TO OPERATE OR NOT TO OPERATE

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Introduction

Pediatric tumors of the urinary bladder are rare, rhabdomyosarcoma being the most frequently occurring lesion. Most are low grade and present at an early stage; reported cure rates approach 70% to 80%. Other bladder tumors are exceedingly uncommon in the pediatric age group. These include transitional cell papillomas, papillary carcinoma, inflammatory myofibroblastic tumors, and very rarely neurofibromas. Neurofibromas of the urinary tract occur infrequently: fewer than 70 cases with bladder involvement have been reported to date and of these only 25 cases occurred in the pediatric population, with only one case described with presentation in infancy. We decided to present a case of a plexiform neurofibromatosis of the bladder in a two months old infant.

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Methods and Materials

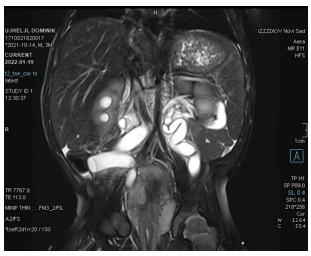
An infant at the age of 2 months was referred for bilateral hydronephrosis and palpable right lower quadrant and suprapubic mass. The initial US examination showed bilateral grade IV hydronephrosis, with a mass in the region of bladder whose origin could not be determined. An initial CT scan of the pelvis was performed and an infravesical and retrovesical mass was clearly seen, again of open etiology (from bowel and/or mesenterial origin, to most likely mesenchimal tumor of the bladder or prostate). Initially, rhabdomyosarcoma was suspected. MRI showed and expansive infiltrative soft tissue tumor that surrounds like a muff caudal aspect of the bladder with both vesicoueretric orifices, prostate and part of the urethra. There is a diffuse thickening of the bladder wall that is up to 18 mm thick. Total dimensions of the tumor were 64x53x56mm. Both ureters are dilated, with bilateral hydronephrosis gr IV. Inguinal lymphnodes slightly enlarged up to 6.8mm on the right and mm on the left. No enlarged mesenterial or retroperitoneal lymph nodes.

Results

Bilateral percutaneous nephrostomy was performed. Needle biopsy of the tumor with a 18 gauge needle was done. Samples were sent for pathohistology. The results showed it was neurofibroma.
Once the percutaneous nephrostomies fell out, surgery was necessary. Initial plan was to remove as much of the tumor as possible, together with bilateral loop ureterocutaneostomies. Intraoperatively prostatectomy had to be done, while we decided to leave the bladder intact, eventhough ex-tempore showed that the posterior bladder wall is also infiltrated with neurofibroma.

MRI findings

The pre and postoperative MRI scans.



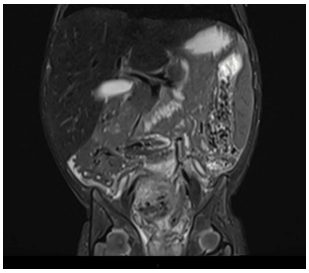


Photo documentation

Photo 1. Elevated bladder after the exstirpation of the tumor and prostate that was completely changed by the tumor

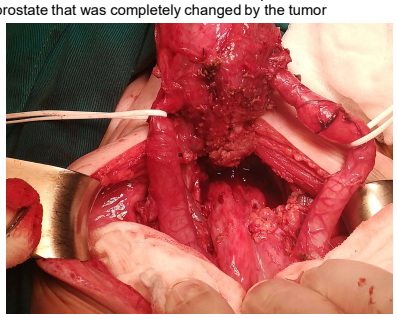


Photo 2. Exstirpated tumor with bilateral ducti deferens

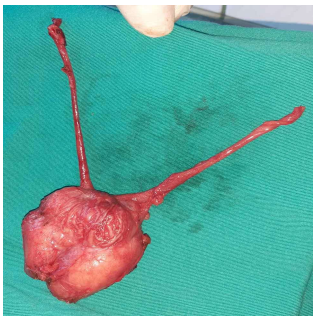
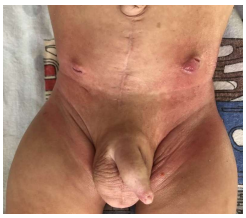


Photo 3. Postoperative aspect of ureterocutaneostomies



Discussion

Most examples of neurofibromas are benign, although approximately 5% of plexiform examples are known to undergo malignant transformation. Only 2 published reports describe neurofibromas of the urinary bladder having undergone malignant transformation. The optimal management of patients with neurofibroma of the bladder is still unclear. Surgical intervention appears the treatment of choice for symptomatic patients. Conservative treatment by transurethral or partial cystectomy may be appropriate for patients without upper tract obstruction. For patients with upper tract obstruction, urinary diversion could be required. It is important to know that the mass, if left in place, often will continue to grow and may cause local pressure effects even if benign.

Conclusions

Neurofibromatosis of the bladder is a rare finding, and surgery could provide a final solution for the patient. In our case we decided to preserve the bladder and to enable urine derivation, as the first step. Patients parents are irrigating the patients bladder in order to preserve the bladder capacity, that is currently 28ml (expected bladder capacity for patients body weight is 55ml). It is stil dificult to evaluate if the patient is continent.

References

Iyimser Üre, Serhat Gürocak, Ipek Isik Gönül, Sinan Sözen, Nuri Deniz Neurofibromatosis Type 1 with Bladder Involvement Case Rep Urol. 2013; 2013: 145076.
A Chakravarti , M A Jones, J Simon. Neurofibromatosis involving the urinary bladder. Int J Urol . 2001 Nov;8(11):645-7
Gulec Mert Dogan, Ahmet Sığırci, Leyla Karaca. Neurofibromas of the bladder in a child with neurofibromatosis type 1 Int Braz J Urol. 2018 Nov-Dec; 44(6): 1256–1257.
Gao B, DeCotiis K, Bobrowski A, Koyle M, O'Kelly F. The association of Neurofibromatosis Type 1 and lower urinary tract dysfunction in the paediatric population - A critical review of literature. J Pediatr Urol. 2020 Jun;16(3):357-365