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Infant prostatic Rhabdomyosarcoma: A diagnostic and therapeutic challenge

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ABSTRACT

The following case report describes a case of prostatic rhabdomyosarcoma in a 6-month-old male who presented with urinary retention and constipation. MRI showed a prostatic mass that was displacing the rectum and bladder, leading to bladder outlet obstruction. A suprapubic tube was placed for urinary diversion and a transvesical approach was used for tissue diagnosis. Biopsy confirmed the diagnosis of prostatic rhabdomyosarcoma. Patient underwent chemotherapy regiment with VAC (vincristine, actinomycin D and cyclophosphamide) and subsequently ifosfamide and doxorubicin. Eventually, due to tumor progression, the patient underwent a radical cystoprostatectomy with pelvic lymph node dissection and ileal conduit.

1. Background

Rhabdomyosarcomas (RMS) are aggressive soft tissue sarcomas that derived from mesenchymal tissue, specifically striated muscle.[1] There is a bimodal age distribution, peaking in the first 2 years and adolescence. About 20% of RMS arise in the Genitourinary System, most commonly in the prostate, bladder, and paratesticular region.[2] Prostatic RMS typically present with mass effect symptoms such as hematuria, azotemia, frequency, urinary retention and/or constipation.

Genitourinary (GU) RMS have different histologic types: embryonal, alveolar, and spindle cell/sclerosing.[3] Embryonal RMS can further be categorized into typical, botryoid and dense subtypes.[1] Embryonal is the most common histology, roughly seen in 90% of all GU RMS cases, is usually found at a younger age and is associated with improved survival. Conversely, alveolar histology RMS is seen in older patients and has a worse prognosis.[4]

Complete resection of pelvic RMS can be attempted upfront only if organ preservation is possible. Initial vincristine, actinomycin and cyclophosphamide (VAC) chemotherapy is standard of care. Following chemotherapy, local control is typically employed with surgery or radiation therapy. In patients with localized pelvic RMS, 5-year failure-free survival (FFS) is estimated at 75%, with most local failures occurring within 3 years of treatment.[2]

2. Case report

The patient is a 6-month-old male, born full term via spontaneous vaginal delivery, with no significant past medical history who presented to the emergency center with constipation and decreased urinary output for two weeks, which acutely worsened with evidence of anuria for greater than 24 hours. Per his parents, he had normal wet diapers until the symptoms began, with a history of constipation treated with Miralax the prior month with mild improvement. The patient had been to another emergency room two days prior and treated for suspected constipation. They deny eating disorders, diet intolerance, or any history of renal or urological disease in the family. Physical Exam revealed no abnormalities besides a soft, distended, palpable bladder.

A Complete Metabolic Panel returned normal values. Urinalysis revealed hematuria of >3 RBC/hpf with no signs of infection. A Renal/Bladder Ultrasound showed a bladder volume of 154 cc, more than double the expected capacity for his age (60 cc). A straight catheter was placed with ease with volume of 150 cc. The emergency team increased his intravenous fluids, bowel regimen and monitored him for voids, however his urinary output did not improve and therefore was admitted to the hospital for further work up with a VCUG and urology assessment.

(Fig. 1) VCUG did not show evidence of ureteral reflux or posterior urethral valves, however demonstrated evidence of bladder outlet

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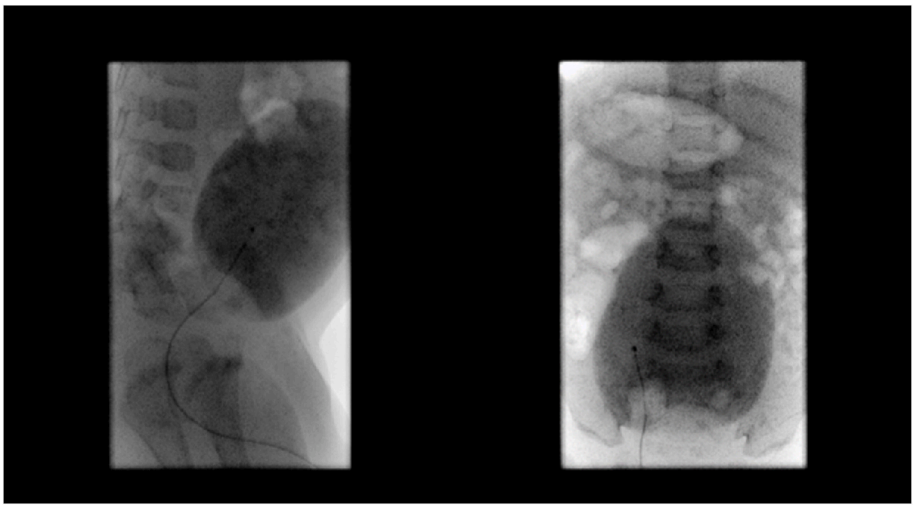

Fig. 1. VCUG

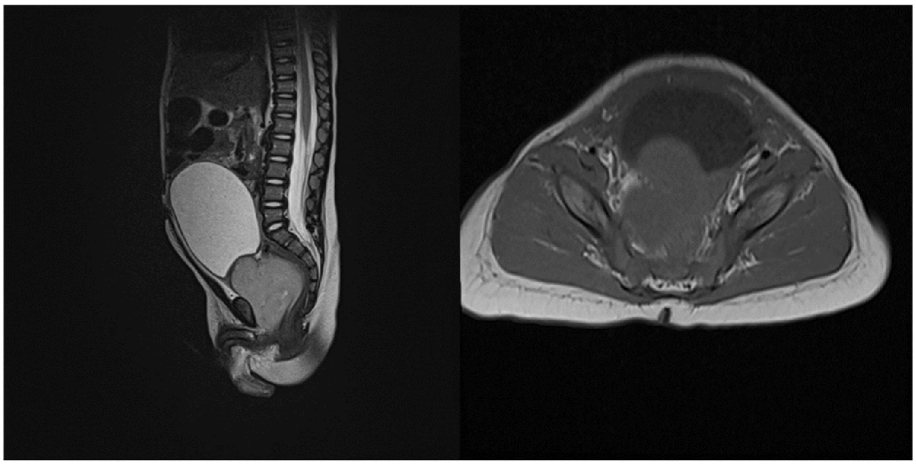

Fig. 2. MRI A) sagittal view B) axial view.

