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gacetamedicaboliviana@gmail.com
Universidad Mayor de San Simón
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Vargas-Rocha, Vladimir E.; Segales-Rojas, Patricia

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Laparoscopic resection of urachal adenocarcinoma

Vladimir E. Vargas-Rocha Vladimir.vargas.r@gmail.com
Caja Petrolera de Salud, Bolivia

 <https://orcid.org/0000-0002-4455-8773>

Patricia Segales-Rojas
Bolivian-Japanese Gastroenterological Institute, Bolivia

 <https://orcid.org/0000-0002-3578-6678>

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Abstract: Urachal adenocarcinoma is a rare neoplastic disease, with an incidence of 1 per 5 million inhabitants. It manifests clinically with hematuria (73%), abdominal pain (14%), dysuria (13%), mucosuria (10%), irritative symptoms (40%), palpable mass in the lower abdomen (17%), bacteriuria (8%), umbilical mucous discharge (2%). We present the case of a 45-year-old patient, diagnosed with urachal adenocarcinoma, who received multiple treatments without response, undergoing radical surgery with en bloc resection of the umbilicus, urachus, peritoneum, posterior fascia of the rectus abdominis muscle and partial cystectomy, by laparoscopic approach, with excellent oncologic and aesthetic results, with rapid recovery and without complications. The role of chemotherapy and radiotherapy and its benefit for the patient is still not clear. We wish to contribute with a new case to the literature, in addition to being able to show that minimally invasive management can be adequate in expert hands, with results equal to open surgery, with the already known benefit of the laparoscopic approach.

Keywords: urachus, carcinoma, bladder neoplasms.

Resumen: El adenocarcinoma de uraco es una enfermedad neoplásica rara, con una incidencia de 1 por 5 millones de habitantes. Clínicamente se manifiesta con hematuria (73%), dolor abdominal (14%), disuria (13%), mucosuria (10%), síntomas irritativos (40%), masa palpable en la parte baja del abdomen (17%), bacteriuria (8%), flujo mucoso umbilical (2%). Presentamos el caso de paciente de 45 años, con diagnóstico de adenocarcinoma de uraco, que recibió múltiples tratamientos sin respuesta, realizándose, cirugía radical con resección en bloque de ombligo, uraco, peritoneo, fascia posterior del musculo recto del abdomen y cistectomía parcial, por abordaje laparoscópico, con excelentes resultados oncológicos y estéticos, con recuperación rápida, y sin complicaciones. El papel de la quimioterapia y radioterapia y su beneficio para el paciente aún no está claro. Con este caso queremos aportar con un nuevo caso a la literatura, además de poder mostrar que el manejo con mínima invasión puede ser adecuada en manos expertas, con resultados iguales a la cirugía abierta, con el beneficio ya conocido del abordaje laparoscópico.

Palabras clave: uraco, carcinoma, tumores de vejiga.

The urachus, also known as the mid umbilical ligament, is a midline structure that extends from the bladder dome to the umbilical scar, it is 5 to 10 cm long, it is an embryonic remnant from the allantois, between the 4th and 7th week of life the cloaca is divided by the uro-rectal septum into two structures: the ano-rectal duct which is located posteriorly and the urogenital sinus which is located anteriorly. The cephalic extension

of the urogenital sinus, which is the precursor of the fetal bladder, communicates with the allantois, which is derived from the yolk sac, at the level of the umbilicus ¹. The epithelial cells that constitute these layers give rise to tumors like urachal adenocarcinoma. ² It is known that just before birth this duct is completely occluded and that it will become a fibrous cord in the adult, known as the median umbilical ligament ³.

This urachal remnant can give rise to various clinical pathologies not only in children but also in adults. Diseases that affect it are uncommon and its abdominal manifestation, symptoms or urinary signs are unspecific, therefore the diagnosis is not always easy.

Failure during closure of the urachal lumen causes different types of anomalies: congenital permeable urachus, urachal fistula, uracovesical diverticulum, urachal cyst, all malignant tumors of the urachus are very rare and there are few reports worldwide and differential diagnosis with vesical dome adenocarcinoma should be considered.

