

Upper tract low-grade papillary carcinoma in a 20-months-old boy: A rare case report

Karim S.M. Saad¹, Mohamed Elsaid Youssif¹, Ahmed Fahmy¹, Seif Al Islam Nafis Hamdy¹, Ahmed Gamal El Din Hanno¹, Ahmed R. El-Nahas²

¹ Urology Department, Alexandria Faculty of Medicine, Alexandria, Egypt

² Urology Department, Urology and Nephrology Center, Mansoura University, Egypt

Abstract Urothelial tumors in children are rare pathology. A 20-months-old male patient presented with upper ureteric papillary tumor in a setting of ureteroscopy after multiple procedures for cystine stones former. Biopsy revealed low-grade papillary carcinoma.

Key words Upper urinary tract; ureter; urothelial cell carcinoma; child.

Copyright © 2015 [pediatricurologycasereports.com](http://www.pediatricurologycasereports.com).

Corresponding Author

Karim S.M. Saad, M.D.

Lecturer of Urology Urology Department

Faculty of Medicine, Alexandria University

25 Victor Emanuel square, smouha, Alexandria,

Egypt

Email: karim.saad.md@gmail.com

Accepted for publication: 26 March 2015

INTRODUCTION

Urothelial tumors in children are extremely rare pathology. They usually occur in the bladder. Urothelial tumors may be flat or papillary as a consequence of the pattern of growth of the intraepithelial lesion, which may lead to invasive urothelial carcinoma [1-3]. It generally presents with hematuria and tends to be unifocal [1,2]. Squamous

cell and more aggressive carcinomas have also been reported [3, 4].

This case report presents a boy with discovered right upper ureteric papillary tumor. Different techniques of yielding good biopsy specimens and probable causes of tumor occurrence are also mentioned. To our knowledge, this is the youngest child reported to have an urothelial tumor.

CASE REPORT

A 20-month-old baby boy with history of recurrent cystine stones, undergone multiple endoscopic procedures. An upper tract urothelial tumor was discovered in the right upper ureter, during a setting for ureteroscopy.

The child presented with history of ureteroscopy for stone removal. At presentation, the child was toxic, feverish (39.5°C). Urinalysis demonstrated pyuria and microscopic hematuria. Urine culture was positive for *Escherichia coli* growth. Patient received antibiotics according to the results. In contrast computed tomography (CT) study, bilateral ureteral stricture was determined (Fig. 1). Left endoureterotomy was done with Semi-Rigid 4.5 French ureteroscopy (Richard Wolf®, Knittlingen, Germany) using Holmium: YAG laser with fixation of 2 ipsilateral JJ (Fig. 2).

At time of right ureteroscopy, a papillary tumor 5 mm in size was accidentally discovered at the site of the right upper ureteric stricture (Fig. 1).



Fig. 1. Contrast CT show bilateral ureteric strictures (yellow arrow).

Biopsy was taken using piranha forceps by grabbing the tumor stem with rotation of the forceps for the tumor to detach, and visual aspiration of tissues for cytology was obtained. Complete ablation of the papillary base was done using Holmium laser (Auriga 30 W StarMedTech®, Starnberg, Germany). A 3 Fr JJ stent was fixed and the procedure was terminated (Fig. 2).



Fig. 2. After ablation of the papillary base, JJ stent is seen.

Biopsy revealed low-grade papillary carcinoma (Fig. 3).

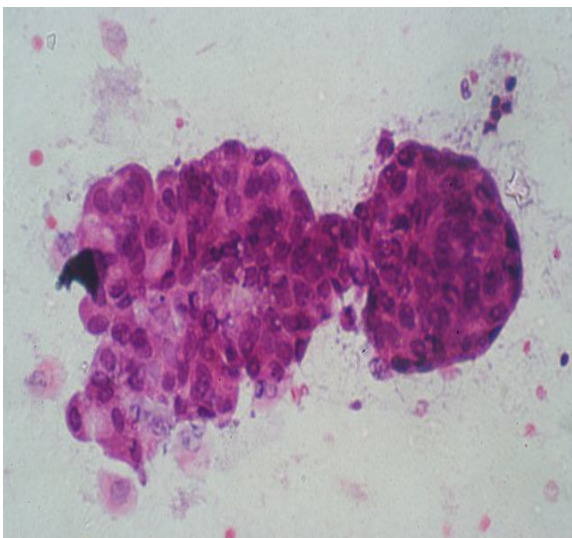


Fig. 3. Low-grade urothelial carcinoma in the ureter, formerly 1973 WHO grade 2 urothelial carcinoma.

