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LAPAROSCOPY

Case Report

Robotic-assisted laparoscopic partial cystectomy for symptomatic urachal hamartoma

Semptomatik urakal hamartom için robot yardımlı laparoskopik parsiyel sistektomi

Richard Shepler[1], Jack M. Zuckerman[1], Dean Troyer[2], John B. Malcolm[1,3]

ABSTRACT

We report a case of an urachal hamartoma in a 30-year-old African American woman. The urachal lesion was excised with a robotic-assisted laparoscopic partial cystectomy. Pathologic analysis revealed cysts, smooth muscle, and ciliated epithelium consistent with a hamartoma. The patient recovered without complication. This case highlights an unusual pathology that is infrequently reported following urachal remnant excision.

Keywords: Hamartoma; laparoscopy; partial cystectomy; urachus

ÖZ

Otuz yaşında bir Afrika kökenli Amerikalı kadında saptanan bir urakal hamartom olgusunu bildiriiyoruz. Robot yardımlu laparoskopik parsiyel sistektomiyle urakal lezyon eksize edilmiştir. Patolojik analiz hamartoma ile uyumlu kistler, düz kas ve siliyer epitelin varlığını göstermiştir. Hasta komplikasyon oluşmadan iyileşmiştir. Bu olgu urakal kalıntının eksizyonundan sonra nadiren bildirilen olağandışı bir patolojiyi aydınlatmaktadır.

Anahtar kelimeler: Hamartoma; laparoskopi; parsiyel sistektomi; urakus

Introduction

The urachus is an early embryonic structure, which is obliterated before birth. It coalesces with the umbilical arteries to form the adult umbilical ligament in two-thirds of individuals. Benign and malignant lesions may arise from the remnants of this structure, with urachal adenocarcinoma being the most common malignant histology.[1,2] The differential diagnosis for urachal lesions also includes urothelial carcinoma, small cell carcinoma, malignant metastasis, or direct invasion from adjacent anatomical structures.

In contrast to carcinoma, hamartomas are tumor-like malformations composed of overgrowth of mature cells and tissues normally present in the affected part. Hamartomas are benign, with growth rates similar to surrounding structures, and they have no propensity for local invasion or metastasis. Hamartomas most frequently occur in the lung, comprising 10% of all solitary pulmonary nodules. Other common locations include the heart, brain, skin, kidneys, spleen, vessels and eyes.[3] Despite their benign nature, these lesions may become symptomatic. When located superficially, such as on the skin or in the head and neck, hamartomas can be disfiguring; in the colon, they may be obstructive; renal hamartomas may result in significant hemorrhage; cerebral hamartomas may induce seizures or cause other central nervous symptoms via mass effect.[4]

We report a case of a urachal remnant hamartoma causing irritative bladder symptoms in a young female patient. Institutional review board approval was obtained prior to performing a chart review.

Case presentation

A 30-year-old African American female presented with dysuria, urinary frequency and nocturia

[1]Department of Urology, Eastern Virginia Medical School, Norfolk, VA, USA

[2]Department of Pathology, Eastern Virginia Medical School, Norfolk, VA, USA

[3]Urology of Virginia, PLLC, Virginia Beach, VA, USA

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Correspondence: Jack M. Zuckerman
E-mail: jmzuckerman@gmail.com