obstruction and impression of the prostate into the bladder (Fig. 2). MRI was recommended, this showed evidence of a well-defined, T2-hyperintense, T1-hypointense mass in the mid and right prostatic region with restricted diffusion and diffuse heterogeneous internal enhancement. It measured 4.9 × 3.6 × 4.9 cm and displaced the bladder anteriorly and superiorly and the rectum to the left. There was abutment of the right inferior pelvic wall and symphysis pubis anteriorly and inferiorly. No definitive invasion of the surrounding structures was seen. An infant resectoscope was not available and hence the decision was made to obtain a tissue diagnosis via transvesical and transtrigonal biopsy with an 18G tru-cut biopsy needle, with subsequent SPT placement. The pathology specimen demonstrated embryonal prostatic RMS. Cytogenetics demonstrated normal FISH, no FOX01 gene rearrangement and positive HRAS mutation. Metastatic work up with a dedicated chest CT with abdomen/pelvis resulted negative (Fig. 3).

Given locally advanced disease the patient was immediately started on VAC chemotherapy regimen. However, given tumor progression he was switched to ifosfamide and doxorubicin. Post-systemic therapy imaging showed poor local tumor control with increase of mass size from 4.9 × 3.5 × 4.8 cm to 6.8 × 4.1 × 6.1 cm with displacement of the bladder and rectum and enlarging bilateral inguinal and right external iliac lymph nodes. The case was discussed during tumor board multidisciplinary meeting and decided with proceeding with a radical cystoprostatectomy and ileal loop. After our team discussed the risks and benefits associated with the procedure and other urinary diversion options available with the family, they opted for an ileal loop diversion.

Pathologic exam of the cystoprostatectomy specimen showed embryonal rhabdomyosarcoma with extensive cytodifferentiation. There were negative margins for the urethra and vas deferens. There was a reported positive margin at the anterior right and left anterior quadrant of the prostate. However, this was likely due to the prostate's capsule tearing during resection. All grossly visible disease was resected and there was no tumor spillage. An extended pelvic lymph node dissection was conducted with no evidence of metastatic disease on pathology. The ileal conduit was performed in the standard fashion.

At 13 months post-operative, the patient remains without evidence of recurrent disease with good clinical improvement.

3. Discussion

This case highlights several important characteristics for young children with GU RMS. First, GU RMS should be on the clinician's differential in a young child with onset of urinary retention as it is often the presenting sign. In this case, the ultrasound and VCUG were ultimately read as normal, however on further inspection, one can appreciate that the bladder neck does not fill appropriately the normal anatomy distorted by a potential mass.

Due to narrow urethral caliber in infants, cystoscopy with tumor

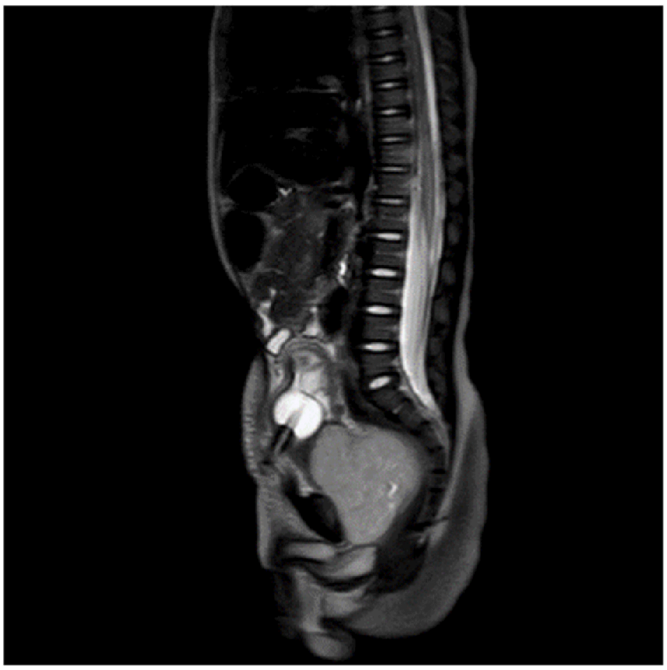

Fig. 3. Chest CT with abdomen and pelvis. Sagittal view.

resection can be challenging. This case highlights that an open transvesical transtrigonal approach through a 2-cm suprapubic incision can be effective in obtaining tissue diagnosis. The biopsy was done in a "needle over finger" fashion, directly visualizing the ureteral orifices as well, thus ensuring safety.

Finally, this case highlights that younger children may have poorer outcomes. Histologically, he had more favorable features (embryonal), and no FOX01 gene rearrangement which would suggest that he should have had a better clinical outcome. However, multiple studies highlight that age <1 year at diagnosis is a significant adverse prognostic factor.[5] Additionally due to very young age, radiation therapy may result in devastating side effects including bowel irradiation and poor growth. In children <1 year at diagnosis, local control, including surgical resection, when necessary, is critically important to successful treatment.

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