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Urachal Cancer in a 47-Year-Old Patient: A Case Report and Literature Review

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Authors' contributions

This work was carried out in collaboration between all authors. Author CC designed the case report and written the case report. Author OB collected the data. Author TTK is pathologist and author OGD written the case and managed the literature searches. All authors read and approved the final manuscript.

Case Report

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ABSTRACT

Aims: Urachal cancer is an uncommon neoplasm associated with poor prognosis. No concensus has been reached regarding diagnostic criteria so far. The management of urachal carcinoma is controversial, too. In this case presentation, we reported a 47 year old female with urachal cancer and treatment approach discussed with published literature.
Presentation of Case: A 47-year-old female patient was admitted with abdominal pain and hematuria. A soft mass was noticed under her navel. Computerized tomography revealed a tumor which is a cystic lesion arising from the urachus and a solid mass component at the urinary bladder dome. The tumor was removed by partial cycectomy. Histological examination showed urachal adenocarcinoma (colonic type and well differentiated), which had invaded the urinary bladder. The patient has been followed up without a recurrence for the next 6 months.
Discussion: Urachal carcinoma is a rare type of, approximately 0.5 to 2%, bladder neoplasms. The pathogenesis of urachal tumours is not fully understood, so far. Currently, the most effective treatment of localized urachal cancer is cystectomy. It is

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unclear whether lymphadenectomy or chemotherapy is usefull for the patients with urachal carcinoma.
Conclusion: Regarding the treatment of urachal cancer, surgery remains the mainstay of therapy. Moreover, the achievement of a complete urachectomy, including umbilectomy and negative surgical margins, is critical for low-stage, localized, well-differentiated urachal cancer.

Keywords: Cystectomy; therapy; urachal cancer; urinary bladder.

1. INTRODUCTION

Urachal cancer is a type of rare tumors. One of the most common presenting symptoms is hematuria (82%). The most prevalent diagnostic test is cystoscopy (100%) (Ashley et al., 2006). The available literature on urachal carcinoma consists of small case series. Five-year survival rate is approximately 43% for urachal cancer (Henly et al., 1993).

They are most frequently seen at the junction of urachal ligament and bladder dome (Sekita et al., 2010). They form 0.5 to 2% of malignant bladder tumors and observed at early ages in proportion to other epithelial carcinomas (Henly et al., 1993). They are most frequently observed as enteric type adenocarcinoma and developed from urachal enteric cell residuals or as a result of metaplasia of urachal ligament during embryonal development (Bratu et al., 2009). They can lead to hematuria, irritative urination symptoms and suprapubic mass by forming a mass on the bladder dome. Localization on bladder dome, to be a sharp demarcation line between tumor and bladder urothelium without any adenocarcinoma in the body to metastasize into the bladder which are important clinicopathologic properties for the diagnosis (Girgin et al., 2003). Currently, there is no consensus on treatments of urachal cancers. In this case presentation, clinic, radiologic, histologic properties of urachal cancer in a young women was presented and treatment approach discussed by accompanying literature.

2. CASE PRESENTATION

A transabdominal ultrasonography (USG) was performed for a 47-year-old female patient at an external center, who experienced several times macroscopic, terminal hematuria without clot for six months, and a mass was detected on the bladder dome. Lobulated mass hanging down from bladder dome towards into the bladder was monitored in the contrast-enhanced tomography at the same external center (Fig. 1). Then, the patient applied to our clinic for further assessment and treatment. The patient had no history of smoking. Family history was unremarkable. The case has no pathologic finding by the physical examination. The chest radiography and urinary cytology were normal. Transurethral resection (TUR) was performed for solid tumoral formation on bladder dome, which was 3x3 cm and including necrotic tissue and cotton wool mucoid material. Middle of tumor showed a crater during cytoscopy performed under general anesthesia. A mobile abdominal palpable mass with 3x3 cm in diameter was present before TUR. And this mass persisted as a smaller mass after TUR by bimanual examination under anesthesia. During TUR, we received randomly bladder biopsies around the mass and we didn't detect any pathological feature. Tumors which originate from bladder dome can have a co-dominance with colorectal cancers. So, we also searched for tumour markers preoperatively. Preoperative tumour markers (CEA and Ca 19-9) of the patient were 3.7 ng/dl and 48 U/ml, respectively.

