Urology Case Reports 43 (2022) 102109

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Urachal rhabdomyosarcoma: A case report of an extremely rare localisation

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ARTICLE INFO

Keywords:
Rhabdomyosarcoma (RMS)
Urachus
Pediatrics
Oncology

ABSTRACT

Urachus is a tubular structure connecting the allatois to the bladder's apex, in the embryonic development. We report a rare case of a 5-year-old boy, with no past medical history, complaining of secondary enuresis, pollakiuria and urgent incontinence. Physical exam revealed a hypogastric mass. Echo guided percutaneous biopsy followed by a histological analysis showed embryonal RMS. The remainder of the staging ruled out metastasis. The patient received neoadjuvant chemotherapy before proceeding to complete tumor excision. Surgical exploration revealed that the tumor was primitively related to the urachus. Total resection was performed. The one year follow-up was uneventful.

1. Introduction

Rhabdomyosarcoma (RMS) is an aggressive childhood tumor that can appear anywhere in the body. It is the most common soft tissue sarcoma in children and adolescents.[1] Four major subtypes are distinguished: Embryonal, alveolar, spindle cell/sclerosing and pleomorphic rhabdomyosarcoma. Embryonal rhabdomyosarcoma (ERMS) and alveolar rhabdomyosarcoma (ARMS) are by far the most frequent.[1] Head, neck and genitourinary are common sites for ERMS while ARMS typically occurs in extremities. RMS arising from the urachus remains rare, and extremely few cases have been reported in literature. We report a case of urachal RMS in a five-year-old boy.

Our aim was to assess the clinical characteristic of RMS in this unusual localisation and its challenging management.

2. Case report

A 5 year-old boy without any medical history presented with secondary enuresis and signs of lower urinary tract: pollakiuria and urgent incontinence since one month with no hematuria. The patient's general condition on admission was fairly good. Physical examination showed a hypogastric voussure. Palpation revealed a well delimited solid mass located in the infraumbilical region, mesuring 10 cm. Rectal examination unveiled an anterior mass. Abdominal ultrasound revealed a voluminous heterogeneous tissular pelvic mass, mesuring 80 × 85 × 64 mm. The mass contained necrotic regions and was vascularized on Color Doppler examination. The bladder was displaced anteriorly and to the left without evidence of intra-luminal flooding. An echo guided percutaneous biopsy followed by a histological analysis with immunohistochemistry staining showed embryonal rhabdomyosarcoma (Fig. 1).

We followed up by a pelvic MRI that showed a voluminous mass, displacing the bladder and infiltrating its lateral wall without further intravesical invasion (Fig. 2a 2b). The mass compresses the sigmoid with loss of the fat plane in between, comes in contact with L5 S1 without bone signal abnormalities and presses against both ureteres causing dilatation upstream (Fig. 2c).

No lymphadenopathy was identified. The remainder of the staging ruled out metastasis. The patient received 4 cycles of neoadjuvant chemotherapy IVA (ifosfamide, vincristine, and dactinomycin). The tumor responded partially, therefore, we continued with the chemotherapy sessions to finally totalize 9 cycles that were well tolerated before complete tumor excision. The surgical exploration revealed that the tumor was primitively related to the urachus. Careful mobilization of the tutor was performed, with en-bloc resection including the bladder dome cuff. The surgery was macroscopically completed. A definitive diagnosis of embryonal rhabdomyosarcoma of the urachus was made. Histopathology of the tumor confirmed its complete removal. Postoperative course was uneventful. Chemotherapy was administrated followed by external abdominal radiotherapy that were well tolerated. At one year follow-up, the patient remains well and free of clinical disease upon physical examination and computed tomography scan.