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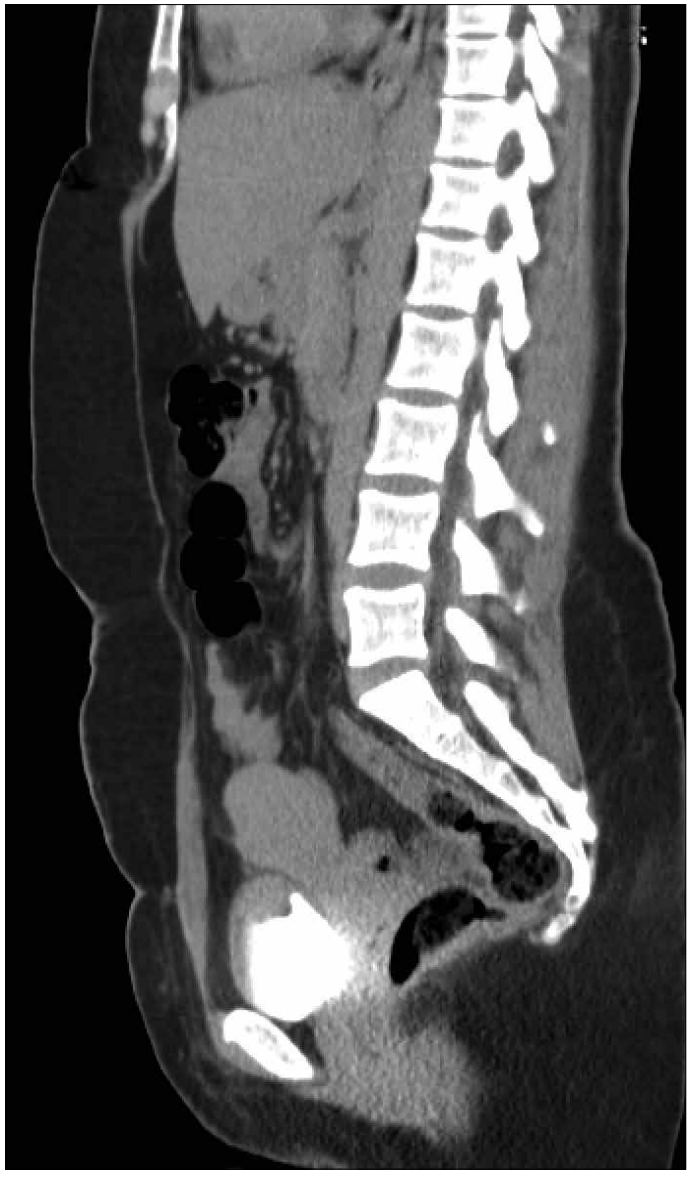

Figure 1. Sagital CT abdomen/pelvis with urachal mass

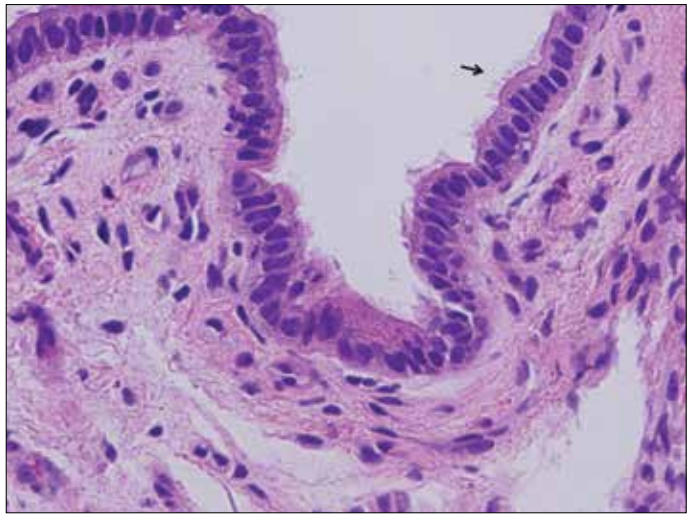

Figure 2. H&E slide demonstrating ciliated columnar cells (arrow)

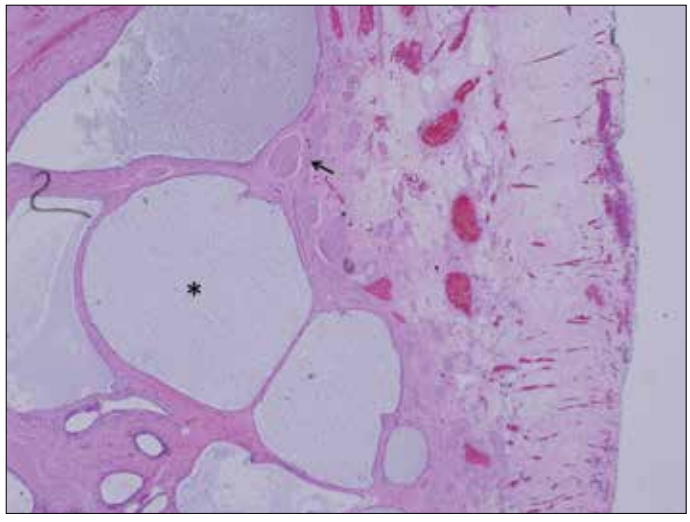

Figure 3. H&E slide demonstrating numerous cysts (asterisk) with smooth muscule bundles (arrow)

for more than a decade. She was referred for urologic evaluation after a transvaginal ultrasound by her gynecologist identified a bladder lesion concerning for malignancy. Urinalysis demonstrated trace blood and leukocyte esterase, with no nitrites or pyuria. Urine culture and cytology were negative. A computed tomography urogram revealed a 2 cm mass at the dome of the bladder, concerning for an urachal remnant tumor (Figure 1). Cystoscopy with cold-cup rigid bladder biopsy of the lesion was performed. Pathology from the bladder dome lesion revealed a benign appearing growth consisting of cysts, smooth muscle, and ciliated epithelium (Figure 2, 3) which is consistent with urachal hamartoma.[5]

