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Low grade papillary urothelial carcinoma in children about a case

Zakarya Alami Hassani[1*], Driss Hanine[2], Yassine Zouirech[3], Abir Manni[4], Freddy Niyongere[5], Naoufal Fejjal[6], Mohamed Rami[7], Rachid Belkacem[8], Mohamed Amine Bouhafs[9]

[1-5, 7-9] Pediatric Urology Department, Rabat Children's Hospital, Ibn Sina UHC / Mohammed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco

[6] Pediatric Plastic Unity, Rabat Children's Hospital, Ibn Sina UHC / Mohammed V university, Faculty of Medicine and Pharmacy, Rabat, Morocco

Abstract
Urothelial tumors of the bladder are a well-known pathology in adults. In children they are very rare and there are no defined guidelines about their management.

We report the case of a 5-year-old boy who presented to our department with gross hematuria. Ultrasonography and cystoscopy with histological study showed a noninvasive low grade papillary urothelial carcinoma. The patient underwent a transurethral resection with no recurrence till date.

Clinically, urothelial tumors of the bladder are mainly revealed by gross and painless hematuria. Definitive diagnosis is obtained by cystoscopy: endoscopic aspect is typical and transurethral biopsy allows the tumor's grading and the elimination of other possible causes of tumor-like lesions. Transurethral resection is the main modality of treatment in children. Due to the significant recurrence rates, a close follow-up is mandatory. This can be done using ultrasonography or cystoscopy. The superiority of either technique is still not evident.

Keywords: urothelial tumors, children, low grade bladder tumors, transurethral resection

Introduction

Tumors of the urinary tract are very rare in children. The first case of urothelial bladder tumor in childhood was reported by Deming in 1924. Since then, about 125 cases have been reported in literature, all identified in a recent review by Lerena and al. [1]. These tumors were shown to have a low grade of malignancy; however, they also showed an important tendency to recur. We report the case of a noninvasive low grade papillary urothelial carcinoma (NILGPUC) in a 5-year-old boy.

Observation

A 5-year-old boy presented to our department for 2 episodes of painless gross hematuria. The first episode occurred 2 years earlier and was treated as a urinary infection. There was no present or previous family history of urinary tract tumor. Physical examination was normal and did not reveal any associated anomaly.

Urinary tract ultrasound revealed a budding bladder lesion of intermediate echogenicity measuring 27 x 20 mm (Figure 1). It was attached to the right posterior bladder wall, with a large implantation base and irregular contours. The mass was avascular to the Doppler and immobile when changing position.

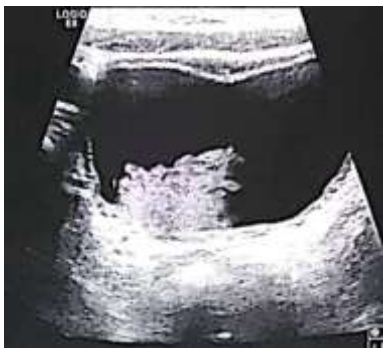

Fig 1: Ultrasonography image of a budding mass growing into the bladder.

At cystoscopy, we visualized a leaf-shaped lesion that resembled a seaweed, of approximately 20 x 20 mm in diameter (Figure 2) just above the right ureteric orifice (Figure 3).

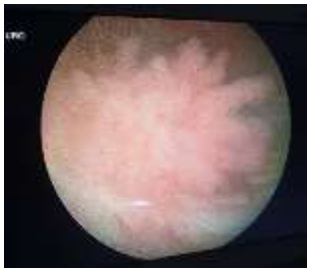

Fig 2: A cystoscopic view of a leaf-shaped lesion corresponding to the tumor.

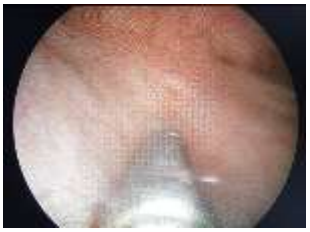

Fig 3: A cystoscopic view showing the lesion lying above the right ureteric meatus.

