CASE REPORT

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Urachal adenocarcinoma – case report and literature review

Adenokarcinom urahusa – prikaz bolesnika i pregled literature

Milan Petrović*, Vladimir Vasić†, Ljubinka Janković-Veličković‡, Srdjan Šterović*, Tomislav Pejčić§, Jovan Hadži-Djokić‖

General Hospital Leskovac, *Department of Urology, Leskovac, Serbia; †Polyclinic "Drašković", Belgrade, Serbia; Faculty of Medicine, ‡Institute of Pathology, Niš, Serbia; Clinical Center of Serbia, §Urological Clinic, Belgrade, Serbia; ‖Serbian Academy of Sciences and Arts, Belgrade, Serbia

Abstract

Introduction. Urachal adenocarcinoma is extremely rare and comprises from 0.35% to 0.7% of all bladder tumors. The most common histologic subtype of urachal tumors is adenocarcinoma which can be associated with intestinal metaplasia and mucin production. **Case report.** We report a case of a 53-year-old patient who attented a urologist because of an intermittent haematuria lasting for three months. The ultrasound examination detected infiltration of the bladder at the fundus, 24 × 29 mm in diameter. By the same wall, next to the tumor, there was an oval hypoechoic lesion about 40 mm in diameter. Computed tomography scan showed a solid, echogenic, strictly limited tumor at the fundus of the bladder, anteriorly, 32 × 35 × 22 mm in diameter which was positive after contrast application. The patient underwent partial cystectomy with complete excision of the tumor lesion 1.5 cm in healthy tissue. Histopathological analysis showed diagnosis of *Adenocarcinoma mucinosum vesicae urinariae infiltrans*. Patohistological findings detected a part of the urachal wall with a thin layer of fibromuscular tissue, chronic inflammation, microcalcifications in the lumen, flattened and desquamated epithelium. One year after the surgery, there were no signs of primary disease or metastases in other organs. **Conclusion.** Urachal adenocarcinoma is extremely rare. Long term survival could be achieved by surgical treatment in the early stage of the disease which consists of complete resection of urachal carcinoma and partial or total cystectomy.

Key words:
urachus; adenocarcinoma; urinary bladder; diagnostic techniques and procedures; urologic surgical procedures.

Apstrakt

Uvod. Adenokarcinom urahusa je izuzetno redak i čini od 0.35% do 0.7% svih tumora mokraćne bešike. Najčešći histološki tip tumora urahusa je adenokarcinom, koji može biti udružen sa intestinalnom metaplazijom i produkcijom mucina. **Prikaz bolesnika.** Prikazujemo slučaj bolesnika starog 53 godine, koji je došao kod urologa zbog intermitentne hematurije koja se javljala u periodu od tri meseca. Na ultrazvučnom pregledu uočena je infiltracija mokraćne bešike uz fundus, dijametra oko 24 × 29 mm. Uz isti zid neposredno uz tumorsku promenu, bila je prisutna ovalna hipoehogena promena dijametra oko 40 mm. Nalaz kompjuterizovane tomografije pokazao je solidnu, ehogenu, jasno ograničenu tumefakciju na krovu mokraćne bešike, anteriorno, dijametra 32 × 35 × 22 mm koja se dobro prebojavala nakon aplikacije kontrastnog sredstva. Bolesniku je učinjena parcijalna cistektomija sa potpunom ekscizijom tumorske promene na 1,5 cm do u zdravo tkivo. Histopatološkom analizom postavljena je dijagnoza *Adenocarcinoma mucinosum vesicae urinariae infiltrans*. Na patohistološkom nalazu uočen je deo zida urahusa sa tankim slojem fibromuskularnog tkiva, hroničnim zapaljenjem u zidu, mikrokalcifikatima u lumenu, aplatiranim i najvećim delom deskvamovanim epitelom. Godinu dana nakon operacije nije bilo znakova recidiva tumora niti pojave metastaza u drugim organima. **Zaključak.** Adenokarcinom urahusa je izuzetno redak. Dugoročno preživljavanje se može postići hirurškim tretmanom u ranoj fazi bolesti, koji obuhvata kompletnu resekciju karcinoma urahusa i parcijalnu ili totalnu cistektomiju.

