The Korean Journal of Pathology 2012; 46: 83-86
http://dx.doi.org/10.4132/KoreanJPathol.2012.46.1.83

■ CASE REPORT ■

Urachal Mucinous Tumor of Uncertain Malignant Potential: A Case Report

Jung-Woo Choi · Ju-Han Lee
Young-Sik Kim

Department of Pathology, Korea University Ansan Hospital, Korea University College of Medicine, Ansan, Korea

Received: December 6, 2010
Revised: February 11, 2011
Accepted: February 22, 2011

Corresponding Author
Young-Sik Kim, M.D.
Department of Pathology, Korea University Ansan Hospital, Korea University College of Medicine, 516 Gojan 1-dong, Danwon-gu, Ansan 425-707, Korea
Tel: +82-31-412-5322
Fax: +82-31-412-5324
E-mail: apysk@korea.ac.kr

Urachal mucinous tumor of uncertain malignant potential is very rare and is characterized by a multilocular cyst showing the proliferation of atypical mucin-secreting cells without stromal invasion. As in ovarian and appendiceal borderline tumors, it represents a transitional stage of mucinous carcinogenesis in the urachus. In addition, this tumor may recur locally and develop into pseudomyxoma peritonei. Due to its scarcity and diagnostic challenges, we report a mucinous tumor of uncertain malignant potential arising in the urachus.

Key Words: Urachus; Mucinous cystadenoma

The urachus is a vestigial remnant, which connects the dome or anterior wall of the urinary bladder to the umbilicus and contains the allantois. After birth, it is closed and detectable only as a fibrous band, the median umbilical ligament.[1] If its lumen persists, it may be the primary site of various tumors, most of which are well-differentiated mucinous adenocarcinoma.[2] In contrast, benign mucinous tumors have rarely been reported in the English literature.[3-5] Their very rare incidence hinders the establishment of classification schemes of urachal mucinous tumors, especially regarding the presence of borderline tumors morphologically similar to those occurring in the ovary and appendix. Herein, we describe a very rare case of urachal mucinous tumor of uncertain malignant potential (mucinous borderline tumor), characterized by the proliferation of atypical mucin-secreting cells but with no evidence of stromal invasion.

CASE REPORT

A 29-year-old female was referred to our hospital because of a intermittent occurrence of right flank pain for 1-week. The patient had neither a family nor personal medical history. Aside from the flank pain, the patient had no other urinary symptoms such as hematuria or mucusuria. Biochemistry and hematologic results were within normal limits. A computed tomography scan of the abdomen and pelvis revealed an ovoid cystic mass located just anterior to the dome of the urinary bladder with a partially calcified wall (Fig. 1A). Radiologically, adenocarcinoma arising from urachal remnant was suggested. The patient underwent a radical excision of this mass including the attached portion of the urinary bladder and median umbilical ligament. At laparotomy, the mass originated from the dome of the urinary bladder without connection to the bladder cavity. There was no evidence of pseudomyxoma peritonei or tumors of other organs including ovary and appendix. The patient has been followed-up postoperatively with no evidence of recurrence for 11 months.

Grossly, a multilocular cystic mass with abundant mucoid materials, measuring 5.5×3.5×1 cm, was found, which was connected to a small cuff of urinary bladder wall and a segment of median umbilical ligament (Fig. 1B). The mass did not show any papillary or solid lesions and did not communicate with the urinary bladder lumen. Histologically, the cyst was lined by a tall columnar epithelium with cellular crowding and stratification (Fig. 2A). The epithelium showed changes ranging from simple benign columnar epithelium to areas of dysplasia. Cytologically, the lining columnar cells had abundant mucin in the cytoplasm and exhibited loss of polarity, nuclear pleomorphism,

