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URACHUS NEOPLASM: RARE ENTITY AMONG BLADDER ADENOCARCINOMAS - A CASE REPORT

Renanna Najara Veras Rodrigues
Hospital Universitário de Aracaju (HU-UFS)
Aracaju - Sergipe

José Vieira Barreto Júnior
Fundação de Beneficência Hospital de Cirurgia
Aracaju - Sergipe

Carlos Kleber Silva Alves
Hospital Universitário de Aracaju (HU-UFS)
Aracaju - Sergipe

Maria Luísa Barreto Paiva
Univesidade Tiradentes (UNIT)
Aracaju- Sergipe

Pamela Viana e Silva
Hospital Universitário de Aracaju (HU-UFS)
Aracaju - Sergipe

Italo Ruan Ribeiro Cruz
Fundação de Beneficência Hospital de Cirurgia
Aracaju - Sergipe

Karen Ito Tabata
Fundação de Beneficência Hospital de Cirurgia
Aracaju - Sergipe

Abstract: The urachus is an embryonic structure, originating from the allantois and cloaca, with the aim of facilitating the exchange of waste during development, which becomes obliterated throughout life. Their urachal remnants are uncommon congenital defects, with urachal carcinoma being a rare diagnostic entity. In this report, we discuss a 61-year-old patient who underwent resection of a urachal tumor, with a good outcome and good postoperative evolution. He is undergoing outpatient follow-up and there was no need for adjuvant treatment. It is important to discuss this rare pathology in order to formulate possible diagnostic hypotheses and treatment strategies.
Keywords: urachus; neoplasia, oncological surgery. (DCD - HEALTH)

INTRODUCTION

The urachus is an embryonic structure formed by a thick fibrous cord, originating from the allantois and cloaca, which connects the dome of the fetal bladder to the navel, with the aim of facilitating the exchange of waste during development. It is composed of layers of smooth muscle, submucosal connective tissue, and urothelial cell tissues. It is located in the pre-peritoneal space, with a tendency to progressively occlude as it develops, during the 12th week of gestation, obliterating itself, producing a fibromuscular channel called the median umbilical ligament. This structure may occasionally remain into adulthood due to failure of its obliteration. However, such malformations are rare and are sometimes underdiagnosed.(SHAO et al., 2022) (RYAN et al., 2023a);(TAKTAK; EL-TAJI; HANCHANALE, 2023).

CASE REPORT

Male patient, 61 years old, complaining of pain in the right inguinal region, of mild intensity and sudden onset, later associated with bladder tenesmus and urinary urgency. Denies any factor of improvement or worsening. He states that he initiated an investigation into the complaint due to the appearance of phlogistic signs in the same location after inguinal hernioplasty, 7 years ago, with a new approach to remove the mesh due to an inflammatory reaction 1 year ago. The physical examination only revealed a palpable mass in the hypogastrium, which was not painful on palpation. During research, he underwent magnetic resonance imaging of the abdominal region, which revealed a multiseptated cystic mass centered in the pelvic cavity (volume of 536.2 cm) without tumor extension to adjacent structures with fatty enlargement of the bilateral inguinal canal and slight thickening of the bladder wall. In addition, he provided a previously performed CT scan of the abdomen, which demonstrated an expansive supravesical formation, maintaining broad contact with the bladder roof, with an aspect that was undetermined according to the method. He denied constitutional symptoms. It presents as comorbidities hypertension and type 2 diabetes mellitus; no family history of cancer. In view of the condition, he underwent resection of Abdominal Wall Tumor + Partial cystectomy with cystorrhaphy (mass in close contact with the bladder roof), under general anesthesia, without complications. The material was sent to Pathology and immunohistochemical analysis, which revealed the finding of colloid-type urachal adenocarcinoma. He was discharged 24 hours after the procedure, using an indwelling urinary catheter, with good postoperative evolution. Probe removed in 14 days on an outpatient basis, without complications. The

patient therefore went to clinical follow-up, with no evidence of distant or local disease in the control exams (tomography and physical examination); according to clinical oncology, with no benefit from undergoing chemotherapy. He continues to undergo oncological monitoring, with a clinical and surgical team.

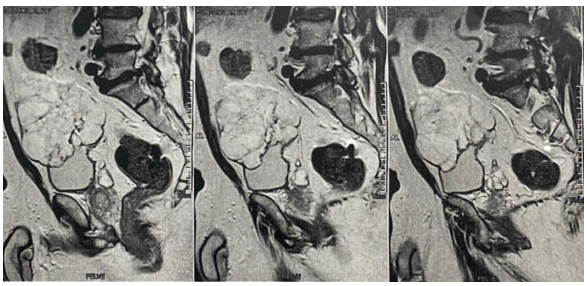

Image 1: Magnetic resonance imaging showing injury (Source: Personal collection).