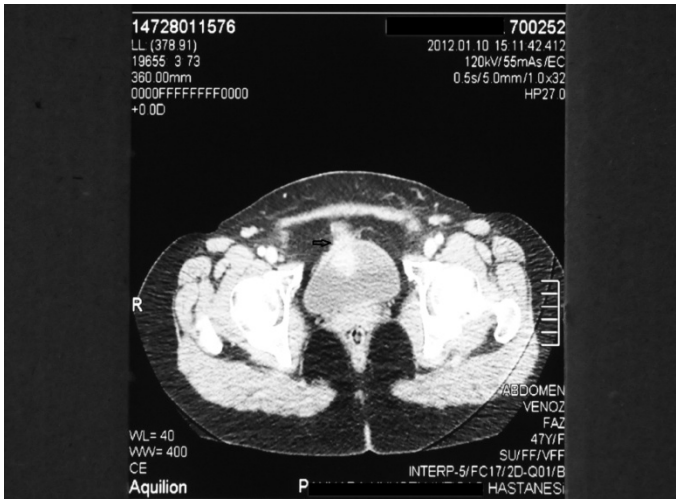

Fig. 1. The view of urachal tumour in computarized tomography
Lobulated mass hanging down from bladder dome towards into the bladder

The patient was clinically assessed as Stage-IIIA urachal adenocarcinoma which has local metastasis into the bladder, and then, an extended partial cystectomy operation including urachus and adjacent fibroadipose tissues was carried out for the patient. A bilateral pelvic lymph node dissection was performed, removing at a minimum all distal common iliac, external iliac, obturator and hypogastric nodes. The patient was discharged in the 7th day after operation. Histopathology of the partial cystectomy material was reported as urachal adenocarcinoma (colonic type, well-differentiated), surgical borders were reported as negative (Fig. 2a-d). Histopathology of lymph nodes reported as negative. The patient who was followed up with cystoscopy for three months completed the 6th month. No local or systemic recurrence had been detected in computarised tomography and no urologic complaints were present. The values of serum Ca 19-9 were 18.8 U/ml and 17.6 U/ml at the 3th month and at the sixth month after the postoperative period, respectively. The values of serum CEA in the same period were 2.6ng/dl and 2.8 ng/dl, respectively.

3. DISCUSSION

Urachal carcinoma is a type of aggressive malignancies about which relatively less known. To date, fewer than 300 cases reported in the english literature. While allantois obliterated with delivery is located as median umbilical ligament between anterior to bladder dome and umbilicus by transforming into urachus that a fibrous structure, it is found as a closed tubular epithelial duct in 30% of the population (Siefker-Radtke et al., 2003). Urachal cancers arise from the urachus which is a vestigial embryonic structure located in the space of Retzius, between the transversalis fascia anteriorly and peritoneum posteriorly and between the dome of the bladder and the umbilicus (Scabini et al., 2009). Patients with urachal cancers often present with higher stage disease at diagnosis because the disease arises outside of the bladder where it does not cause any symptoms. Symptoms often occur only after the disease has progressed further and grown into the bladder secondarily. Symptoms commonly included irritative voiding symptoms, discharge of mucous-like material and gross hematuria. Some patients also reported umbilical pain and umbilical discharge (Siefker-Radtke, 2006). Hematuria was the most common presenting symptom, followed by a palpable suprapubic mass and mucosuria (Sheldon et al., 1984). In our case, patient has

macroscopic and terminal hematuria due to the growth of the disease into the bladder (stage: T3a).