After 1 month follow up, diagnostic ureteroscopy with selective urine cytology was free.

DISCUSSION

Urothelial lesions of the upper urinary tract are very rare, accounting for about 5% of urothelial malignancies and 10% of renal tumors diagnosed every year in the United States [5]. The spectrum of urothelial pathology in the renal pelvis and ureter is similar to that seen in the bladder [6]. According to WHO (2004)/ISUP, neoplasms of the urothelium can be flat (planophytic), papillary (exophytic) and inverted (endophytic), depending on their growth pattern relationship with the surface of the surrounding urothelial mucosa [7]. Papillary lesions and neoplasms grow exophytically into the lumen of the urinary system. Papillary tumors recognized in WHO classification includes urothelial papilloma, papillary urothelial hyperplasia, papillary urothelial neoplasm of low malignant potential (PUNLMP), low-grade papillary carcinoma and high-grade papillary carcinoma [8,9].

The majority of low-grade papillary carcinoma patients would have been considered as grade 1 of 1973 WHO and around 70% of grade 2 lesions in the 1973 WHO system. WHO grade 1 neoplasm

showing slight cytologic atypia and mitoses are diagnosed in the 2004 WHO system as LG PUC. At low magnification, there is a generally ordered appearance of the cells within the epithelium. The nuclei tend to be uniformly enlarged but retain the elongated to oval shape of normal urothelial cells. The chromatin remains fine with small nucleoli. Mitoses may be present but are few and remain basally located. These tumors have a significantly higher recurrence rate than that of PUNLMP [7,10].

Symptoms of transitional cell carcinoma may include blood in the urine and pain in the flank or abdomen. Sometimes there are no symptoms at all, and the cancer found incidentally during examination for an unrelated medical problem. To diagnose or rule out transitional cell carcinoma, medical history and symptoms are noted, and blood tests, urine tests and radiologic imaging such as a CT scan or MRI are studies. If transitional cell carcinoma is suspected, ureteroscopy is recommended identifying the therapeutic approaches. During ureteroscopy, a biopsy of the lesions is taken for further examination by a pathologist, who can confirm the grade of the cancer. However, because of the technical difficulties during the application of ureteral and pelvis biopsies, ureteroscopic biopsy specimens are generally smaller than

cystoscopic samples, where the instrument is larger and visualization of the lesion was more accurate. For this reason, specimens taken from the upper urinary tract with the help of ureteroscopy are particularly challenging for pathologists when it comes to accurate tumor diagnosis, grading and staging [11]. Urine cytology evaluating the size and shape of the cells in the urine may also be used in diagnosing transitional cell carcinoma.

The standard management of upper urinary tract urothelial malignancies is radical, open surgery. However, most lesions in the upper urinary tract at the time of presentation are low grade and superficial [12-14]. The evolution in the technology of endoscopes, lasers, special instruments and optics, endoscopic treatment has gained popularity in the treatment of upper tract urothelial carcinoma [15]. For low-grade and smaller cancers, minimally invasive endoscopic management can be applied. The major long-term sequelae of endoscopic therapy is postoperative ureteral stricture. This complication is frequently not secondary to ureteroscopic manipulation but due to the type of energy applied to remove the tumor [16]. YAG laser energy in the treatment of upper urinary tract urothelial lesions has been defined with a low stricture rate [17,18]. *Grasso et al* [16] suggested that

ureteroscopic treatment of malignant urothelial lesions in the upper urinary tract should be reserved for a selected patient population. As with bladder cancers, the initial stage and grade of tumor is the key to defining the ultimate success of treatment.