Urachal adenocarcinoma is a malignant neoplasm that corresponds to only 0.7% of malignant bladder tumors and 35% of bladder adenocarcinomas. ⁴ This tumor is more frequent in men with a ratio of 5:1, ⁵⁻⁸ with a predominance of presentation between the 5th to 6th decade of life ^{6,7}.

Case presentation

A 45-year-old female patient, started her condition 6 years ago, with several urinary tract infection episodes, receiving multiple courses of antibiotics, with partial improvement, later accompanied by hematuria, mucosuria, and pain in the hypogastrium, with periumbilical pruritus and sensation of vesical oppression, with severe low irritative urinary symptoms, urinary urgency, tenesmus, pyuria and dysuria, For this reason, a control ultrasound was performed in a private environment, diagnosing a vesical tumor, so it was decided to perform a transurethral resection, in a private setting, with a histopathological report of bladder adenocarcinoma, without other data, completing the study protocol with a hired abdominopelvic tomography, in which a tumor in the bladder dome was found, which infiltrated the anterior abdominal wall, being subsequently treated with 3 cycles of chemotherapy, with unknown schedule and duration, without response to management, Afterwards, the patient underwent a 3 cycle chemotherapy course, the schedule and duration are unknown, with no response, having presented for 2 years abundant purulent discharge through the umbilicus (see image 1).



Figure 1.

Fistula in the umbilicus, with outflow of purulent material from urachal tumor.

own elaboration

With cures and drains without improvement, later presenting difficulty in urination and copious mucosuria, the patient attended the clinic for treatment. Initially a contrasted thoraco-abdominal-pelvic tomography with elimination phase was performed (see image 2).

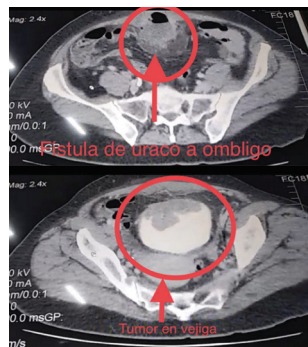


Figura 2.

Axial abdominopelvic tomography with contrast, urachus tumor draining into the umbilicus and invading the bladder.

own elaboration

Documenting a tumor involving the roof of the bladder, anterior wall of the rectus abdominis muscle and fistulizing to the umbilicus, without lymph nodes or metastasis in other sites, A control cystoscopy was performed (see image 3).

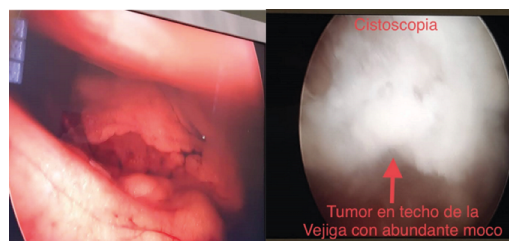


Figure 3.

Control cystoscopy, showing evidence of bladder dome tumor, with abundant mucus and cellular detritus, involving a large part of the bladder roof.

own elaboration

Showing evidence of tumor involving the roof and dome of the bladder in its entirety, with abundant mucus secretion, friable tissue

and easy bleeding, with sediment throughout the bladder, along the lateral walls, floor, neck and retrocollar with no evidence of tumor activity, with both orthotopic ureteral meatuses excreting clear urine. Based on these findings, it was decided to perform radical surgery, by radical umbilectomy, with en bloc resection of the urachus, parietal peritoneum, posterior aponeurosis of the rectus abdominis muscle, wide partial cystectomy and laparoscopic extended pelvic lymphadenectomy, the procedure was performed by a minimally invasive approach, by means of 4 laparoscopic ports (see image 4).



Figure 4.