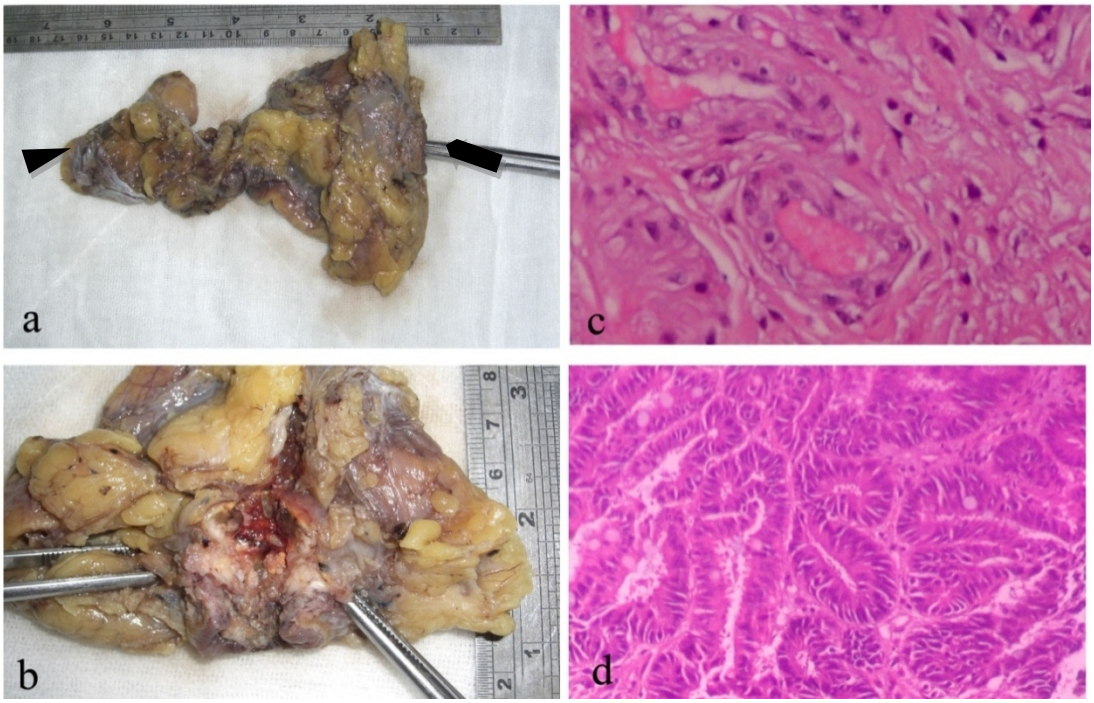

Fig. 2. a) ***Macroscopic view of the surgical resection specimen extended from the bladder dome mass (arrow) up to the umbilicus (arrowhead). b) Gross apperiance of the solid and infiltrative tumor in the urachus. The tumor has ill-defined borders and light pink-yellowish color. c) Light microscopic view of urachal remnants of the embryonic allantoic stalk connecting the umblicus to the bladder (stained with Haematoxylene and Eosine). d) Light microscopic view of urachal adenocarcinoma (stained with Haematoxylene and Eosine). Microscopically tumor is composed of enteric type well-differentiated pseudostratified columnar epithelium with hyperchromatic nuclei forming glands that resembled colonic adenocarcinoma.***

The diagnosis of urachal carcinoma is confirmed usually, but not always, by cystoscopy and endoscopic biopsy. Adopting a more practical approach, Henly et al urge that if a biopsy of the bladder dome reveals adenocarcinoma, urachal cancer should be the working diagnosis until proven otherwise (Henly et al., 1993). As in other mucinous adenocarcinomas radiologically, also psammomatous calcifications in urachal cancers may be detected with CT, MRI, ultrasonography (USG) and rarely direct radiography, and be observed as soft tissue mass containing calcifications at the mid line anterior to the bladder (Nese et al., 2003; Fancher et al., 2010). In addition, neovascularization was reported to be seen in Doppler USG. While IVU most often is normal, irregularity in bladder contours, repression in ureters may be detected sometimes (Nese et al., 2003; Fancher et al., 2010). A mass ranging to the anterior, including punctate calcification focuses at the center, with irregular and lobulated contour was also detected in our patient on the anterior bladder wall (Fig. 1).