pISSN 1738-1843
eISSN 2092-8920

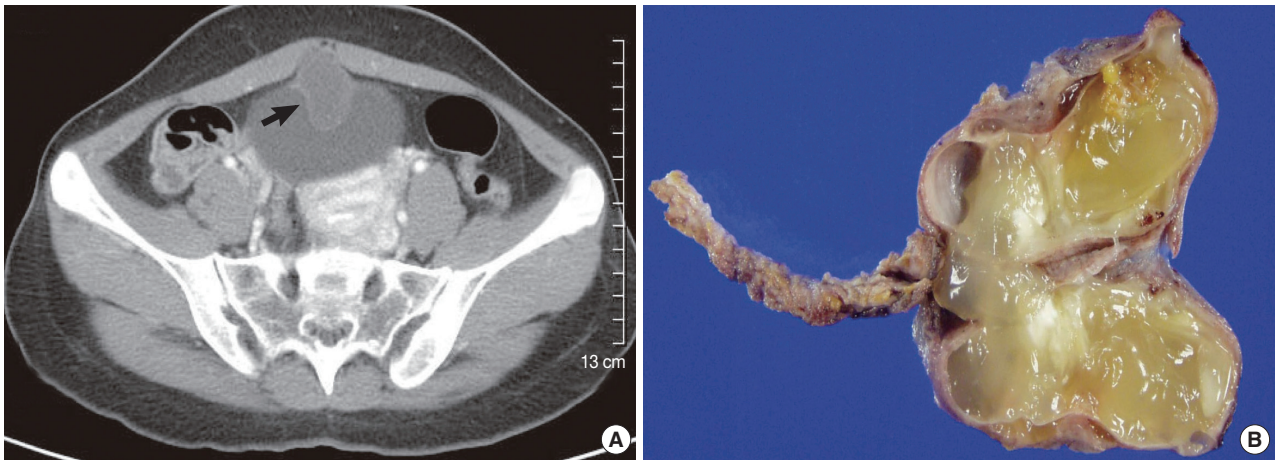

Fig. 1. (A) Computed tomography scan of the pelvis reveals an irregular cystic mass (arrow) in the dome of the urinary bladder. (B) Grossly, the multilocular cyst contains abundant mucin.

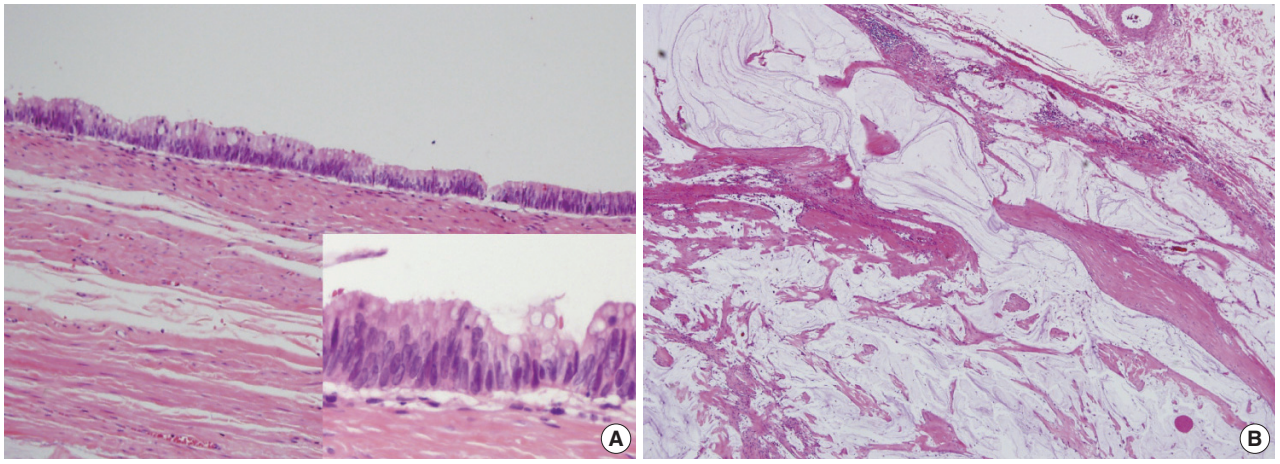

Fig. 2. (A) Microscopically, the lining epithelium consists of stratified atypical columnar cells containing mucin. (B) The underlying stroma shows extensive mucin extravasations without tumor cells.