Patient position, in forced trendelenburg at 45°, 4 trocars were placed, with the leg in abduction.
own elaboration

2 trocars of 10 mm and 2 trocars of 5 mm, with a bleeding of 200 ml, surgical time of 2 hrs 30 min, the procedure was performed without complications, the piece was extracted through the umbilicus, after resecting the entirety of it, and the patient was discharged after 48 hrs, with a tube for 10 days, with adequate oral tolerance. The catheter and stitches were removed 10 days later, and a control bladder ultrasound was performed one month after the surgery, resulting in a bladder without alterations and a vesical capacity of 430 ml.

The histopathological report of the specimen was: mucinous adenocarcinoma, mucoproduktive type of the urachus with extension into the muscularis propria and mucosa of the bladder wall, tumor size 10 x 7 x 2 cm, surgical margin of the bladder 1 cm from the tumor, umbilical skin with fistulous tract with foci of suppurative necrosis and granulation tissue (see image 5).

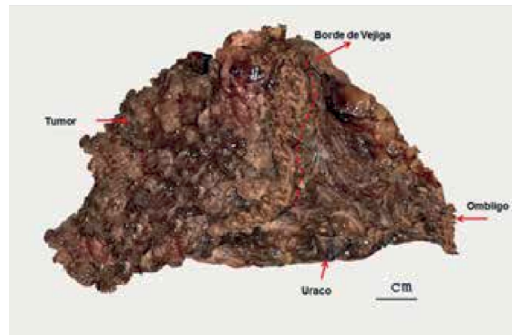


Figure 5.

Surgical specimen, complete tumor, with free bladder border and tumor.

own elaboration

Without evidence of neoplastic cells, pelvic lymph nodes negative for neoplastic cells. The patient is currently under follow-up with good evolution, without data of tumor recurrence, with well healed wounds (see image 6). A control cystoscopy was performed 3 months after surgery, with findings of bladder with fundus, lateral walls, floor, neck and posterior neck without signs of tumoral activity, with no alterations or recurrence findings in the study.



Figure 6

Healed wounds in controls, no signs of recurrence.

own elaboration

Discussion

In relation to urachal pathology, they can be divided into two: congenital and acquired. Among those of congenital origin is permeable urachus, which consists in the persistence of the communication between the dome of the bladder and the umbilicus, clinically characterized by urine leakage through the umbilicus since birth. Other acquired urachal anomalies are characterized by partial reopening of the urachal lumen: These include 1) urachal fistula, which is the presence of a cyst communicating with the umbilicus; 2) uracovesical diverticulum, which is a cystic structure communicating with the bladder; 3) urachal cyst, which does not communicate with either the bladder or the umbilicus; 4) alternating fistula, which is the presence of a urachal cyst that drains

intermittently into the bladder or the umbilicus^{9,12,13}. These pathologies can appear at any age, but they usually occur during childhood¹¹.

Due to the location and accessibility of the urachus, which is located in the anterior wall of the abdomen, it is far from the interference of intestinal gases, for this reason ultrasound is a very accepted technique for a very good diagnostic approach^{10,14}, but tomography is the imaging test that provides greater accuracy, both for the visualization and delimitation of the alterations that can be found in the urachus, it is also very useful for the study of adjacent structures, allowing differential diagnosis with other pathologies^{15,16}.

Surgery, with complete resection of the urachus is the most appropriate treatment⁴. It is known that in the hands of expert urologists and with selected patients, a laparoscopic surgical approach can be chosen, thus providing a shorter hospital stay and rapid recovery^{17,18}. As in our case, a minimally invasive approach was used, with excellent oncologic and esthetic results in the patient, with a rapid recovery to her normal activities, this case being the first report in relation to this type of management in our country.

Diagnosis is sometimes difficult and therefore delayed, due to the scarce initial clinical manifestations. Clinically it can manifest with hematuria(73%), abdominal pain (14%), dysuria (13%), mucosuria(10%)³, irritative symptoms(40%), palpable mass in the lower abdomen(17%), bacteriuria(8%), umbilical mucous discharge(2%)^{12,13}. The diagnosis can be reached by a similar route to that used for bladder tumors: cystoscopy and transurethral resection are fundamental in the diagnosis. Abdomino-pelvic plain radiography frequently reveals calcifications. CT and MRI are fundamental when planning the approach for surgical treatment, since they provide an idea of the relationship between the tumor and adjacent structures in terms of their possible involvement, or the presence of lymph nodes²¹.