Immunostains do not unequivocally discriminate between an urachal and a colonic malignancies although diffuse strong immunoreactivity for 34BE12 would support and diffuse nuclear reactivity for ß-catenin would militate againts a diagnosis of urachal carcinoma (Gopalan et al., 2009).

Siefker-Radtke et al emphasizes the clinical similarity of urachal cancer and adenocarcinoma of the colon. Not only is the histology similar, but they found that carcinoembryonic antigen and CA 19-9 are commonly increased in urachal cancer (Siefker-Radtke et al, 2003). Because of the common biological pathways with colon adenocarsinomas, Ca 125 also increases in urachal tumours. While Ca 19-9 was determined as high in our patient, CEA was in normal ranges. The value of Ca 19-9 began to decrease in early postoperative period and it was normal in the third month Guaraccia et al. have treated a 69 years old female patient by resection and adjuvant chemotherapy (5-fluorouracil, doxorubicin and mitomycin). After the operation, the value of CA 125 began to decrease in early postoperative period and it was normal in the second month (Guarnaccia et al,1991). In a similar study of M.D. Anderson cancer clinics, it was determined that CA125 and Ca 19-9 increased (Siefker-Radtke et al, 2003).

Several staging systems recently proposed (Sheldon et al., 1984; Pinthus et al., 2006; Ashley et al., 2006), but commonly accepted proposal by Sheldon et al as follows: Stage I- no invasion beyond the urachal mucosa; Stage II- invasion confined to the urachus; Stage III- local extension to the (a) bladder, (b) abdominal wall, and (c) viscera other than the bladder; and Stage IV- metastasis to (a) regional lymph nodes and (b) distant sites. Bratu et al. classified urachal cancers, histopathologically. The most encountered type is mucine-positive (colonic type) adenocarcinomas (Bratu et al., 2009). In our case, well-differentiated urachal adenocarcinoma in the colonic type was detected (Fig. 2). Furthermore, our case was in stage III which is the most seen stage. The case was evaluated as local advanced urachal carcinoma, because there was no lymph node involvement and surgical margin was negative (Sheldon et al., 1984).

Currently, there is no certain consensus about classification of urachal cancers and the treatment is controversial. Although the radical cystectomy is the standard treatment, the extended partial cystectomy together with umbilicus, urachus and adjacent fibroadipose tissue is also recommended (Bratu et al., 2009; Herr, 1994). There is also no consensus on concomitant excision of umbilicus. In a large series reported by Siefker Radtke et al., umbilicus was reported to be excised in cases (54%) where partial cystectomy was carried out (Siefker-Radtke et al., 2003).

Urachal cancers invade as repression into the bladder by forming a demarcation line between bladder epithelium and tumor, without showing an invasion into the bladder wall. This property provides concomitant excision of tumor with adjacent normal bladder tissue and show that partial cystectomy is an appropriate treatment alternative in appropriate cases (Herr, 1994). Because local recurrence will be in question after inadequate excision (38 to 50%), even though a group of authors recommends the radical cystectomy for all cases of urachal cancer, other authors recommend the partial cystectomy in appropriate cases (Henly et al., 1993; Herr, 1994; Kakizoe, 1983). Herr et al. reported that there is a recurrence only in 2 of 12 cases with urachal cancer, where a partial cystectomy was performed and whose majority consists of moderate and low stage tumors (Herr, 1994). Also, that supports that conservative surgery may be an effective and adequate approach in appropriate cases In another study, the cure was reported to be achieved in 14 (88%) of 16 well-differentiated cases with urachal adenocarcinoma, who were treated with partial cystectomy, and the

partial cystectomy was considered to be a good alternative for these cases by reporting that prognosis of cases, particularly, with localized, well-differentiated, colonic-type urachal adenocarcinoma (Santucci et al,1997). Our case was evaluated as non-metastatic local advanced urachal cancer. We performed extended partial cystectomy and bilateral lymph node dissection, because our patient was well-differentiated and localized.

