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Radiology

Bladder Rhabdomyosarcoma: A Case Report

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Abstract

Case Report

Tumors of the urogenital sinus in children are very rare. Rhabdomyosarcoma is the most common tumor. In girls, it includes tumors of the uterus, vagina, cervix, vulva and bladder. Rhabdomyosarcomas account for 8% of all pediatric malignancies. Rhabdomyosarcoma of the bladder is a rare condition. We report an observation of a bladder rhabdomyosarcoma in a one-year-old girl, who consulted for hematuria with impaired renal function. The aim of this paper is to review the clinical symptomatology and radiological appearance of this condition in children, following a literature review of the condition, and to discuss therapeutic options. Rabdomyosarcoma should be considered in the presence of dysuria associated with hematuria in children. Its diagnosis is suspected by imaging and confirmed by anatomopathology.

Keywords: Rhabdomyosarcoma, bladder, CT, child.

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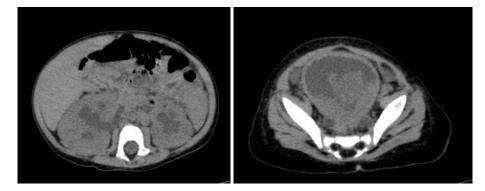
INTRODUCTION

Rhabdomyosarcoma (RMS) is a tumor with striated muscle differentiation developing at the expense of non-bone supporting tissue. It accounts for 4% of solid malignancies in children and 60% of softtissue sarcomas. RMS is the third most common tumor after neuroblastoma and Wilms' tumor. Two-thirds of RMS are diagnosed in children under 6 years of age, with a further peak in adolescence. Rhabdomyosarcoma of bladder location is exceptional in children.

The aim of this study was to report a case of rhabdomyosarcoma of the bladder in children while carrying out a review of the literature, with emphasis on the radiological and evolutionary aspects of this pathology.

OBSERVATION

We report the case of a one-year-old girl, consulted for hematuria with impaired renal function. The patient underwent an abdomino-pelvic CT scan, which revealed a voluminous lesional process in the bladder trigone, budding intra luminally with a cauliflower-like appearance, iso dense on spontaneous contrast, heterogeneously and annularly enhanced after injection of PDC, delineating areas of necrosis with circumferential and irregular thickening of the posterior bladder wall. This process invades the ureteral meatus, causing major upstream ureterohydronephrosis.



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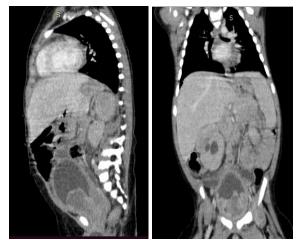


Figure 1: CT axial slices with sagittal and coronal reconstructions, without and after enhancement: arterial and portal times

Voluminous lesion process at the bladder trigone (arrow), budding intra luminally, iso dense to spontaneous contrast heterogeneously enhanced and annular after PDC injection, delimiting necrotic areas. Invades ureteral meatus, causing major upstream ureterohydronephrosis. Circumferential and irregular thickening of the posterior bladder wall.

DISCUSSION

Bladder tumours, whether benign or much more frequently malignant, remain rare conditions in children [1]. Malignant bladder tumours are mainly mesenchymal in nature, such as rhabdomyosarcomas, which account for around 5% of all solid tumours in children [6]. Bladder and/or prostate localization account for 11% of other rhabdomyosarcoma localizations. [6].

These malignant tumors are more frequent in boys than in girls, with a sex ratio of 1/4 [5]. They are characterized by a frequent association with congenital malformations or other solid tumors (osteosarcomas, adrenocorticalomas, brain tumors), within the same family, as well as the frequency of breast cancer in mothers of affected children, suggesting the possibility of a genetic predisposition [5]. We found no particular family history or other congenital malformation in our patient. Clinically, these tumors present with nonspecific symptoms. These include: episodes of acute or subacute retention of urine, which is very often the first clinical sign of vesicoprostatic rhabdomyosarcoma in boys [5]; recurrent urinary tract infections, sometimes with haematuria, which is often a sign of onset; more rarely, a hypogastric mass, which may be discovered incidentally in cases of bladder globe.

Anatomopathologically, it is an embryonic mesenchymal tumour, most often endo-vesical in development, almost exclusively of the botryoid type, located in the "cauliflower-shaped" or "cluster-shaped" bladder fundus, more rarely in the bladder dome. Radiological findings, particularly bladder ultrasound, reveal an intra-vesical mass. Extension workup should systematically look for lymph node involvement. CT scans provide the best possible indication of tumour extension, particularly in the lymph nodes. It should also look for pulmonary and bone metastases.

The prognosis of rhabdomyosarcoma depends on age, tumor stage (tumor extension, lymph node involvement) and histological type. The S.I.O.P. (International Society of Pediatric Oncology) distinguishes 4 stages of increasing severity from 1 to 4. Stage I: Tumor localized to the organ, without lymph node invasion, without metastases (T1, Mo, No), Stage II: Invasion of one or more contiguous organs without lymph node invasion, without metastases (T2, Mo, No), Stage III: Regional lymph node invasion, without metastases (T1, T2, Mo, No) and Stage IV: distant metastases (T1, T2, No, N1, M1).

Favourable prognostic factors in bladder RMS are: absence of metastasis, embryonic histology, tumour less than 5 cm in diameter and removable, and age of the child at diagnosis less than 10 years. These factors are independent. The prognosis is particularly grave because of the vesicoprostatic localization compared with other sites (orbit, testicles, vagina) [7]. The treatment of rhabdomyosarcoma combining surgery, radiotherapy and chemotherapy is disappointing, despite the combination of different therapeutic modalities: the overall survival curve for all teams barely reaches 50-60% at 5 years [5].

CONCLUSION

Bladder rhabdomyosarcoma in children is a rare tumour, and its diagnosis must be suspected in the presence of an obstructive lower urinary tract syndrome, whether or not associated with haematuria and/or infection, on the basis of medical imaging data. If possible, cystoscopy with biopsy confirms the diagnosis on anatomopathology. After surgical treatment, the evolution is marked by local recurrence or metastasis. These recurrences can be prevented by chemotherapy and/or radiotherapy. But despite these therapeutic combinations, the prognosis for rhabdomyosarcoma remains poor.

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