3. Discussion

Since it's developed from mesenchymal cells, RMS can occur anywhere in the body. Over the last 30 years prognosis of RMS is becoming

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https://doi.org/10.1016/j.eucr.2022.102109
Received 27 March 2022; Received in revised form 26 April 2022; Accepted 4 May 2022
Available online 10 May 2022

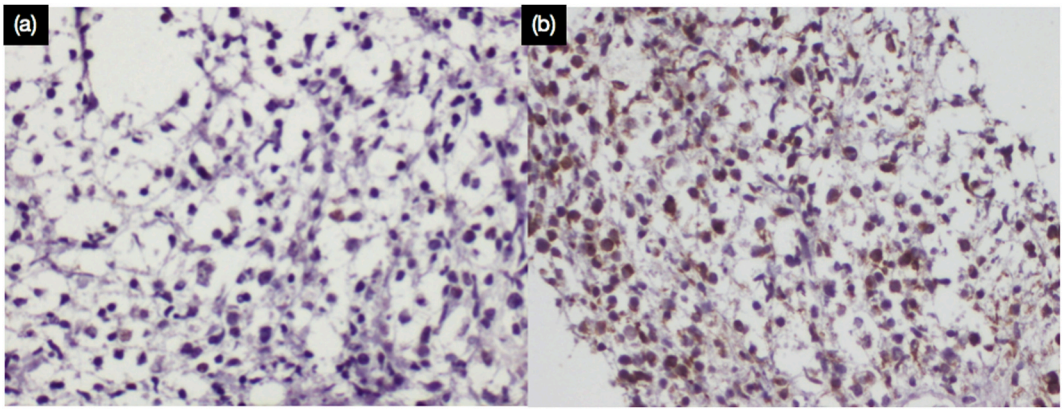

Fig. 1. Tumoral cells showed positive myogenic (a) desmin (b) markers in histopathology.

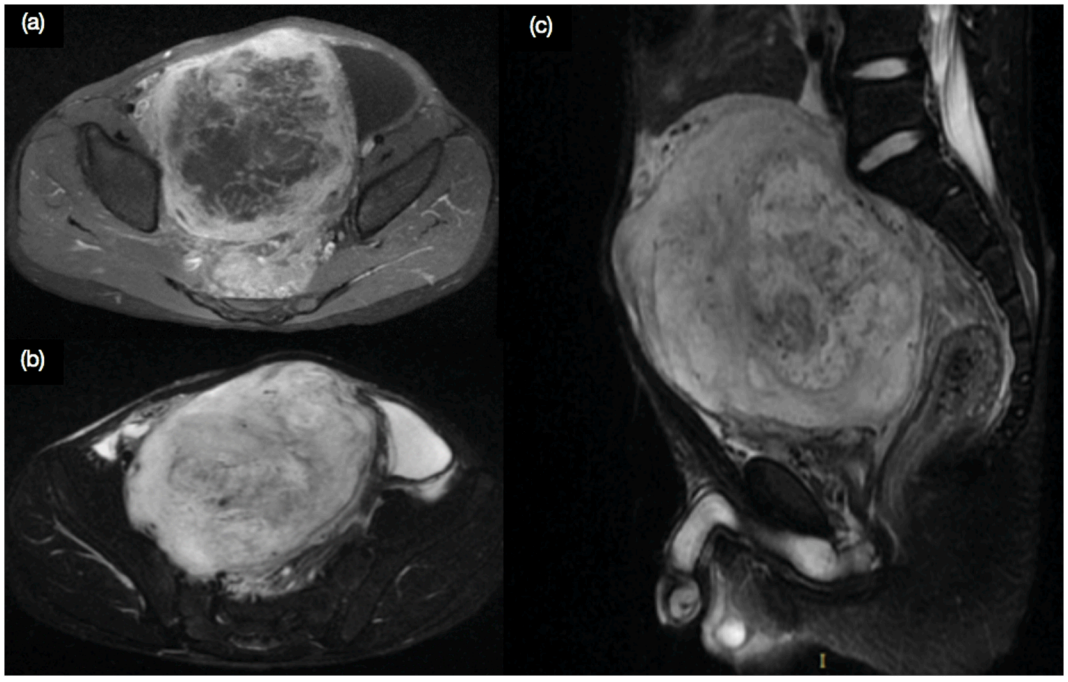

Fig. 2. Pelvic MRI large anterior medial mass displacing the bladder.

