and before resorting to more complex options such as fluoroscopy, laparoscopy or open surgical exploration (7.8).

REFERENCES AND RECOMMENDED READINGS (*of special interest, **of outstanding interest)


URACHAL ADENOCARCINOMA. CASE REPORT AND BIBLIOGRAPHIC REVIEW

Gabriel Ogaya Pinies, Felipe Herranz Amo, Gregorio Escribano Patiño, Enrique Lledó García, Roberto Molina Escudero, Adrian Husillos Alonso and Carlos Hernandez Fernandez.


Summary.- OBJECTIVE: Literature review of adenocarcinoma of the urachus in connection with two cases recently diagnosed and treated in our center.

METHODS/RESULTS: We report 2 cases of urachus Adenocarcinoma treated in our institution, both underwent extended partial cystectomy including excision of the urachus up to the umbilicus.

CONCLUSION: Urachal adenocarcinoma is an exceptional tumor, of poor prognosis, the treatment of which is surgical.

CORRESPONDENCE

Gabriel Ogaya Pinies
Servicio de Urología
Hospital General Universitario Gregorio Marañón
Doctor Esquerdo, 46
28007 Madrid (Spain)
gabrielogaya@gmail.com.

Accepted for publication: September 13th, 2010
Resumen.- OBJETIVO: Revisión de la literatura del Adenocarcinoma de uraco a propósito de 2 casos diagnosticados y tratados recientemente en nuestro centro.

MÉTODOS/RESULTADOS: Describimos 2 casos de adenocarcinoma de uraco tratados en nuestro, ambos sometidos con cistectomía parcial extensa incluyendo resección de el uraco hasta el ombligo.

CONCLUSIONES: El adenocarcinoma de uraco es un tumor excepcional, de mal pronóstico, cuyo tratamiento recae fundamentalmente en la cirugía (cistectomía parcial), y donde los principales factores pronóstico de supervivencia libre de enfermedad son el grado de diferenciación tumoral y los márgenes libres de la pieza quirúrgica.

Palabras clave: Uraco. Adenocarcinoma de uraco.

INTRODUCTION

Urachus or median umbilical ligament, is a tubular structure located in the mid line and extends from the umbilicus to the bladder’s dome. It’s the remaining of two embryological structures: the cloaca, which is the cephalic extension of the urogenital sinus (forerunner of fetus’s bladder) and the allantoids. It has three histological layers: transitional or cuboidal epithelium, submucosal connective tissue and the most external portion (represented by the muscular tissue) (1,2).

Adenocarcinoma of the bladder is a rare malignancy with poor prognosis, representing 0.2-0.5% of bladder tumors. The mucosa of the bladder is covered with transitional cells in absence of glandular epithelium. The most accepted theories in order to explains the presence of adenocarcinoma in an organ that do not contains glandular tissue are:

1) The metaplastic changes from urothelium to mucinous epithelium or glandular and

2) The persistence of intestinal endoderm tissue (3).

The annual incidence of urachus carcinoma is 1 in 5 million people in the general population or 0.01% among all adult cancer types (4). In Spanish literature there are only 18 reported cases of urachal adenocarcinoma, 5 of them published in the last 10 years (2,7,11,12).

Case 1

43 years old male, referred from primary care physician, because an isolated episode of monosymptomatic hematuria. On abdominal ultrasound a bladder mass of 45x45x25mm was found. The abdominopelvic CT scan shows endoluminal bladder mass with exophytic growth, hypodense center 45 x50 x 30 mm, without increased size lymph nodes, no sign of metastatic disease. The cystoscopy showed the imprint of a body of probable extrinsic origin of about 40mm in diameter, located in the bottom-front of the bladder.

Partial cystectomy was performed which included extensive excision of the urachus towards the umbilicus, with bilateral iliobturatriz lymphadenectomy.

The histological analysis of the specimen showed a moderately differentiated intestinal-type adenocarcinoma, without infiltration of resected lymph nodes (pT3N0M0).

Case 2

36 years old male, referred to the urology clinic for evaluation of episodes of recurrent hematuria and abdominal fullness.

Ultrasound was performed as part of the hematuria study, showing: Normal size and parenchyma on both kidneys, distended bladder with hypoechoic nodular structure of 40mm in diameter, with well defined and sharp edges. Abdominopelvic CT scan was performed, which shows a solid tumor of 35 mm in diameter

![FIGURE 1. Sagital CT scan. Demonstrate mass at the anterior superior portion of the bladder with endoluminal and exophytic growth, extending up to the umbilicus.](image-url)
attached to the anterosuperior aspect of the bladder that is extended up to the umbilical region, over a distance of 52mm. This lesion had an intravesical growth with polypoid excrescence.

The patient underwent extensive partial cystectomy, including the urachus to the umbilicus, plus bilateral lymphadenectomy, with an uneventful postoperative.