To the best of our knowledge this is the youngest and only patient presented with an upper tract low-grade urothelial tumor. This may be due to multiple endoscopic

procedures and sustained irritation of ureteric stenting of the delicate ureter of a child. Along with repeated UTI and inflammation of the mucosa all of these can be predisposing factors for urothelial metaplasia.

Acknowledgements

The author(s) declare that they have no competing interests and financial support.

REFERENCES

1. Alanee S, Shukla AR. Bladder malignancies in children aged <18 years: results from the Surveillance, Epidemiology and End Results database. *BJU Int.* 2010;106 (4):557-60.
2. Paner GP, Zehnder P, Amin AM, Husain AN, Desai MM. Urothelial neoplasms of the urinary bladder occurring in young adult and pediatric patients: a comprehensive review of literature with implications for patient management. *Adv Anat Pathol.* 2011;18(1):79-89.
3. Sung JD, Koyle MA. Squamous cell carcinoma of the bladder in a pediatric patient. *J Pediatr Surg.* 2000;35(12):1838-9.
4. Lezama-del Valle P, Jerkins GR, Rao BN, et al. Aggressive bladder carcinoma in a child. *Pediatr Blood Cancer.* 2004;43(3):285-8.
5. Jemal A, Siegel R, Ward E, Hao Y, Xu J, Murray T, Thun MJ. Cancer statistics, 2008. *CA Cancer J Clin.* 2008;58(2):71-96.
6. Holmäng S, Johansson SL. Synchronous bilateral ureteral and renal pelvic carcinomas: incidence, etiology, treatment and outcome. *Cancer.* 2004;101(4):741-7.
7. Mazzucchelli R, Cheng L, Lopez-Beltran A, Scarpelli M, Montironi R. Classification and grading of noninvasive and invasive neoplasms of the urothelium. *Anal Quant Cytol Histol.* 2012;34(3):111-9.
8. Montironi R, Mazzucchelli R, Scarpelli M, Lopez-Beltran A, Cheng L.

- Morphological diagnosis of urothelial neoplasms. *J Clin Pathol.* 2008;61(1):3-10.
9. Laishram RS, Kipgen P, Laishram S, Khuraijam S, Sharma DC. Urothelial tumors of the urinary bladder in Manipur: a histopathological perspective. *Asian Pac J Cancer Prev.* 2012;13(6):2477-9.
10. Montironi R, Lopez-Beltran A, Scarpelli M, Mazzucchelli R, Cheng L. Morphological classification and definition of benign, preneoplastic and non-invasive neoplastic lesions of the urinary bladder. *Histopathology.* 2008;53(6):621-33.
11. Gittes RF. Retrograde brushing and nephroscopy in the diagnosis of upper-tract urothelial cancer. *Urol Clin North Am.* 1984;11(4):617-22.
12. Tawfik ER, Bagley DH. Upper-tract transitional cell carcinoma. *Urology.* 1997;50(3):321-9.
13. Nielsen K, Ostri P. Primary tumors of the renal pelvis: evaluation of clinical and pathological features in a consecutive series of 10 years. *J Urol.* 1988;140(1):19-21.
14. Murphy DM, Zincke H, Furlow WL. Primary grade 1 transitional cell carcinoma of the renal pelvis and ureter. *J Urol.* 1980;123(5):629-31.
15. Fajkovic H, Klatte T, Nagele U, et al. Results and outcomes after endoscopic treatment of upper urinary tract carcinoma: the Austrian experience. *World J Urol.* 2013;31(1):37-44.
16. Grasso M, Fraiman M, Levine M. Ureteropyeloscopic diagnosis and treatment of upper urinary tract urothelial malignancies. *Urology.* 1999;54(2):240-6.
17. Razvi HA, Chun SS, Denstedt JD, Sales JL. Soft-tissue applications of the holmium:YAG laser in urology. *J Endourol.* 1995;9(5):387-90.
18. Erhard MJ, Bagley DH. Urologic applications of the holmium laser: preliminary experience. *J Endourol.* 1995;9(5):383-6.

Access this article online

<http://pediatricurologycasereports.com>

Quick Response Code