Table 1
Clinical and anatomic pathology characteristics of Urachal RMS in literature.

	year	gender	Age at diagnosis (years)	Symptoms	Tumor size mm	histology
Ransom et al	1930	F	0.34	Abdominal mass	115	NOT
Cheikhelard et al.	1983	F	5.5	NOT	50	NOT
Schulz et al	O	M	2	0	0	embryonal
Yokoyama et al	1995	M	2	Abdominal mass	92.5	embryonal
Fernández et al.	2002	F	6	intractable constipation	140	embryonal
Cheikhelard et al.	2003	F	3.3	Peritoneal rupture	100	alveolar
Cheikhelard et al.	2005	F	2.6	dysuria	150	embryonal
Cheikhelard et al.	2005	M	4.2	dysuria	100	embryonal
Cheikhelard et al.	2005	M	5.3	Abdominal mass	106	embryonal
Cheikhelard et al.	2006	M	2.5	obstructive renal insufficiency	210	embryonal
Cheikhelard et al.	2010	M	4.5	Abdominal pain	70	embryonal
Cheikhelard et al.	2010	M	6	Abdominal pain	140	embryonal
Our case	2021	M	5	secondary enuresis	85	embryonal

NOS: not otherwise specified.

better due to multimodal therapies.[1] Clinical approach of RMS in children strongly depends on the location of the tumor, local and distant sites extension, pathological stage, and histological subtype.

The urachus origin for RMS is extremely rare and insufficiently identified. Few single cases were reported in literature with only one pediatric series including eight cases of urachal RMS in children.[2]

Information about the only twelve cases of urachal RMS reported in english literature are detailed in Table 1.

During embryogenesis, fetal bladder and allantois are connected by the urachus. The urachal lumen is obliterated making the umbilical ligament.[3] When obliteration is unfinished urachal remnants occur. Virtually they are present in all newborns and will regress with age. Management used to be a prophylactic urachal excision to avoid malignancy in adulthood. It was later dismissed since no evidence of relationship between patent urachus and the onset of cancer was retained.[2]

Malignant Urachal Neolplasias are extremely rare. They occur more frequently in male adults.[4] 80% of them are adenocarcinomas. Nevertheless sarcomas are the most frequent in patients younger than 20 years of age.[4] In children urachal tumors include RMS, leiomyosarcoma, inflammatory myofibroblastic tumor, neuroblastoma and yolk sac tumor but no adenocarcinoma.[2] Ominous prognosis is the only point that pediatric and adult urachal cancer can share. Cheikhelard et al. found that the urachal location worsens the prognosis of RMS in children.[2]

Of the twelve cases reported in literature only one was diagnosed ARMS, nine were ERMS and the two oldest cases not otherwise specified.[2] Our case is the tenth ERMS.

The preperitoneal location allows a major asymptomatic spreading in the Retzius space. Silent progression means delay of diagnosis. Symptoms appear when the tumour reaches large volumes impending compression of neighbouring organs. Urachal tumors are discovered as a hypogastric palpable mass without hematuria, except adenocarcinomas.[4] Width of the mass can make it impossible to define its primary origin. Management of RMS should provide a cure with the least morbidity. Radical cystectomy en bloc is no more indicated as a primary treatment. The most appropriate surgical intervention is believed to be total mass resection while preserving quality of life. Since urachus RMS are often extensive, surgery will be preceded by chemotherapy to reduce the volume and to provide a non aggressive resection. In our case, 9 cycles of neoadjuvant chemotherapy as a first approach with IVA, according to the European Pediatric Soft Tissue Sarcoma Study Group 2005.[5] Surgery and radiotherapy are the pillars of local treatment in non-mestastatic RMS. Their aim is to cure patients while insuring minimal long term sequelae.

4. Conclusion

Although rhabdomyosarcoma is the most frequent soft tissue tumor in children, those arising from the urachus are extremely rare. They are assembled in the “abdominal and other locations” group for oncological treatment purpose, and therefore not well characterized. They seem to be of a higher risk than other urogenital locations, with a poor prognosis, most probably due to its long asymptomatic evolution.

Consent

Signed consent was obtained from the patient.

Declaration of competing interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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