CLINICAL CASE

Bladder rhabdomyosarcoma in a 6-month-old girl: a case report

I. Camacho-Guerrero*, L. Moussali-Flah, J.L. Jiménez-Mariscal, O. Burgos-Santos, J. Domínguez-Bravo and L. Ramírez-Patiño

Hospital Pediátrico de Tacubaya, Mexico City, Mexico

Received 13 October 2015; accepted 25 January 2016
Available online 7 March 2016

Abstract
Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood, more frequently affecting males, with an incidence of 8 children per one million. It is found in the genitourinary tract in 26% of cases. Differential diagnoses are ureteral polyps and ureterocele. The most frequent subtype in children is embryonal rhabdomyosarcoma and the botryoid variant with a polypoid form arises from the cavitated structures. The characteristics of a case of botryoid rhabdomyosarcoma in the bladder cavity of a 6-month-old infant girl and the importance of its differential diagnoses are described. We present herein a rare case of bladder tumor in the bladder cavity for a better understanding and recognition of this type of disease in pediatric urology.

© 2016 Sociedad Mexicana de Urología. Published by Masson Doyma México S.A. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

PALABRAS CLAVE
Rhabdomiosarcoma; Rhabdomiosarcoma embrionario; Sarcoma botrioides

Caso de rabdomiosarcoma vesical en niña de 6 meses
Resumen
El rabdomiosarcoma es la variedad de sarcoma de tejidos blandos más común en la niñez, la cual afecta más frecuentemente a varones y tiene una incidencia anual de 8 niños por millón; lo encontramos en el tracto genitourinario en el 26% de los casos. Existen diagnósticos diferenciales como son el pólipo ureteral y el ureterocele. El subtipo más frecuente en niños es el embrionario, siendo la variedad botrioides la que surge de estructuras cavitas; tienen forma polipoide. En este artículo describimos las características de un caso de rabdomiosarcoma variedad botrioides en cavidad vesical en una lactante de 6 meses de vida y la importancia de sus diagnósticos diferenciales. Se presenta un caso raro de tumor vesical en vejiga. Se da a
Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood and is characterized by local extensive invasion and early hematologic and lymphatic dissemination. It belongs to the class of tumors with small, round, blue cells and its histologic characteristic is striated skeletal muscle. The clinical and histologic characteristics are presented herein of a case of botryoid (from the Greek botrys, grapes) embryonal rhabdomyosarcoma in a 6-month-old infant girl seen at the Pediatric Urology Service of the Hospital Pediátrico de Tacubaya.

Case presentation

A 6-month-old firstborn infant girl had no remarkable prenatal history. At 6 months of age the patient was constipated for 12 days and was treated by a general physician with rectal glycerin and amikacin, showing partial clinical improvement. The same physician later treated her for bowel movement alterations with trimethoprim/sulfamethoxazole with no improvement and she was taken to a government hospital. The patient continued to reject an oral diet and had persistent constipation and hyporexia. Physical examination revealed skin and tegument pallor ++, a regular hydration status, neurologic integrity, and no cardiopulmonary alterations. She had important abdominal distension and apparent pain upon palpation. She also presented with reduced diuresis, for which she was admitted to the hospital. The patient was initially managed with Foley catheter placement, intravenous hydration, antibiotics, and hemodynamic stabilization. Urinalysis results were consistent with urinary tract infection. The patient was evaluated by a pediatrician who ordered ultrasound imaging of the urinary tract, which revealed bilateral hydronephrosis. Suspecting a surgical pathology the patient was referred to the pediatric urology service at the Hospital Pediátrico Tacubaya.

Another bladder ultrasound was done that reported an echogenic heterogeneous mass occupying the bladder cavity with regular and lobulated edges and the presence of the Foley catheter balloon (fig. 1).

Kidney ultrasound identified mild bilateral hydronephrosis, and no duplex collecting system or parenchymatous lesion (fig. 2).

Lateral cystourethrogram showed a filling defect at the bladder floor level with a lobulated aspect and a slight prolongation toward the prostatic urethra (fig. 3).

The presumptive diagnosis of intravesical tumor was established, with bladder rhabdomyosarcoma as the likely diagnosis and bladder polyp and ureterocele as the differential diagnoses.