and hyperchromatism in dysplastic areas, but there were no mitotic figures. No stromal invasion of the tumor cells was observed, although mucin was extravasated into the cyst wall (Fig. 2B), and foci of dystrophic calcification were present. The underlying stroma was composed of hyalinized connective tissue. On the basis of the proliferation of atypical mucinous cells and the absence of stromal invasion, the tumor was diagnosed as a mucinous tumor of uncertain malignant potential. Its association with urachal remnant and location just anterior to the dome of urinary bladder confirmed the primary urachal origin of this tumor.

DISCUSSION

Tumors arising in the urachus are generally classified into two categories: benign and malignant. Urachal mucinous tumors with features morphologically similar to ovarian and appendiceal borderline tumors are extremely rare. Only four cases have been reported in the English literature.[6-9] The authors of those cases raised the possibility that, as in ovarian and appendiceal borderline tumors, there is a transitional stage of mucinous carcinogenesis from benign to intraepithelial or infiltrative carcinoma, and those reported cases could represent the premalignant phase in this progression sequence. That hypothesis is also supported by other reports that urachal adenomas may coexist with *in situ* and invasive adenocarcinoma.[10,11] All mucinous tumors, regardless of the source, should be accepted as having an aggressive biological potential such as local recurrence and the development of pseudomyxoma peritonei.[12] Therefore, it is not surprising that a urachal adenoma with numerous mu-

 http://dx.doi.org/10.4132/KoreanJPathol.2012.46.1.83

Table 1. Reported cases of urachal mucinous tumors with uncertain malignant potential

References	Age (yr)/Sex	Size (cm)	PMP	Diagnosis	Treatment	Follow-up
Carr and McLean[6]	72/M	4	Absent	Mucinous tumor of uncertain malignant potential	Partial cystectomy	Not mentioned
Paul *et al.*[7]	68/M	3	Absent	Stage 0 mucinous adenocarcinoma *in situ*	Partial cystectomy	1 yr, WOR
Shinohara *et al.*[8]	54/M	6	Present	Mucinous cystic tumor with low malignant potential	Partial cystectomy	7 yr, WOR
Stenhouse *et al.*[9]	54/M	11	Present	Mucinous neoplasm of uncertain malignant potential	Not mentioned	6 mo, WOR
Present case	29/F	5.5	Absent	Mucinous tumor of uncertain malignant potential	Partial cystectomy	11 mo, WOR

PMP, pseudomyxoma peritonei; WOR, without recurrence.

cinous implants, although histologically benign, might lead to death.[13] A recent report described a complex mucinous cystadenoma of undetermined malignant potential on the basis of only extensive mucin extravasation into perivesical soft tissue, even in the absence of dysplasia in the lining of the epithelium and pseudomyxoma peritonei.[5]

Characteristics of the so-called borderline urachal mucinous tumors reported thus far are listed in Table 1. All the cases described marked nuclear pleomorphism, stratification, and various degrees of mitoses in the lining epithelium but no stromal invasion. Two cases revealed the occurrence of pseudomyxoma peritonei, which required adjuvant therapy such as intraperitoneal lavage in addition to surgical removal. However, complete surgical excision was of paramount importance, as it was in the benign tumors. No recurrence was found in all of those cases, although there were limitations that the number of cases was too small to be representative and the follow-up periods were not long enough. Notably, the patient of our case was in the third decade, whereas the other reported ones were in their sixth to eighth decades.