Henly et al. reported 5-year survival rates as 50% after radical cystectomy, as 43% after partial cystectomy (Henly et al, 1993). Siefker-Radtke et al. (2003) reported 5-year survival rate similarly as 44% after partial and radical cystectomy. To be shown that negative surgical borders are achieved after conservative surgical treatment is very important for a long-term survival. Negative surgical borders were also histopathologically noted to be in our case after extended partial cystectomy. Herr et al found that the survival of patients with advanced pathologic stages, nodal metastases and with positive surgical margin was poor. The major finding of their study is that en bloc resection of the urachal tumor and urachus coupled with extended partial cystectomy cures 70% of patients with clinically localized urachal carcinoma (Herr et al., 2007). When Ashley et al. searched 66 patients with urachal cancer retrospectively, they found no difference in terms of survival in patients with parsial and radical cystectomies. They also compared Mayo and Sheldon Staging systems and they found both of the systems compatible (Ashley et al., 2006). Also, an extended partial cystectomy with bilateral pelvic lymph node dissection was carried out in our case because there is a well-differentiated, colonic-type urachal adenocarcinoma with local metastasis into the bladder, systemic metastasis is not monitored and the patient is young.

On the other hand, Gopalan et al. have reported that it is the type of surgery that is performed rather than margin status that affects local recurrence: despite all patients showing negative resection margins, the incidence of local recurrence was higher in patients who underwent a partial cystectomy alone (37.5%) than in those who had more radical surgery (27%) (Gopalan et al., 2009). Cho et al did not suggest parsial cystectomy. They showed that mucinous adenocancers and tumours larger than 4cm have worse prognosis than the rest (Cho et al., 2011).

The roles of radiation therapy and chemotherapy in the management of urachal carcinoma are unclear. Usually, patients with nodal involvement, positive surgical margin and metastasis are treated with adjuvant chemotherapy. Ashley et al. found in their study that adjuvant chemotherapy (cysplatin 100% of patients, paclitaxel 50% of patients and doxorubicin 33%of patients) does not improve survival in cancer patients (Ashley et al, 2006). In a study of MD Anderson cancer center, data of 42 urachal cancer patients were reviewed retrospectively and nodal involvement and surgical margin were investigated as important markers in the survival of the patients. In this study, patients were not treated with chemotherapy initially, they were cured with chemotherapy after metastasis. The best answer to the chemotherapy was 5-fluorouracil and cysplatin treatment (Siefker-Radtke et al., 2003).

Kume et al. performed a chemotherapy protocole including cysplatine to a 64 year old patient with metastatic urachal cancer. But, metastatic lesions did not treated with this therapy and the disease continued to progress. Tumour imitated colorectal cancers, so they used another agent called as irinotecan. An apparent shrinkage had been determined especially in the lesions of lung. But, the patient didn't tolerate the side effects of the treatment. Researchers mentioned that biological pathway of the cancer is quiet important in the election of chemotherapeutic agent (Kume et al., 2006). In our case presentation, we did

not want to give adjuvant chemotherapy because of the lack of nodal involvement and positive surgical margin.

In our case, who was followed up with cystoscopy for three months was completed at the 6th month. No local or systemic recurrence and urologic complaint are present, after partial cystectomy.

Articles about urachal cancers in literature are usually the series composing of cases in a limited number or as case presentation. Therefore, it is not possible to reach a clear result about surgical approach. There is no randomized, prospective study comparing approaches of surgical treatment. Therefore, treatment approach can be decided in the light of literature and according to clinicopathological features, also, by discussing with the patient.

4. CONCLUSION

In our opinion, nonmetastatic, well-differantiated and localized urachal cancers can be treated successfully with parsial cystectomy. If tumour has not lymph node involvement and positive surgical margin, it may not be treated with adjuvant chemotherapy. Tumour markers can be important in the follow of the disease. Undoubtly, studies with more patients are needed to confirm these knowledges.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

This study is not animal experiments or experimental study. This is a case report. Informed consent form was obtained from the patient.

CONFLICT OF INTEREST

No conflict of interest was declared by the authors.

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