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Atypical Presentation of an Urachal Carcinoma as an Urachal-Colonic Fistula: A Rare Case Report

Abdelhamid Garmane[1*], A. Chehboun[1], A. El Hajjami[1], B. Boutakioute[1], M. Ouali Idrissi[1], N. Cherif Idrissi El Ganouni[1]

[1]Radiology Departement, AR-RAZI Hospital, CHU Mohamed VI, Cadi Ayad University Marrakech, Morroco

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***Corresponding author:** Abdelhamid Garmane
Radiology Departement, AR-RAZI Hospital, CHU Mohamed VI, Cadi Ayad University Marrakech, Morroco

Abstract | **Case Report**

Urachal carcinoma is a rare and aggressive tumor, involving the urachus and the bladder. Symptoms of urachal carcinoma usually appear at later stages of the disease; therefore, these tumors are diagnosed in advanced stages, providing limited options for curative treatment. We report the clinical case of a 39-year-old man with a urachal carcinoma which presented as a mass of the abdominal wall invading the transverse colon, creating an enterocutaneous fistula.

Keywords: Urachal tumor, Colo-Urachal fistula, CT.

INTRODUCTION

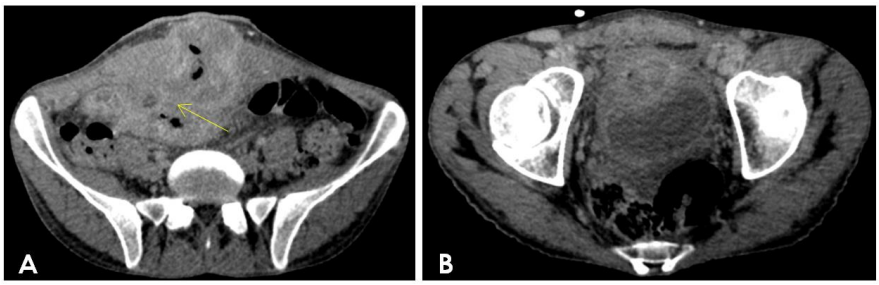
Carcinoma of the urachus is a rare and very aggressive tumor that affects the urachus and bladder. Because it is an extremely rare cancer, it is often misdiagnosed, making it difficult to determine its true incidence in the general population, It accounts for less than 1% of all bladder cancers [1, 2] and 0.01% of all cancers [3]. Symptoms usually appear late and the diagnosis is often made at an advanced stage of the disease [4, 5]. We present an atypical case of Urachal carcinoma and a review of the most relevant literature.

CASE REPORT

We report a case of a 39-year-old man, with no medical history, who presented with a growing anterior abdominal mass that extend from the ombilic to the pelvic region that is median with purulent secretions and digestive fluid, in a context of fever and weight loss.

An abdominal ultrasound showed a large heterogeneous mass extending between the antero-superior wall of the bladder and the umbilicus, with a heterogeneous and largely necrotic echostructure and mixed vascularization on color Doppler, associated with a cutaneous fistula.

A CT scan was performed confirming the ultrasound aspect, showing a voluminous infiltrative mass developed from the bladder that has an irregular thickening, extending to the umbilical region, with heterogenous enhancement after contrast injection, and individualization of an abcess with cutaneous fistula (Figure 1) associed with an urachal-transverse colon fistula (figure 1).