Ključne reči:
urahus; adenokarcinom; mokraćna bešika; dijagnostičke tehnike i procedure; hirurgija, urološka, procedure.

Correspondence to: Vladimir Vasić, Marijane Gregoran 15/04, 11 000 Belgrade, Serbia. E-mail: vladimirdrvasic@gmail.com

Introduction

Urachal adenocarcinoma is extremely rare and comprises from 0.35% to 0.7% of all bladder tumors. It represents 22%–35% of all bladder adenocarcinomas [1]. The urachus is composed of three layers: outer muscular, middle layer consisting of a connective tissue and internal canal which is lined with transitional cell epithelium. It is located between transverse fascia anteriorly, parietal peritoneum posteriorly, cranial umbilicus and bladder caudal. Urachal neoplasms can arise in any of these layers, and can be epithelial or mesenchymal [2]. The criteria for the diagnosis of urachal cancer are not strictly defined. Most investigators agree with following: tumor in the dome of the bladder; absence of cystitis cystica and cystitis glandularis; predominant invasion of the muscularis or deeper tissues with a sharp demarcation between the tumor and surface of bladder urothelium. The presence of urachal remnants within the tumor and extension of tumor into the bladder wall, with involvement of the space of Retzius and anterior abdominal wall or umbilicus are also criteria for diagnosis. There is a lack of evidence for a primary neoplasm of another localization [1, 3]. In most cases, the adenocarcinoma of urachus presents with a higher stage of disease at the time of diagnosis because it develops on the outside of the bladder where it does not cause any symptoms. After the disease has progressed and grown into the bladder, dysuria, haematuria, abdominal and umbilical pain can occur secondarily [3]. Surgical treatment consists of partial or total cystectomy with *en bloc* resection of the median umbilical ligament and umbilicus [4]. Currently, there is no standard radiotherapy or chemotherapy regimen for the treatment of urachal cancers [5].

Case report

We report a case of a 53-year-old patient who attented urologist because of intermittent haematuria lasting for three months. The patient denied dysuric symptoms, fever, abdominal or umbilical pain. Case history did not mention a loss of body weight. Standard laboratory blood tests were normal. Physical examination showed regular findings. Abdomen was below the chest and there was no pain at superficial or deep palpation.

The ultrasound examination detected infiltration of the bladder at the fundus, 24 × 29 mm in diameter. By the same wall, next to the tumor, there was an oval hypoechoic lesion about 40 mm in diameter. Computed tomography (CT) scan of abdomen and pelvis (Figure 1) showed a solid, echogenic, strictly limited tumor at the fundus of the bladder, anteriorly, 32 × 35 × 22 mm in diameter, which was positive after contrast application. Superior to the tumor, intraabdominaly, outside the bladder, continued encapsulated hypodesic, strictly limited lesion about 66 mm in craniocaudal diameter. Superior lesion was in close relation with the intestinal wall, but there was no evidence of infiltration.

The patient underwent partial cystectomy with complete excision of the tumor lesion 1.5 cm in healthy tissue (Figure 2). Intraoperative and postoperative course was uneventful without complications. One year after the surgery there were no signs of primary disease or metastases in other organs.

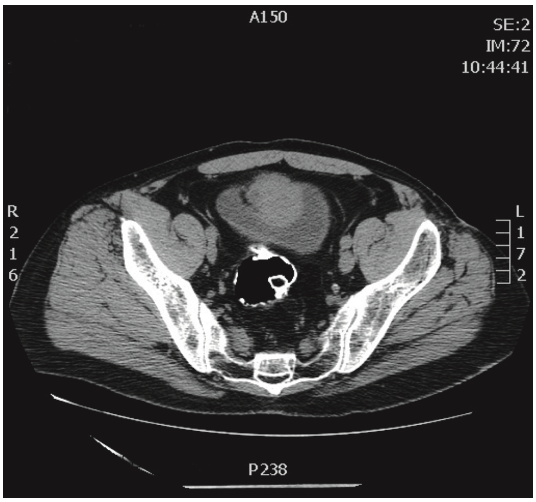

Fig. 1 – Computed tomography scan: a solid, echogenic, strictly limited tumor at the fundus of the bladder, anteriorly, 32 × 35 × 22 mm in diameter.