Surgical exploration of the bladder was carried out with excisional biopsy. The surgical findings were an 8 cm exophytic, lobulated, gelatinous tumor dependent on the bladder neck, with a brownish fluid in the bladder cavity (fig. 4).

The cytology study of the samples was positive for malignant cells and the histopathology study reported a high-grade fusocellular sarcoma with surgical margin lesion and ulcerated mucosa.

Postoperative computed axial tomography scan revealed generalized thickening of the bladder wall that could be related to neoplastic extension.

Figure 1. Bladder ultrasound showing a mass with heterogeneous echogenicity occupying part of the lobulated bladder cavity.
Figure 2  Kidney ultrasound showing mild bilateral hydronephrosis.

Figure 3  Lateral cystourethrogram showing a filling defect at the level of the bladder floor with a slight prolongation toward the prostatic urethra.

Figure 4  The surgical exploration revealed an 8 cm, lobulated, gelatinous tumor dependent on the bladder neck, with a brownish fluid in the bladder cavity.

The patient was referred to the tertiary care pediatric oncology surgery service for multidisciplinary management, follow-up studies, and definitive treatment.
Discussion

Rhabdomyosarcoma is the most common of sarcoma-type tumors in childhood, with an incidence of 8 children per million. Its prevalence is slightly higher in the male sex (ratio 1.4:1). Seventy percent of all rhabdomyosarcomas occur in children under the age of 10 years.

Striated skeletal muscle is a histologic characteristic and the tumors are derived from rhabdomyoblasts or primitive muscle cells. They can appear in any part of the body. They belong to the class of small round blue-cell tumors that includes neuroblastoma, Ewing's sarcoma, peripheral neuroectodermal tumors, non-Hodgkin's lymphoma, and leukemia.

In 1958, Horn and Entenline identified 4 subtypes: embryonal 54%, alveolar 19%, botryoid 4.5%, and pleomorphic < 1%. Currently only 3 main groups are considered, based on histology study and outcome: favorable prognosis (botryoid or fusiform cell), intermediate prognosis (embryonal), and poor prognosis (alveolar and undifferentiated).

Their distribution in relation to frequency is: head and neck 35%; genitourinary tract 26%; extremities 19%; and others 20.

Some hereditary syndromes can predispose to the development of rhabdomyosarcoma, such as Li-Fraumeni syndrome, Werner syndrome, tuberous sclerosis, neurofibromatosis, and nevoid basal cell carcinoma syndrome.

The fusocellular and botryoid variety is the most frequent in childhood. It has a polypoid appearance with small, round cells and a distinctive myxoid matrix of cells in the submucosa known as the cambium layer.

This type of tumor represents close to 4.5% of all cases of rhabdomyosarcoma. The clinical presentation of the botryoid variant of embryonal rhabdomyosarcoma of the bladder and prostate corresponds to 10% of the 26% of the genitourinary rhabdomyosarcomas. Symptomatology is based on hematuria and obstruction of the bladder exit. Early hematologic dissemination to the bone marrow and lung is more common in these tumors of the prostate than of the bladder.

Treatment is based on clinical stage, primary tumor location, and disease extension.

Rhabdomyosarcomas of the prostate are candidates for bladder sparing and prostatectomy with reconstruction of the urethra or bladder neck, chemotherapy, and radiotherapy.

The clinical case presented herein shows the process involved in reaching early diagnosis and opportune treatment, as well as the differential diagnoses that need to be ruled out when diagnosing an intravesical mass.

One of the differential diagnoses was bladder polyp. It is very rare in children and we ruled it out in the ultrasound study that revealed a solid heterogeneous mass without the characteristic thin stem.

Ureterocele was ruled out in the excretory urography and cystourethrography images because there was an absence of filling at the level of the bladder cavity. The existence of a duplex collecting system was not corroborated, and as we know, the majority of cases of ureterocele are related to a duplex collecting system. The ultrasound images we ordered reported a mass with solid and heterogeneous echogenicity.

Ethical responsibilities

Protection of persons and animals. The authors declare that no experiments were performed on humans or animals for this study.

Data confidentiality. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Financial disclosure

No financial support was received in relation to this article.

Conflict of interest

The authors declare that there is no conflict of interest.

References