The pathological study showed a moderately differentiated adenocarcinoma of the urachus, infiltrating extrinsically the peritoneal lining of the bladder, but not involving the muscular layer. The surgical margins of resection were free of tumor. Lymph nodes were negative.

DISCUSSION

Most tumors coming from the bladder are transitional cells carcinomas, been Urachus carcinoma a rare aggressive neoplasia. In the literature the mainstream of published series contains less than 10 cases, and only few of those contain enough cases in order to perform statistical analysis (5).

The Urachus adenocarcinomas are mostly located at the bladder dome. This type of tumors generally escapes from clinical detection, allowing them to grow for long periods of time before the diagnosis, facilitating local invasion and systemic spread, before any therapeutic intervention (6).

In the vast majority of the series published, hematuria is the most common symptom, been present in 80%of patients at the time of diagnosis, followed by lower urinary track symptoms. The mucosuria is found between 9% and 15% of patients, and its presence should arouse immediate suspicion for urachus carcinoma (5-7).

Regarding gender distribution, there is not an agreement on the literature. The larger series reported a 1:1 male-female distribution vs. the one observed by Ashley et al. of 2.2:1 male-female (6). It usually occurs in patients under 60 years of age, as in Mostofi et al (8), where they observed a mean age of 45.8 years old and Grignon et al of 51.5 years old (5). However, neither sex nor age have proven a factor in predicting survival (6,9).

The imaging test can also be used to facilitate the diagnosis of urachal tumors. Urachal malignant tumors arise in 90% of cases at the juxtavesical portion, and extends up to the umbilicus and inferiorly through the bladder. Urachal carcinoma may be solid, cystic or a combination of both, showing low attenuation on CT scan in 60% of cases, reflecting its mucinous component. Peripheral punctate or curvilinear calcification is observed in 50% - 70% of cases. The presence of a caudal part of the tumor involving the bladder wall, and a cystic encapsulated supravesical portion, are considered highly characteristic findings on CT scan (10).

Urachal tumors can be divided into five histological subtypes (5):

- *Intestinal type*, when its architecture reminiscent of colon adenocarcinoma.
- *Mucinous type*, when the tumor is characterized by a cell or group of cells in a matrix of extracellular mucin.
- *Signet ring type*, when tumor cells spread diffusely through the tissue.
- *Unspecified adenocarcinoma*, when the pattern does not fit into any category
- *Mixed type*, when the tumor shows two or more of any of these patterns and none of those represents more than 75% of the material submitted.

In cases where diagnostic doubts exist about whether the source is colonic or urachal adenocarcinoma, immunohistochemical techniques may be used, as CK7 and CK20 positive profile point to a transitional urologic origin, while generally, a negative CK7 profile, usually is more oriented to a colonic source 11.12.

Sheldon (4) made a staging classification of adenocarcinoma of the urachus:

- *Stage I*: Tumor is located in the urachal mucosa;
- *Stage II*: Tumor invades the submucosa or muscular layer of the urachus but is limited to the urachus;
- *Stage III*: Tumor extends outside of the urachus, IIIA to the bladder, IIIB to the abdominal wall, IIIC to the peritoneum and IIID to some different viscera;

![FIGURE 2. Axial CT scan. Solid-looking lesion on anterior bladder, with exophytic growth.](image)
Stage IV: tumor with distant metastases. Extended partial cystectomy including the umbilicus and urachus with peritoneum, is considered as the ultimate treatment of this entity. In the other hand lymphadenectomy, chemotherapy or radiotherapy have failed to proof benefits in terms of patients survival with urachal adenocarcinoma (13).

The cancer-specific overall survival at 5 years is ranged between 26% -49% (5,6). Prognostic factors for cancer-specific survival with statistical significance are: the degree of dysplasia and surgical margins, this according to Ashley et al (6) who were the first to perform a multivariable analysis on prognostic variables. In their study showed that there is a relative risk (RR) of death 3.8-specific cancer when surgical margins were positive (p <0.001) and a RR of 3.7 when the tumor was high grade (p <0.001).

In the largest series published, local recurrence occurred in 15% of patients, especially during the first 6 months after surgery. The surgical re-intervention in these cases resulted in a survival of 67% of patients at 15 years. Distant metastases were detected in 59% of patients at some point during the follow-up, of these, 90% died of the disease in about 1 year (6).

CONCLUSIONS

Urachal adenocarcinoma is an exceptional tumor, of poor prognosis, whose treatment lies with surgery (extended partial cystectomy with umbilicus and urachus with peritoneum), and where the main predictors of disease-free survival are the degree of tumor differentiation and the free margins of the surgical specimen.

REFERENCES AND RECOMMENDED READINGS

(*of special interest, **of outstanding interest)

Sheldon CA, Clayman RV, Gonzalez R, Williams
Ashley RA. Urachal Carcinoma: Clinicopathologic