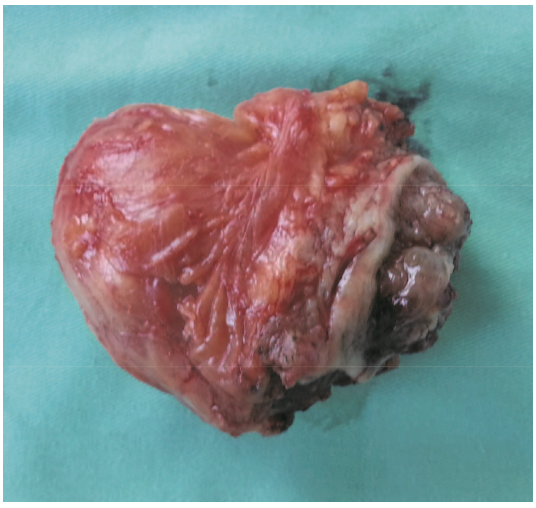

Fig. 2 – Surgical findings after partial cystectomy and complete excision of the tumor.

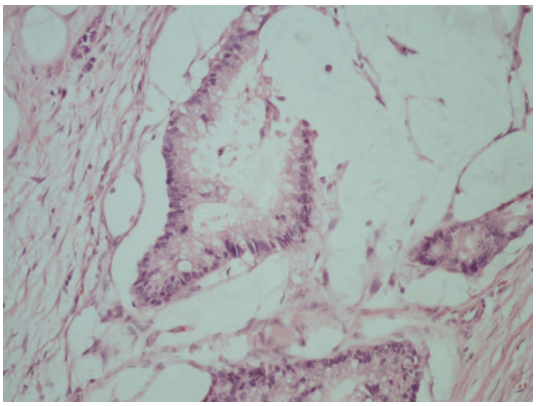

Fig. 3 – Adenocarcinoma mucinosum focus with extracellular mucin (hematoxylin eosin – HE × 200).

Histopathological (HP) analysis showed diagnosis of *Adenocarcinoma mucinosum vesicae urinariae infiltrans* (Figure 3). HP findings detected a part of the urachal wall with thin layer of fibromuscular tissue, chronic inflammation, microcalcifications in the lumen, flattened and desquamated epithelium (Figure 4). The level of histological malignancy showed grade 2. Tumor had infiltrated the connective and adipose tissue around the bladder, but had not penetrated the serous membrane. Invasion of the veins was registered at several places.

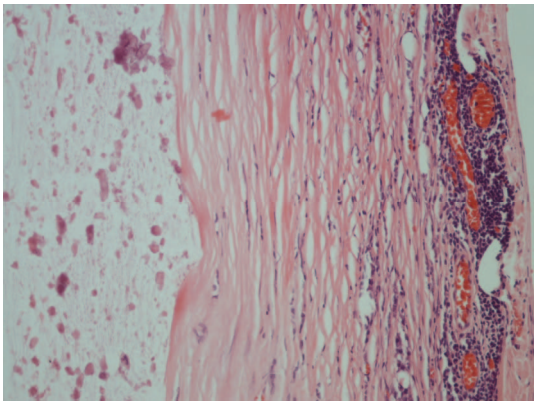

Fig. 4 – Part of wall of urachus with a thin layer of fibromuscular tissue (hematoxylin eosin – HE × 200).