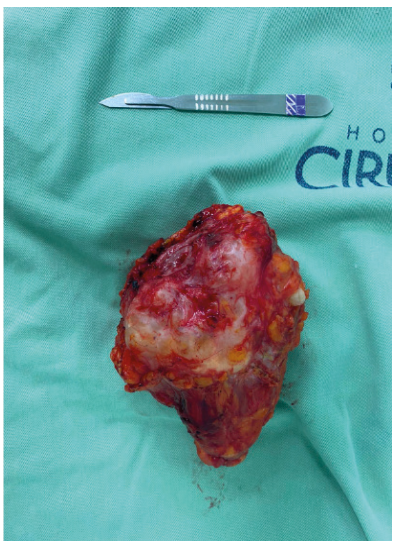

Image 2: Tumor resection product (Source: personal collection).

DISCUSSION

Urachal remnants are uncommon birth defects, with urachal carcinoma being a rare urological entity, accounting for only 0.35–0.7% of all bladder cancers.(SHAO et al., 2022)

This develops mainly across the midline from the dome of the bladder to the umbilicus, as well as in the retropubic space. (KUMAR et al., 2023). The pathology is more common during the 5th and 6th decades of life, frequently occurring in male patients, with a ratio of 5:1, and an incidence of 9.0% for men and 2.2% for women. Second REYES-ARROYO et al., 2021"Regarding the clinical presentation of urachal cancer, the symptoms are, in order of frequency, macroscopic hematuria (73%), poorly localized abdominal pain (14%), intermittent dysuria (13%), mucosuria (10%), irritative symptoms of the lower urinary tract (40%), palpable tumor in the lower abdomen (17%), bacteriuria (8%) and umbilical or bloody mucous secretion (2%)". Still in this regard, according to the Sheldon system, a classification method and the first staging system described that allowed a modern approach to the surgical intervention of these tumors, urachal cancer can be divided into 4 stages ranging from the neoplasm confined to the mucosa of the urachus to distant metastasis (TAKTAK; EL-TAJI; HANCHANALE, 2023).

Regarding diagnosis, it is possible to use cytology, imaging and cystoscopy. Radiographic images with computed tomography provide broad support in surgical planning. On CT, adenocarcinoma may appear solid, cystic or a combination of the two, while on Magnetic Resonance Imaging, the tumor is reported as heterogeneous, with high intensity on T2 (Canadian Association). Due to the rarity of this neoplasm and the non-specificity of symptoms, diagnosis tends to be late, resulting in a poor prognosis. Another point that makes a conclusive diagnosis difficult is its similarity to adenocarcinoma of various origins in terms of molecular biology and pathology. Immunohistochemistry can be of great value for identifying urachal carcinoma, since the majority of cases are positive for the biomarkers CK20 and CDX2; CK7 and β-catenin are expressed at a lower frequency (KUMAR et al., 2023). However, according to the Canadian Urological Association (2019),

there are no specific markers to confirm or refute urachal cancer, as its cells show great similarity to the gastric and colonic mucosa.

Therapeutic strategies tend to vary according to the clinical stage at the time of diagnosis, with the treatment of choice for non-metastatic cancers being the surgical approach, in which partial cystectomy with removal of the urachal ligament is most commonly performed, as it is not possible to relate the increased survival time with an approach in which radical cystectomy is performed. Another option described is partial or radical cystectomy plus omphalectomy and bilateral pelvic lymphadenectomy, taking into account that radical prostatectomy must be performed in men(TAKTAK; EL-TAJI; HANCHANALE, 2023);RYAN et al., 2023). However, Ashley et al. conducted a study in which records of 130 patients with urachal masses were analyzed, 66 of whom were diagnosed with urachal carcinoma, treated at the Mayo Clinic between the years 1951 and 2004, which demonstrated a lack of therapeutic benefit in relation to adjuvant therapy (chemotherapy, radiation and both) or regional lymphadenectomy. Furthermore, in the aforementioned study, although with a small sample, local recurrences were noticed in 15% of patients (approximately 6 months after surgery). Of the group of patients who underwent repeated surgical resection, 67% of them were cured within 15 years (4 of 6), while radiotherapy did not save any (0 of 2). Such data prove that the roles of radiotherapy and chemotherapy are still unclear.

Furthermore, urachal adenocarcinomas tend to have a worse prognosis when compared to non-urachal adenocarcinomas, and may metastasize early to the inguinal iliac lymph nodes, omentum, liver, lungs and bones (Fundamental Urology, book). The main factors to be evaluated include: stage of the disease, status of the surgical margin, pathological grade of the tumor, presence of positive lymph nodes and type of surgery, with a high stage being associated with a poor prognosis.

CONCLUSION

As urachal carcinoma is a rare pathology among urological neoplasms, and in view of the poor prognosis of the disease in question, better knowledge of the disease is necessary to enter into differential diagnoses during oncological investigations, in addition to better monitoring of patients affected by the disease, as well as new prospective and multicenter studies in order to establish more modern and effective treatment protocols.

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