Despite the presumed benign histology of the mass, the patient was experiencing bothersome lower urinary tract symptoms refractory to anticholinergic medications. For this reason, after adequate patient counseling, we elected to proceed with a robotic-assisted laparoscopic partial cystectomy, including excision of the urachal remnant and associated mass. The patient was placed in low-lithotomy, steep Trendelenberg position and surgically prepped with chlorhexidine and alcohol. A six-port setup was utilized for the procedure, with a camera port at the umbilicus, three robotic arms and two assistant ports. We traditionally utilize monopolar shears in the right robotic arm and a PK dissector and Prograsp in the left two robotic arms. The Prograsp is useful for posterior and cephalic retraction on the urachus during dissection. The bladder was dissected from the anterior abdominal wall and the urachus was transected high on the abdomen. We did not completely excise the umbilicus as

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would be done for malignant pathology given the benign nature of this lesion. The urachus and associated mass was dissected towards the bladder until adequate mobility was achieved. A grossly benign margin of bladder mucosa was excised along with the lesion, and the bladder was closed in two layers. The operative time for this procedure was 2.5 hours and estimated blood loss was 100 mL. Surgical pathology confirmed the diagnosis of benign hamartoma.

The patient recovered uneventfully in the hospital and was discharged home on post-operative day one. She had a negative cystogram and then catheter removal 5 days following surgery. At three month follow up the patient was voiding well. Her lower urinary tract symptoms had completely resolved and she had returned to all her daily activities. No follow up imaging or surveillance is planned.

Discussion

Various syndromes are associated with the formation of hamartomas. Tuberous sclerosis complex leads to formation of hamartomas in a variety of sites, including the heart, brain, retina, and kidney.[6] Hamartomas have been associated with the Phosphatase and Tensin homolog (PTEN) gene. Germline mutations in PTEN have been described in a variety of rare syndromes that are collectively known as PTEN hamartoma tumor syndromes. Phenotypic manifestations of this mutation are, among others, Cowden syndrome and Bannayan-Riley-Ruvalcaba syndrome.[7]

Cowden syndrome shows autosomal dominant inheritance. It is characterized by hamartomas in a variety of tissues, as well as characteristic dermatologic manifestations, which include trichelemmomas, oral fibromas, and punctate palmoplantar keratosis. Genitourinary manifestations include uterine fibroids and ovarian cysts in the female and testicular lipomatosis in the male. Patients with Cowden syndrome also have an increased risk of breast, endometrial, thyroid, kidney, and colorectal cancers.[8] In Bannayan-Riley-Ruvalcaba syndrome, in addition to hamartomas, patients exhibit multiple subcutaneous lipomas and macrocephaly. Genitourinary manifestations include penile lentigines (pigmented macules).[9]

Partial cystectomy is becoming a more commonly performed procedure in the treatment of localized bladder lesions, including lesions with malignant pathology. A 2015 review by Knoedler and Frank[10] nicely outlined the published literature on partial cystectomies for localized muscle invasive urothelial carcinoma. As nicely explained in this article, proper patient selection is paramount when considering patients for partial cystectomy. The ideal patient is one with a solitary, anterior lesion without history of tumors in other locations. In this ideal patient, cancer specific survival and metastasis free survival have been shown to be comparable with radical cystectomy and are associated with significantly reduced complications in the partial cystectomy cohort.[11] Although most published series report results following open partial cystectomy, robotic approaches to urologic surgery are expanding are we feel comfortable performing a partial cystectomy with a robotic approach. Small series describing the robotic technique have been published with acceptable outcomes, albeit short follow up.[12]

While the pathology in our patient was benign, urachal adenocarcinoma is most appropriately treated with partial cystectomy and en block resection of the urachus.[13] With his in mind, we performed a similar procedure utilizing a minimally invasive approach with a few important differences. We omitted the obligatory umbilical resection as well as the pelvic lymph node dissection. We were confident in our transurethral biopsy and did not feel that the additional morbidity of those aspects of the typical cancer surgery was warranted. In general, robotic surgery has the advantage of reduced patient morbidity from a larger incision, faster return of bowel function and shorter time to hospital discharge. We have anecdotally found this to be true in our practice, although it inevitably comes with the price of higher overall health care costs.

Urachal hamartoma is an infrequently reported pathology. Although benign, they may be associated with significant irritative bladder symptoms. Robotic-assisted laparoscopic excision can offer a minimally invasive treatment with short convalescence and excellent functional bladder outcomes.

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