Discussion

Urachal adenocarcinoma is extremely rare and in most cases it occurs in patients over 30 years of age, in 1 per 5 million people [6]. Gupta et al. [7] noticed a small number of patients younger than 30 years in whom the urachal adenocarcinoma was diagnosed. Based on a study by Ashley et al. [8], it was observed that the urachal carcinoma occured more frequently in older male than in female population. The most common symptoms are hematuria, dysuria, abdominal and umbilical pain. Ultrasound examination allows localization of the tumor and detection of highly echogenic calcifications as well as of the solid components of the tumor and lesions in the anterior abdominal wall. Williams and Chavda [9] pointed out the importance of CT or magnetic resonance imaging scans of the abdomen and pelvis, which may also provide information on local extent, lymph node involvement and metastases. Further metastatic evaluation could be obtained by chest radiography or bone scanning.

Urachal tumors can be of mesenchymal or epithelial origin. Gopalan et al. [1] noticed that the most common histologic subtype of urachal tumors was adenocarcinoma, which can be associated with intestinal metaplasia and mucin production. A very few number of studies [10] showed that squamous cell carcinoma, transitional cell carcinoma and anaplastic carcinoma can arise from the urachus. Prakash et al. [2] reported the case of a complex mucinous cystadenoma of urachus. Histopathologically, the tumor was characterized by the absence of cellular atypia as seen in adenocarcinomas. Urachal cystadenoma had a low malignant potential because it can result in *pseudomyxoma peritonei* if ruptured. Yu at al. [11] emphasized that the benign urachal neoplasms, such as adenomas, fibromas, fibroadenomas, hamartomas and fibromyomas, are extremly rare.

Criteria for the diagnosis of urachal carcinoma were established by Wheeler and Hill [12] and modified by Mostofi et al. [13]: tumor in the dome of the bladder; absence of cystitis cystica and cystitis glandularis; predominant invasion of the muscularis or deeper tissues with a sharp demarcation between the tumor and surface bladder urothelium; presence of urachal remnants within the tumor; extension of tumor into the bladder wall with involvement of the space of Retzius, anterior abdominal wall or umbilicus and no evidence of a primary neoplasm elsewhere.

Most of the patients at the time of diagnosis are in advanced stages of the disease because symptoms occur after tumor infiltration of bladder and/or other organs. Siefker-Radtke et al. [14] showed early peritoneal dissemination of urachal adenocarcinoma and metastases predilective in the bones, lungs and liver. Lee [15] reported the case of urachal adenocarcinoma that metastasized in both ovaries. If there is a bilateral mucinous adenocarcinoma of the ovaries, the presence of urachal adenocarcinoma as primary tumor must be ruled out.

The stage of the disease is determined by a system set up by Sheldon et al. [16]. The early stage of the disease implies the presence of a tumor in the urachal mucosa, while the advanced stage considers the presence of tumor in the bladder, abdominal wall, peritoneum and the presence of metastases in regional lymph nodes and distant sites. Mayo Clinic proposed a modification of this system [5].

Due to the low incidence of urachal adenocarcinoma, setting of standard treatment protocols is somewhat difficult. If the diagnosis is established in early stage of the disease, long term survival could be achieved by surgical treatment. Williams and Chavda [9] showed high survival rate of patients who underwent complete resection of urachal carcinoma and partial or total cystectomy. Asano et al. [17] pointed out the importance of intrapelvic and iliac lymphadenectomy, but consensus on this issue has not been reached, and lymphadenectomy is not routinely recommended. Elser et al. [5] reported the case of urachal adenocarcinoma which was resistant to multiple chemotherapy protocols. This is in concordance with literature data which indicate modest response of the tumor to chemotherapy and radiotherapy.

Conclusion

Urachal adenocarcinoma is extremely rare. The most common symptoms are hematuria, dysuria, abdominal and umbilical pain. Most of the patients at the time of diagnosis are in advanced stages of the disease because symptoms occur after the tumor infiltrated bladder and/or other organs. Long term survival could be achieved by surgical treatment in the early stage of the disease, which consists of complete resection of urachal carcinoma and partial or total cystectomy.

