

## URACHAL ADENOCARCINOMA: A RARE CASE REPORT

## Urology

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## ABSTRACT

Urachal adenocarcinoma is a rare and aggressive form of bladder cancer involving the urachus, a fibrous remnant of the allantois that extends from the bladder to the umbilicus. We report this case of a 47 year old women with primary urachal mucinous cystadenocarcinoma treated with en bloc resection (urachal mass and urachectomy up to umbilicus) with partial cystectomy. The patient presented with abdominal pain and intermittent hematuria with imaging reveals a large mass involving the dome of the bladder extending from the urachus.

## KEYWORDS

Urachus, Bladder, mucinouscystadenocarcinoma

## INTRODUCTION :

Urachal cancer is a very rare type of cancer arising from the urachus or its remnants.[1] The disease might arise from metaplastic glandular epithelium or embryonic epithelial remnants originating from the cloaca region.[2]

It occurs in roughly about one person per 1 million people per year varying on the geographical region.[3] Detailed diagnostic and staging schemes were proposed by Sheldon et al in 1984, which remain widely used today.[1]

We report the case of a 47-year-old female patient with a stage I (Mayo Clinic) primary urachal adenocarcinoma with good outcomes after surgery.

## Case Report:

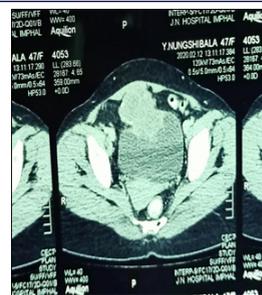
A 47-year-old female patient presented in our urology OPD with chief complaints of abdominal pain and intermittent hematuria from the last 3 months. On physical examination, a palpable firm and non tender lump was present in the midline of the suprapubic region.

The USG whole abdomen was advised to further assess the mass and the ultrasound (US) examination revealed a heterogeneous mass in fundal region of the urinary bladder measuring 5.7×4.3×7.9cm with bilateral kidneys and ovaries normal as shown below in the diagram(fig.1)

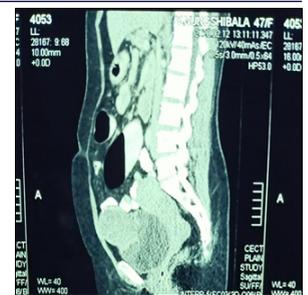


**Fig 1(Ultrasound Delineating Bladder Fundal Mass)**

Contrast enhanced computed tomography (CECT) scan of the abdomen was planned subsequently and was reported as a heterogeneous enhancing mass lesion arising from anterosuperior wall of the urinary bladder with intravesical irregular margins suggestive of growth and exophytic component shows peripheral calcification and central cystic component extending to the anterior wall of the abdomen with obliterated fat planes, radiological features suggestive of urachal carcinoma (Fig 2a & 2b).



**Fig 2a-Transverse Section**



**Fig 2b- Coronal Section**

After getting admission in urology ward, the patient was planned for cystoscopy under local anaesthesia and the cystoscopy findings shows infiltrative lesions involving the dome of bladder with friable mucosa and rest of bladder mucosa and bilateral ureteral orifices were normal.

Other baseline investigations were done for the fitness for surgery and after getting appropriate consent for the surgery, the patient was planned for surgery. The patient was also explained about the need for augmentation cystoplasty if found decreased bladder capacity intraoperatively.

The patient was posted for surgery and en bloc resection (urachal mass and urachectomy up to umbilicus) with partial cystectomy was done and specimen was sent for histopathology.

During the postoperative period the patient remains uneventful.

Postoperative specimen photos (Fig 3a & 3b).

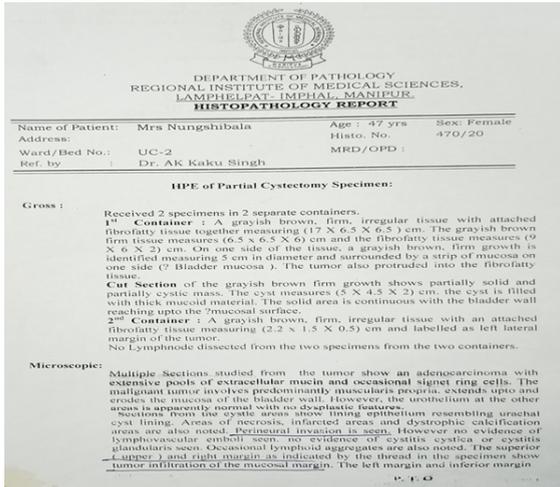


**Figure 3a (resected Whole Specimen)**



**Figure 3b (bladder Side Margin)**

Histopathology of resected tumor report came out as urachal mucinous cystadenocarcinoma (Sheldon stage 3a).



