case of pediatric bladder cancer.

Case presentation

A 1-year-old boy was brought to hospital by his parents with voiding difficulty since 3 months prior to admission. The complaints accompanied with intermittent fever since 1 month before, there is no history of reddish urine or sign of renal insufficiency. On physical examination, the bladder was fully distended with initial urine about 100 ml after foley catheter insertion, yellowish.

Biochemical and haematological laboratory findings were within normal limit, while urinalysis revealed microscopic hematuria. Imaging by ultrasound and CT scan revealed dilatation of in left pelvocalyceal system, and mass inside the bladder from anterior bladder wall extending to the prostate and bladder neck (Fig. 1). We performed diagnostic cystoscopy and mass biopsy to determine the origin and pathology of the mass. From cystoscopic finding, the bladder mass was arising only from anterior bladder wall, extending to the prostate and the bladder neck with pathology result of chronic granuloma. Due to discrepancy between the clinical condition and histopathologic finding and favorable location of the tumor at anterior wall of the bladder, we decided to performed partial cystectomy.

Intraoperatively we found a 3 × 2 cm pedunculated mass from anterior wall of the bladder extending to the bladder neck and obstructing the left ureteral opening. There was no sign of abdominal wall infiltration or ureteral orifices obstruction. The specimen consisted of a large gray-yellow tissue, encapsulated by a pseudocapsule. The inner mass was tender, flesh-fish-like and grayish-brown in color. The final pathologic diagnosis from partial cystectomy specimen showed composition of sheets, which were randomly arranged and tightly packed. Small blue cells were arranged in serpiginous aggregates (blastemal component) and primitive neuroepithelial cells between fibromyxoid tissues (Fig. 2). In the specimen there were epithelial component, blastemal and stromal that consist of fibromyxoid tissue. There were cells enlarge nucleus, pleomorphic and rough chromatin, eosinophilic cytoplasm. Nucleus to cytoplasmic ratio increased and mitosis feature were abundant (Fig. 3). Pathological features suggested extrarenal Wilms' tumor without lymphovascular infiltration and favorable histopathology with free margin tumor. Postoperatively the patient had vesicocutaneous fistulae that developed at second post operation day, but require no further surgical management. The fistula resolved spontaneously at 10th post operation day with just maintaining a larger foley catheter.

Discussion

Nephroblastoma or Wilms' tumor is one of the most common

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https://doi.org/10.1016/j.eucr.2019.100898
Received 10 March 2019; Received in revised form 13 April 2019; Accepted 24 April 2019
Available online 26 April 2019

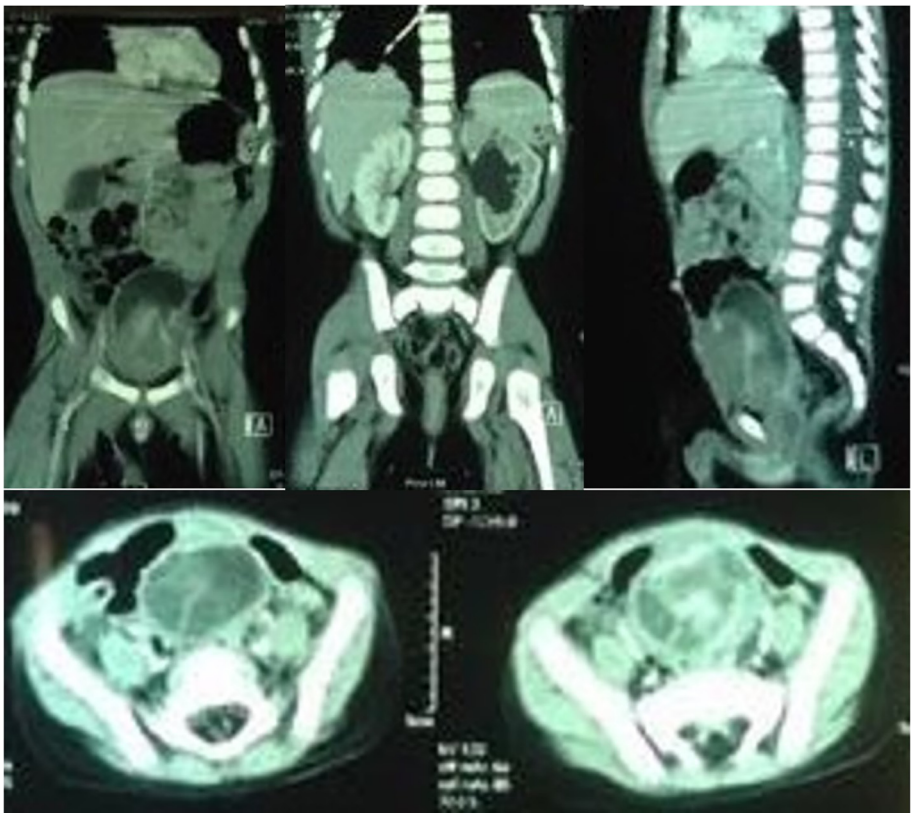

Fig. 1. Abdominal-pelvic CT scan revealed dilatation of in left pelvocalyceal system, and mass inside the bladder from anterior bladder wall extending to the prostate and bladder neck.

