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A NON-HEMATURIA URACHAL ADENOCARCINOMA IN A FORM OF AN INFECTED UMBILICAL MASS: CASE REPORT

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ABSTRACT

We present a case report of a 72 year-old Asian female who presented with an enlarging infected umbilical mass without any urinary symptom, and received an initial diagnosis of either umbilical-urachal sinus infection or infected urachal remnants. But histopathology surprisingly revealed a primary mucinous adenocarcinoma arising from urachus. Clinicians should be alert that urachal carcinoma may be formed as infected umbilicus mass and without hematuria.

INTRODUCTION

Urachal adenocarcinoma is a rare tumor with insidious course and variable presentation. Several case reports and cohort studies have shown hematuria to be the most common presenting symptoms in 82 % of urachal carcinoma [1,4]. The urachus is a vestigial structure that connects the dome of the bladder to the umbilicus, and it normally closes by the fourth month of fetal life [2,3]. In Sheldon et al.'s report, most of urachal carcinoma was arising from the dome of urinary bladder extending towards abdominal wall via urachus [1,2].

Persistent malformations of the urachus commonly present in childhood and occasionally in the adulthood, and include patent urachus, urachal cysts, and urachal sinuses, all of which can result in secondary infection or give rise to tumors that most frequently were adenocarcinoma [5,10]. The treatment options depend on the diagnosis and whether the tumor was benign or malignant. Traditionally, the umbilical-urachal sinus is surgically excised using a transverse or midline infraumbilical incision [5,10]. But infected urachal

remnants are initially treated with drainage and antibiotics, followed by surgical excision [5]. Treatment of urachal adenocarcinoma consists of open radical or partial cystectomy with pelvic lymph node dissection, and total excision of the umbilicus and the urachal ligament [2,6].

The case:

A 72 year-old Asian female presented to our department with a painful pigmented 0.5-cm umbilical mass which was first noted 3 months prior to the admission. She had been treated with antibiotics and analgesics at local clinics as the umbilical mass was first suspected to be infected. Two weeks before admission, odorless and clear watery discharge was noted at the umbilicus and the mass had since enlarged from 0.5 cm to 1.5~2.0 cm in diameter, with heterogeneous brownish pigmentation and rough surface with erythematous changes. Patient denied appetite change, gross hematuria or any other urinary symptom. Abdominal physical examination revealed an erythematous, round, rough, firm,



and pigmented umbilical mass grossly which was tender to touch. The biochemical and hematological analysis were within normal limit. The patient underwent wide excision umbilectomy with excision of urachus partially for the impression of either umbilical-urachal sinus infection or infected urachal remnants. The distal urachus, urinary bladder, peritoneum and intra-abdominal cavity all appeared grossly normal during operation. The urachus beneath abdominal wall extended from umbilicus to the dome of urinary bladder without any palpable mass or uneven surface. Histological examination of the biopsy specimen revealed a mucinous adenocarcinoma with free surgical margins. The tumor arose from upper dermis and almost reached epidermis of umbilicus (Fig.1). Grossly, there showed a tumor of 2.5x2.2x2cm in size extending to

almost whole layer of the abdominal wall. Microscopically, the peritoneal surface shows moderate chronic inflammation and upper dermis shows tumor cell invasion (Fig.2).

The patient was further evaluated with an abdominal ultrasound which revealed multiple small hypoechoic masses (1~2cm) over both of the liver lobes suspected to be metastatic lesions. The results of whole body bone scan and chest X-rays were negative for metastatic lesion. The patient was then referred to medical center for further staging the tumor by lymph node dissection for histological analysis. (Table 1) Chemoradiation therapy was provided, and short survival two year after her surgical treatment. We obtained patient's informed consent to report this case.

Table 1. The staging system for urachal cancer

Stage	Definition
I	Confined to urachal mucosa
II	Invasion confined to urachus itself
IIIA	Local urachal cancer extention to bladder
IIIB	Local urachal cancer extention to abdominal wall
IIIC	Local urachal cancer extention to peritoneum
IIID	Local urachal cancer extention to viscera other than bladder
IVA	Metastatic to lymph nodes
IVB	Distant metastasis
*See Sheldon et al.,1984 (2)	

Figure 1. The tumor arose from upper dermis and almost reached epidermis of umbilicus

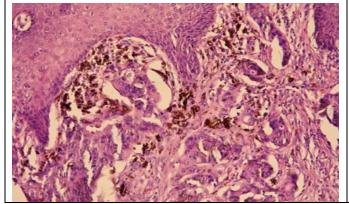
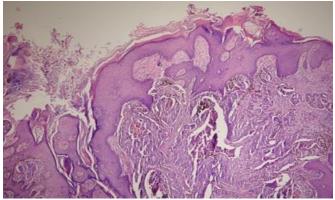


Figure 2. The peritoneal surface shows moderate chronic inflammation and upper dermis shows tumor cell invasion



DISCUSSION

To our knowledge, our study may be the first to show the urachal carcinoma in the form of an infected umbilicus mass. Hematuria was the common presentation in more than two thirds of reported cases [3]. But for this patient, the urinary bladder was intact and no hematuria was reported. Our patient received wide excision umbilectomy with excision of urachus due to superficial invasion with clinical manifestation as umbilical-urachal sinus infection and or infected urachal remnants, and primary adenocarcinoma from the urachus was impressed. Urachal carcinomas frequently metastasize to iliac and

inguinal lymph nodes, omentum, liver, lung, and bone [2,7]. It is important to remove all anomalous tissue of urachus or umbilicus for treatment [2,3], in order to prevent recurrence or transformation into a malignant adenocarcinoma[4]. Negative margin status has been identified as an important factor for long term-survival and adequate local treatment is of paramount value [6]. In the initial management of urachal carcinoma, umbilectomy with partial cystectomy may be considered; this can enhance quality of life without necessarily influencing survival adversely [8].



CONCLUSION

The symptoms of hematuria will depend on tumor origin. If the urachal carcinoma invasion the urinary bladder, hematuria will be presented [1,4]. In our case, the tumor grew over umbilical region and formed an infected mass, pathologic analysis revealed the cancer cell was invasion to the skin layer and mostly invaded the abdominal wall layer. The treatment modality of this patient will be another controversial issue, the extend of excision should be wider or other novel preoperative evaluation in the literature should be applied. Overall survival for all stages of urachal adenocarcinoma is 62 months with 34% of the patients being alive after 5 years [3]. Misdiagnosis of urachal carcinoma is still a reality. Accurate diagnosis needs an appropriate imaging studies and alertness of atypical unreasonable symptom sign. Appropriate local treatment is the key for improved survival. Pathologic stage is an important prognostic factor in urachal carcinoma. Surface urothelial involvement by carcinoma and presence of cystitis cystica/glandularis do not necessarily exclude the diagnosis of urachal carcinoma [9]. Clinicians should be alert of these rare tumors over the umbilicus without typical hematuria.

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CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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