**Follow Up:**

Patient was followed-up at 2 week and then at 3 months. She was stable with healthy scar at the incision site and no palpable lump on examination. Repeat CT urogram was done at 3 month, report was normal with no residual disease(fig.4) and patient is on regular followup in our department.



**Fig.4...CT urogram at 3 month (no residual disease)**

**DISCUSSION:**

The urachus is the embryologic remnant of allantois and the adjacent ventral cloaca. It is a tubular structure in which lumen becomes obliterated with the advancing age. But its patency with the urinary bladder may persist in a small proportion of adults [4].

Urachal tumors are rare and devastating tumors of the bladder which were first described by Hue and Jacquin in 1863. They account for only 0.5% of all bladder malignancies, and 20–40% of primary bladder adenocarcinomas [5-7].

Hematuria is the most common presenting symptom in about 90%of patients [8]

The MD Anderson Cancer Center (MDACC) suggested 5 criteria for the diagnosis of urachal cancers. These criteria include a midline location of the tumor; a sharp demarcation between the tumor and normal surface epithelium; an enteric histology; the absence of urothelial dysplasia, cystitis cystica or cystitis glandularis transitioning to the tumor; and the absence of a primary adenocarcinoma of another origin [9,10].

Wheeler and Hill in 1954 proposed 5 criteria: location in the bladder dome or anterior wall; invasion of the bladder wall from outside to inside; absence of cystitis cystica or cystitis glandularis; presence of

embryonic remnants; absence of a primary adenocarcinoma of another origin [11]. All of these criteria were present in our reported case.

Partial cystectomy with en bloc urachectomy up to the umbilicus is considered the gold standard for the treatment of urachal carcinoma when the disease is surgically resectable. Partial cystectomy is performed to ensure negative margins. En bloc resection of the urachal ligament and umbilicus is recommended because tumors can occur anywhere along the urachus, including at the umbilicus (7%) [7].

In 1984, Sheldon et al. [1] proposed a system for clinical staging of urachal adenocarcinoma. In this system, early stage urachal cancers are localized to the urachal mucosa, whereas late stage disease involves local structures, like the bladder, abdominal wall or peritoneum, and metastases to regional lymph nodes or distant sites (Table 1). The Mayo clinic has suggested recently a more simplified system (Table 2) [12]. But none of them are validated.

**Table 1** clinical staging system by Sheldon et al. [1].

- Stage I Urachal cancer confined to urachal mucosa
- Stage II Urachal cancer with invasion confined to urachus itself
- Stage IIIA Local urachal cancer extension to bladder
- Stage IIIB Local urachal cancer extension to abdominal wall
- Stage IIIC Local urachal cancer extension to peritoneum
- Stage IIID Local urachal cancer extension to viscera other than bladder
- Stage IVA Metastatic urachal cancer to lymph nodes
- Stage IVB Metastatic urachal cancer to distant sites

**Table 2** clinical staging system by Mayo clinic [12].

- Stage I Urachal cancer confined to the urachus and/or bladder
- Stage II Urachal cancer extending beyond the muscular layer of the urachus and/or bladder
- Stage III Urachal cancer infiltrating the regional lymph nodes
- Stage IV Urachal cancer infiltrating the non-regional lymph nodes or other distant sites

Recent case reports h show the benefit of combined chemotherapy in isolated cases of urachal cancers, most of them adenocarcinomas: the association of 5-FU, cisplatin or oxaliplatin, irinotecan and bevacizumab in different combinations demonstrated usually a partial and limited response [13–16].

There is currently no standard adjuvant or metastatic chemotherapy protocol for the treatment of urachal adenocarcinoma.

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