R E F E R E N C E S

1. *Gopalan A, Sharp DS, Fine SW, Tickoo SK, Herr HW, Reuter VE*, et al. Urachal carcinoma: A clinicopathologic analysis of 24 cases with outcome correlation. Am J Surg Pathol 2009; 33(5): 659–68.
2. *Prakash RM, Vijayalaxmi VS, Maitreyeer R, Ranjit PK*. Complex mucinous cystadenoma of undetermined malignant potential of the urachus: A rare case with review of the literature. Malaysian J Pathol 2014; 36(2): 145–8.
3. *Ormeci T, Kiremit MC, Erkurt B, Ormeci A*. An Unusual Long-TermSurvey of a Patient with Widespread Malignant Urachal Tumor, NotGivenChemotherapy or Radiotherapy. Case Rep Radiol 2015; 2015: ID 183787.
4. *Spiess PE, Correa JJ*. Robotic assisted laparoscopic partial cystectomy and urachal resection for urachal adenocarcinoma. Int Braz J Urol 2009; 35(5): 609.
5. *Elser C, Sweet J, Cheran SK, Haider MA, Jewett M, Sridhar SS*. A case of metastatic urachal adenocarcinoma treated with several different chemotherapeutic regimens. Can Urol Assoc J 2012; 6(1): E27–E31.
6. *Lee RS, Kang H, Kang HM, Yu DY, Choi IC, Choi HK*, et al. The Youngest Korean Case of Urachal Carcinoma. Case Rep Urol 2015; 2015: ID 707456.
7. *Gupta S, Bhaijee F, Harmon EP*. Mucinous neoplasm arising in a urachal cyst: a first in the pediatric population. Urology 2014; 83(2): 455–6.
8. *Ashley RA, Inman BA, Sebo TJ, Leibovich BC, Blute ML, Kwon ED*, et al. Urachal carcinoma: clinicopathologic features and long-term outcomes of an aggressive malignancy. Cancer 2006; 107(4): 712–20.
9. *Williams CR, Chavda K*. En Bloc Robot-assisted Laparoscopic Partial Cystectomy, Urachal Resection, and Pelvic Lymphadenectomy for Urachal Adenocarcinoma. Rev Urol 2015; 17(1): 46–9.
10. *Herr HW, Bochner BH, Sharp D, Dalbagni G, Reuter VE*. Urachal carcinoma: Contemporary surgical outcomes. J. Urol 2007; 178(1): 74–8. discussion 78.
11. *Yu JS, Kim KW, Lee HJ, Lee YJ, Yoon CS, Kim MJ*. Urachal remnant diseases: Spectrum of CT and US findings. Radiographics 2001; 21(2): 451–61.
12. *Wheeler JD, Hill WT*. Adenocarcinoma involving the urinary bladder. Cancer 1954; 7(1): 119–35.
13. *Mostofi FK, Thomson RV, Dean AL*. Mucous adenocarcinoma of the urinary bladder. Cancer 1955; 8(4): 741–75.
14. *Siefker-Radtke AO, Gee J, Shen Y, Wen S, Daliani D, Millikan RE*, et al. Multimodality management of urachal carcinoma: The M. D. Anderson Cancer Center experience. J Urol 2003; 169(4): 1295–8.
15. *Lee W*. Urachal adenocarcinoma metastatic to the ovaries resembling primary ovarian mucinous carcinoma: A case report with the immunohistochemical study. Int J Clin Exp Pathol 2011; 4(1): 118–23.
16. Sheldon CA, Clayman RV, Gonzalez R, Williams RD, Fraley EE. Malignant urachal lesions. J Urol 1984; 131(1): 1–8.
17. *Asano K, Miki J, Yamada H, Maeda S, Abe K, Furuta A*,et al. Carcinoma of urachus: report of 15 cases and review of literature: is total cystectomy the treatment of choice for urachal carcinoma? Nihon Hinyokika Gakkai Zasshi 2003; 94(4): 487–94. (Japanese)

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