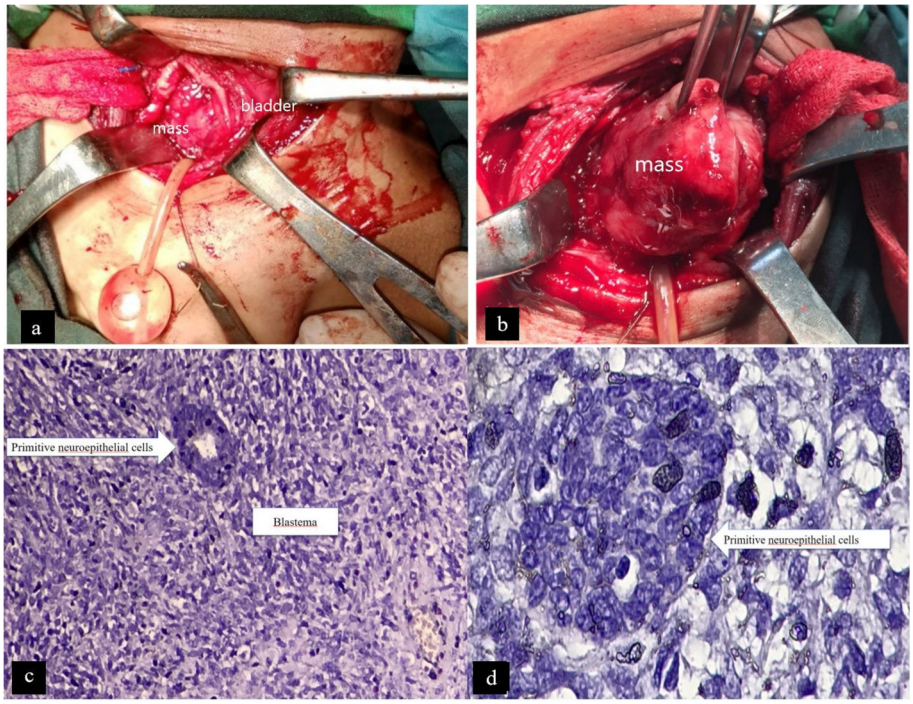

Fig. 2. (a) and (b) intraoperative findings of partial cystectomy, (c) Histopathologic finding revealed Small blue cells were arranged in serpiginous aggregates (blastemal component) and primitive neuroepithelial cells between fibromyxoid tissues. (d) Primitive neuroepithelial cells.

childhood malignancies, which accounts for almost 95% of renal malignancies in pediatrics. Extrarenal nephroblastoma is a rare entity, which was first described by Moyson et al. The estimated rate of occurrence of nephroblastoma outside the kidneys is almost 0.5–1% of all cases of Wilms' tumor. Primary bladder Wilms' tumor is very rare. In a review report by Shojaeian et al. from 1961 to 2015, there were 80 cases of ERWTs, but none of them occur in bladder.[2] Previous reports about bladder wilms' tumor described cases of bladder extension

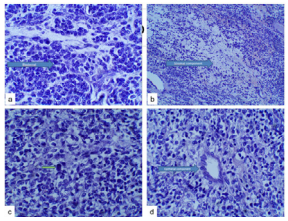

Fig. 3. a. blastemal component (100x with HE); b. stromal component (40x, with HE); c. Enlarged hyperchromatic nucleus; d. Epithelial component (40x, HE).

through the ureter from primary Wilms' tumor in the kidney (Botyroid Wilms' tumor).[3] To our knowledge, this is the second reported case of primary Wilms' tumor occurring in the bladder. Previous case was reported by Zhang et al., where they encountered a case of simultaneous Wilms' tumor in the bladder and left kidney.[1]

In this case, classical clinical picture of bladder malignancy (visible hematuria) was not found. Instead, the initial clinical picture resembles chronic obstruction in pediatric. The diagnosis of bladder neoplasm is made by from ultrasound and abdominal pelvic CT scan. Cystoscopy and histological evaluation of resected tissue revealed a chronic granuloma, a benign lesion that not resemble the clinical picture of the patient. The final histopathology result of extrarenal Wilms' tumor from partial cystectomy specimen was different from initial pathology from bladder resection. The pathologic result from first resection could be misinterpreted since lack of tissue resected or shallow resected area, since wilms' tumor originated from mesenchymal tissue, not epithelial tissue. Wilms' tumor is believed to develop from residual immature kidney cells: pluripotent embryonic renal precursor cells or metanephric blastemal.[4]

The consideration for performing partial cystectomy was due to the discrepancy between pathologic result and clinical manifestation. Considering the high suspicion of malignant bladder mass from CT scan we convinced that transurethral resection and conservative management was not enough for this patient. Favorable tumor location at anterior wall of the bladder made us take our step to performed partial cystectomy. Surgical excision remains the key step in the treatment of ERWT, especially when performed radically, but the role of intraoperative frozen section in an unidentified childhood bladder mass or ERWTs has not been discussed clearly before and is not considered as a part of surgical principle, while total excision is the mainstay of treatment in most pediatric solid tumors when applicable.[5]

Source of support

None.

References

1. Zhang DY, Lin T, Wei GH, et al. A rare case of simultaneous occurrence of Wilms' tumor in the left kidney and the bladder. *Pediatr Surg Int.* 2010;26(3):319–322.
2. Shojaeian R, Hiradfar M, Sharifabad PS, Zabolinejad N. *Wilms Tumor. Ch 6: Wilms' Tumor: Challenges in Diagnosis, Embryology, Treatment and Prognosis.* Brisbane (AU): Codon Publications; 2016.
3. Ritchey M, Daley S, Shamberger RC, et al. Ureteral extension in wilms tumor: a report from the national wilms tumor study group (NWTSG). *J Pediatr Surg.* 2008 Sep;43(9):1625–1629.
4. Hennigar RA, O'Shea PA, Grattan-Smith JD. Clinicopathologic features of nephrogenic rests and nephroblastomatosis. *Adv Anat Pathol.* 2001;8(5):276–289.
5. Ngan KW, Shaari S, Subramaniam T. Juxtarenal/pararenal Wilms' tumour in a six-year-old Malay girl. *Singap Med J.* 2009 Sep;50(9):e329–e331.