Most of the tumor was endoscopically excised (Figure 4). Biopsies were sent to two different histopathology laboratories.

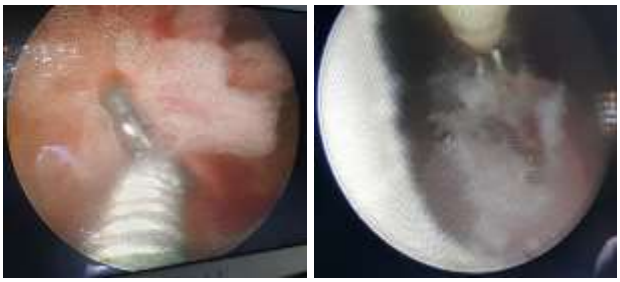

Fig 4: Transurethral resection of the tumor

Initially, the results were discordant, one study revealed a papillary urothelial neoplasm of low malignant potential (PUNLMP), the other was in favor of noninvasive low grade papillary urothelial carcinoma (NILGPUC). A review of histological slides was requested and both laboratories agreed to conclude in favor of a urothelial tumor proliferation with moderate cytologic atypia and Ki-67 expression (Figure 5) but no stromal invasion, all in favor of a low grade papillary urothelial carcinoma (NILGPUC).

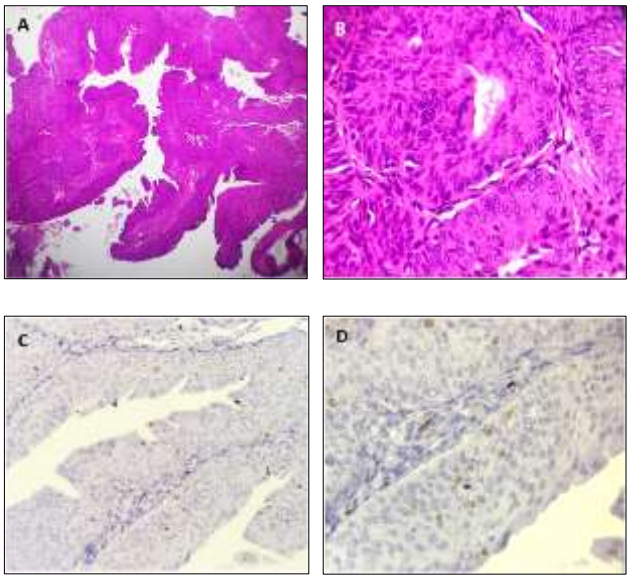

Fig 5: A-B: Low (x40) and higher (x200) magnification showing the histopathology with papillomatous pattern (A-B); C-D: Low (x40) and higher (x200) magnification showing cellular proliferation and Ki-67 expression (C-D).

A cystoscopic reassessment was performed one month later showing a few papillomatous islets lying above the right ureteric meatus. In order to avoid injuring the meatus, a guide was put in place (Figure 6), then the islets were resected and the bed cauterized. A ureteral probe was left in place for 3 postoperative days because of a possible edema. The resected islets were sent for histopathological studies showing inflammatory tissue with no sign of cytologic atypia.

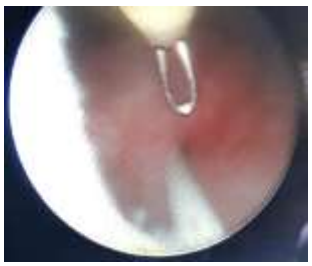

Fig 6: A cystoscopic view showing a guide put into the ureteric meatus to avoid its injury.

Discussion

Bladder tumors are rare in children and both malignant and benign tumors have been reported [2]. Most of urothelial bladder tumors in children are at low risk of malignancy. However, they incur a risk of recurrence. Hence the importance of a good grading allowing a good prediction of tumor prognosis.