The differential diagnosis of urachal mucinous tumor of uncertain malignant potential includes villous adenoma, urachal mucinous adenocarcinoma, urothelial malignancy with components of adenocarcinoma, and metastatic mucinous tumor from a variety of body sites including breast, pancreas, ovary, prostate gland, and gastrointestinal tract. Villous adenoma is a benign glandular neoplasm lined by pseudostratified columnar epithelium and is characterized by papillary architecture instead of formation of a cyst containing abundant mucinous materials. Mucinous adenocarcinoma, either primary or secondary, can be easily confirmed by clinical information and the presence of stromal invasion. Criteria to classify a tumor as urachal in origin are as follows: 1) presence of urachal remnants, 2) tumor growth in the bladder wall and 3) in the dome of urinary bladder, 4) absence of dysplastic intestinal metaplasia or dysplastic mucosal change, and 5) exclusion of other primary malignancies.[14,15]

We herein describe a rare case of mucinous tumor of uncertain malignant potential arising in the urachus. To the best of our knowledge, this is the first reported case in Korea and the fifth case in the English literature. We expect that this report will help to clarify the biological behavior and therapeutic implications of urachal mucinous tumors.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Taylor S, Bacchini P, Bertoni F. An isolated vertebral metastasis of urachal adenocarcinoma. Arch Pathol Lab Med 2004; 128: 1043-5.
2. Ohira S, Shiohara S, Itoh K, Ashida T, Fukushima M, Konishi I. Urachal adenocarcinoma metastatic to the ovaries: case report and literature review. Int J Gynecol Pathol 2003; 22: 189-93.
3. Elliott GB, Freigang B. Observations on the nature of mucin secreting urachal cystadenoma. Ann Surg 1963; 157: 613-7.
4. Hull MT, Warfel KA. Urachal cystadenoma with abundant glycogen: ultrastructural study. Ultrastruct Pathol 1994; 18: 499-502.
5. Schell AJ, Nickel CJ, Isotalo PA. Complex mucinous cystadenoma of undetermined malignant potential of the urachus. Can Urol Assoc J 2009; 3: E39-41.
6. Carr NJ, McLean AD. A mucinous tumour of the urachus: adenoma or low grade mucinous cystic tumour of uncertain malignant potential? Adv Clin Path 2001; 5: 93-7.
7. Paul AB, Hunt CR, Harney JM, Jenkins JP, McMahon RF. Stage 0 mucinous adenocarcinoma *in situ* of the urachus. J Clin Pathol 1998; 51: 483-4.
8. Shinohara T, Misawa K, Sano H, Okawa Y, Takada A. Pseudomyxoma peritonei due to mucinous cystadenocarcinoma *in situ* of the urachus presenting as an inguinal hernia. Int J Clin Oncol 2006; 11: 416-9.
9. Stenhouse G, McRae D, Pollock AM. Urachal adenocarcinoma *in situ* with pseudomyxoma peritonei: a case report. J Clin Pathol 2003; 56: 152-3.
10. Cheng L, Montironi R, Bostwick DG. Villous adenoma of the urinary tract: a report of 23 cases, including 8 with coexistent adeno-

carcinoma. Am J Surg Pathol 1999; 23: 764-71.
11. Seibel JL, Prasad S, Weiss RE, Bancila E, Epstein JI. Villous adenoma of the urinary tract: a lesion frequently associated with malignancy. Hum Pathol 2002; 33: 236-41.
12. de Bree E, Witkamp A, Van De Vijver M, Zoetmulde F. Unusual origins of Pseudomyxoma peritonei. J Surg Oncol 2000; 75: 270-4.
13. Eble JN, Hull MT, Rowland RG, Hostetter M. Villous adenoma of the urachus with mucusuria: a light and electron microscopic study. J Urol 1986; 135: 1240-4.
14. Abenoza P, Manivel C, Fraley EE. Primary adenocarcinoma of urinary bladder: clinicopathologic study of 16 cases. Urology 1987; 29: 9-14.
15. Jones WA, Gibbons RP, Correa RJ Jr, Cummings KB, Mason JT. Primary adenocarcinoma of bladder. Urology 1980; 15: 119-22.

 http://dx.doi.org/10.4132/KoreanJPathol.2012.46.1.83