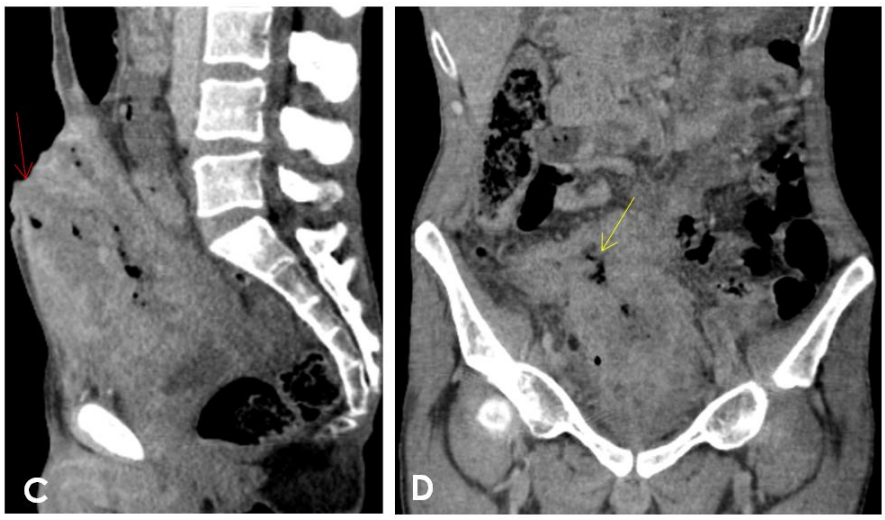


Figure 1: Axial (A, B), sagital (C) and coronal (D) CT sections on portal veinous injection demonstrating an heterogenous tumor between the umbilicus and the bladder protruding and infiltrating the skin surface of the peri-umbilical region, with cutaneous fistula (red arrow), and infiltration of the transverse colon with colo-Urachal fistula (yellow arrow)

A per cutaneous biopsy was performed confirming an urachal carcinoma. There were no detected metastatic sites on the chest, abdomen and pelvis scan.

The patient died before the operation due to septic shock.

DISCUSSION

Urachal cancer is a rare form of bladder cancer that arises from the urachus, a vestigial musculofibrous band that extends from the dome of the bladder to the umbilicus [8]. They were originally described by Hue and Jacquin in 1863. They account for only 0.5% of all bladder cancers, and 20% to 40% of primary bladder adenocarcinomas [9].

The most common histologic subtype is the adenocarcinoma with enteric features, with or without mucin production [1]. Approximately 70% of urachal adenocarcinomas are mucin-producing tumors [4, 5, 7]. Other rarer types are: sarcomas, transitional cell carcinomas, squamous cell carcinomas.

The criteria for the diagnosis of urachal carcinoma were defined by Sheldon *et al.*, and Mostofi *et al.*, and revised by Gopalan *et al.*,

These included a dome tumor or elsewhere in the midline of the bladder, absence of cystitis cystic and cystitis glandularis, a sharp demarcation between the tumor and normal surface epithelium, enteric-type histology, and absence of a primary neoplasm elsewhere [3, 6, 12, 13]. These criteria were further adapted and published by the World Health Organization [14].

Few cases are reported in literature making it difficult to determine the prognostic of these cancers. Because early urachal cancer is not accompanied with symptoms, patients often present at the time of diagnosis with higher stage and poor prognosis, the most common symptom is hematuria [10]. In our case, it was the suprapubic mass with cutaneous fistula with purulent secretions and digestive fluid that revealed the tumor.

Imaging plays also an invaluable role at urachal cancer workup. On Ultrasound (US), it is commonly recognized as a midline soft tissue mass or a fluid-filled cavity with mixed echogenicity and calcifications. The CT scan is decisive in making a positive diagnosis of the tumor, specifying its solid, cystic or mixed nature, the presence of calcifications which are generally peripheral but may also be central or both. It specifies bladder invasion (present in the majority of cases), locoregional lymph node invasion or metastatic invasion. It allows guided biopsies to be performed and to detect possible recurrences after treatment [15]. Magnetic resonance imaging (MRI) does not provide any additional morphological information compared to CT. It can be useful in case of contraindication to the iodinated contrast agent or if there is a diagnostic doubt [15]. It is usually detected as a midline mass, superior to the bladder dome and adjacent to the abdominal wall. In the majority of cases, the tumor is mixed solid and cystic, the latter representing its mucin composition.

Urachal-transverse colon fistula is extremely rare, reported in only few cases.

The gold standard surgical approach for the management of localized urachal cancer is an excision of the urachus, umbilicus, and cystectomy combined with bilateral pelvic lymphadenectomy [11].

CONCLUSION

Urachal carcinoma is a rare and aggressive tumor, transverse colon fistula is even less common as we believe. The diagnosis is based on imaging especially on CT scan showing the tumor and its extension. Proper surgical intervention has proven critical to the survival of patients.

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