In the 1973 World Health Organization (WHO) classification of urothelial neoplasms, papillary tumors fell in to G1 and G2 groups of tumors. Changes have been made in 2016 WHO classification (Figure 7) to reduce variability and increase reproducibility [3]. G1 lesions are now divided into papillary urothelial neoplasms with low malignant potential (PUNLMP) and noninvasive low grade papillary urothelial carcinoma (NILGPUC). G2 lesions are divided into NILGPUC and noninvasive high-grade papillary urothelial carcinoma (NIHGPUC). G3 lesions are described as NIHGPUC. The European association of urology recommend the use of both 1973 and 2016 WHO classifications.

Table 1: World Health Organization (WHO) classification of tumors of the urothelial tract (8).

Infiltrating urothelial carcinoma	Noninvasive urothelial neoplasms
▪ Nested, including large nested ▪ Microcystic ▪ Micropapillary ▪ Lymphoepithelioma-like ▪ Plasmacytoid / signet ring cell / diffuse ▪ Sarcomatoid ▪ Giant cell ▪ Poorly differentiated ▪ Lipid-rich ▪ Clear cell	▪ Urothelial carcinoma in situ ▪ Noninvasive papillary urothelial carcinoma, low grade ▪ Noninvasive papillary urothelial carcinoma, high grade ▪ Papillary urothelial neoplasm of low malignant potential ▪ Urothelial papilloma ▪ Inverted urothelial papilloma ▪ Urothelial proliferation of uncertain malignant potential ▪ Urothelial dysplasia

Epidemiologically, there is a clear male predominance with a sex ratio of 3: 1. There is also an ethnic difference as white patients are more affected [1]. Many risk factors have been identified for urothelial tumors in adults: smoking, professional exposure, low fluid consumption. However, none of these have been cited in pediatric cases, our case included.

Clinically, urothelial tumors of the bladder are mainly revealed by gross and painless hematuria. Some cases of

incidental discovery have also been reported [4]. Ultrasonography can identify bladder lesions, but it cannot specify their nature and degree of malignancy. Only the histological study can confirm the benignity or malignancy of a bladder lesion.

Definitive diagnosis is obtained by cystoscopy, the endoscopic aspect is typical: a mushroom-like growth or a leaf-shaped lesion resembling a seaweed, localized in the trigone area. Transurethral biopsy shows a thickened multilayered urothelium with minimal or no cytologic atypia in case of PUNLMP, and an orderly arrangement of urothelial cells and cytologic atypia in NILGPUC. Transurethral biopsy can also eliminate other possible causes of tumor-like lesions as inflammatory tumors. If a pediatric resectoscope is not available, a transurethral needle biopsy can be used as described by Lightfoot and al [5].

There are no defined guidelines concerning low grade urothelial tumors management in the pediatric population as it is mainly based on the experience with adult patients. Transurethral resection (TUR) is the main modality of treatment in children. Post-operative single-dose intravesical chemotherapy and intravesical immunotherapy are used after TUR in adults to prevent recurrence, however there is insufficient knowledge about their indications and usage in the pediatric population [6].

Urothelial tumors are mostly non-invasive at diagnosis; however, they present a significant risk of recurrence after resection. Reported recurrence rates range from 2.6% to 13% in patients aged <20 years [7]. In adults, follow-up can be assured by urine cytology, however the sensitivity of the latter in well differentiated tumors decreases to 6-38%. As the majority of urothelial tumors in children are well differentiated, urine cytology is not recommended. In children, surveillance methods are still subject to debate. For Polat [4], ultrasonography may be sufficient for follow-up, for other authors cystoscopy is the gold standard despite its disadvantages such as the requirement of general anesthesia and the possibility of urethral trauma.

Conclusion

Urothelial tumors are rare in children, and should be thought of in any case of gross hematuria. Cystoscopy allows diagnosis and transurethral resection. After TUR, recurrence rates remain significant and require close monitoring.

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