Based on the architecture, arrangement and mucin content of urachal carcinoma cells, urachal carcinomas are pathologically subdivided into tubular, papillary, mucinous, colloid and signet ring adenocarcinomas. The pathologic features of the tumor are the variables that best predict its survival and aggressiveness. Based on these data, mucin-predominant tumors have a better prognosis than papillary, tubular or signet ring tumors, which have an adverse and aggressive prognosis²².

The differential diagnosis between adenocarcinoma of urachal origin and adenocarcinoma of colon origin that metastasizes to the urachus, in those cases where there is doubt, can be made with immunohistochemical techniques. An immunohistochemical profile of CK7+ and CK20+, points to a transitional and urological origin, as opposed to a colonic origin, which usually presents a CK7- pattern²³.

Sheldon et al²⁴ in his paper makes a stage classification of urachal adenocarcinomas: Stage I: refers to a tumor that is located in the mucosa of the urachus; Stage II: tumor that invades the submucosa or the muscular layer of the urachus, but is located in the urachus;

Stage III: tumor extending outside the urachus, IIIA extending into the bladder, IIIB extending into the abdominal wall, IIIC extending into the peritoneum and IIID extending into some other viscera; Stage IV: a tumor with distant metastasis.

The treatment of urachal tumors is partial cystectomy, with en bloc resection of the urachal tissues, from the bladder to the umbilicus, of the posterior lamina of the sheath of the rectus abdominis muscles, as well as extended bilateral pelvic lymphadenectomy ²⁵⁻²⁷.

It should be noted that the prognosis is not good, because most patients are diagnosed in advanced stages. Between 43 and 50% of patients survive 5 years after surgery. These tumors are considered radioresistant and present dubious and poor chemosensitivity ²⁸.

Adjuvant polychemotherapy does not improve the results either. The most commonly used chemotherapy regimens are the reference ones for transitional cell bladder cancer, FAM, 5-fluorouracil, Doxorubicin and Mitomycin C, CISCA, Cisplatin, Cyclophosphamide and Doxorubicin, modified FAM, which substitutes Adriamycin for Mitoxantrone and M-VAC, Methotrexate, Vinblastine, Adriamycin and Cisplatin. Recently, good results were reported in urachal metastatic adenocarcinomas, associating surgery, radiotherapy and polychemotherapy, with disease-free periods of 10 years ^{29,30,31}.

The organs most frequently affected by metastasis are the lung (50%), regional lymph nodes (46%), bone, predominantly the spine (30%), intestine (30%), brain (20%) and liver (16%) ³².

Conclusion

Urachal carcinoma is a rare pathology and is usually diagnosed in advanced stages. For initial management, a wide surgical resection of the tumor mass is indicated with a high risk of recurrence and distant metastasis in the first year, which decreases survival at five years, so adjuvant methods such as radiotherapy and chemotherapy are used to improve life quality.

The literature on urachal adenocarcinomas has not found a relationship between the size and prognosis of the disease. This association appears when a relationship is made with the pathologic stage, especially in pT3c tumors. It is important to note that with respect to management, the results are still inconclusive and are divided between partial and radical cystectomy, both with good results in the short term, but poor in the long term.

In our case it is a patient without comorbidities, with a tumor stage pT3a our case was managed with partial cystectomy, with adequate clinical response. with adequate clinical response. The patient does not require adjuvant surgery for the moment and should continue to be followed up and monitored for local or systemic recurrence, with cystoscopy and colonoscopy controls.

Currently, there is no standard protocol for the treatment of urachal adenocarcinoma with adjuvant chemotherapy. Chemotherapy and radiotherapy and their benefit for the patient is still unclear.

Our aim with this case is to contribute to the literature, besides being able to show that minimally invasive management can be adequate in expert hands, with results equal to those of open surgery, with the already known benefit of the laparoscopic approach.

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