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URACHAL ADENOCARCINOMA: A RARE CASE REPORT

Urology

Dr. Nawaz Ali* Senior Resident department of urology RIMS. *Corrresponding Author

Prof. Ak .Kaku Singh Professor & HOD department of urology RIMS.

Dr. Kh. Somarendra Assistant professor department of urology RIMS.

KEYWORDS

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 measuring 5.7×4.3×7.9cm with bilateral kidneys and ovaries normal as shown below in the diagram(fig.1)

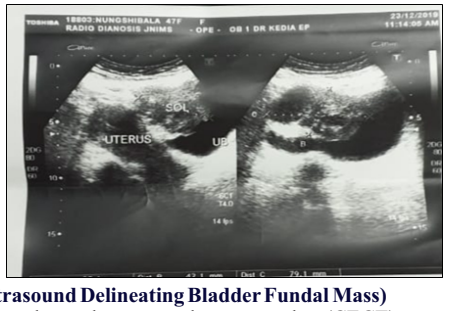


Fig 1(ultrasound Delineating Bladder Fundal Mass)

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 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).

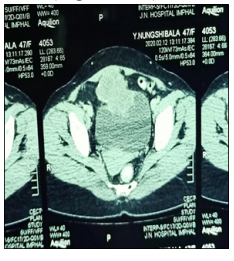


Fig 2a-Transverse Section

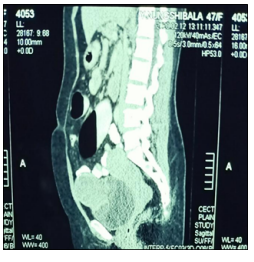


Fig 2b- Coronal Section

After getting admission in urology ward, the patient was planned for cystoscopy under local anaesthesia and it 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 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).

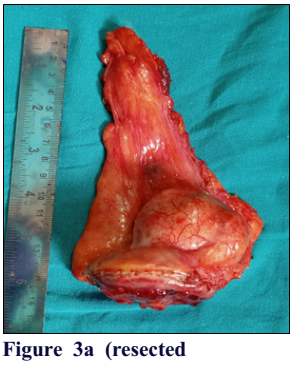


Figure 3a (resected Whole Specimen)

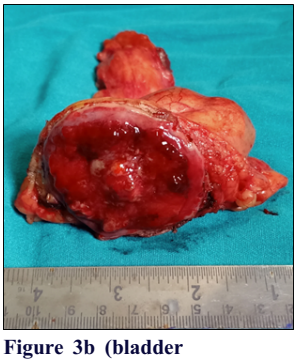
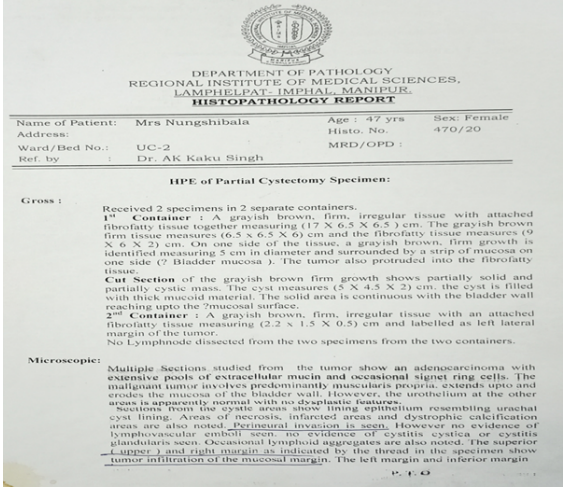


Figure 3b (bladder Side Margin)

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



DEPARTMENT OF PATHOLOGY
REGIONAL INSTITUTE OF MEDICAL SCIENCES,
LAMPHELPAT- IMPHAL, MANIPUR.
HISTOPATHOLOGY REPORT

Name of Patient: Mrs Nungshibala Age : 47 yrs Sex: Female
Address: Histo. No. 470/20
Ward/Bed No.: UC-2 MRD/OPD :
Ref. by : Dr. AK Kaku Singh

HPE of Partial Cystectomy Specimen:

Gross :

Received 2 specimens in 2 separate containers.
1st Container : A grayish brown, firm, irregular tissue with attached fibrofatty tissue together measuring (17 X 6.5 X 6.5) cm. The grayish brown firm tissue measures (6.5 x 6.5 X 6) cm and the fibrofatty tissue measures (9 X 6 X 2) cm. On one side of the tissue, a grayish brown, firm growth is identified measuring 5 cm in diameter and surrounded by a strip of mucosa on one side (? Bladder mucosa). The tumor also protruded into the fibrofatty tissue.
Cut Section of the grayish brown firm growth shows partially solid and partially cystic mass. The cyst measures (5 X 4.5 X 2) cm. the cyst is filled with thick mucoid material. The solid area is continuous with the bladder wall reaching upto the ?mucosal surface.
2nd Container : A grayish brown, firm, irregular tissue with an attached fibrofatty tissue measuring (2.2 x 1.5 X 0.5) cm and labelled as left lateral margin of the tumor.
No Lymphnode dissected from the two specimens from the two containers.

Microscopic:

Multiple Sections studied from the tumor show an adenocarcinoma with extensive pools of extracellular mucin and occasional signet ring cells. The malignant tumor involves predominantly muscularis propria, extends upto and erodes the mucosa of the bladder wall. However, the urothelium at the other areas is apparently normal with no dysplastic features.
Sections from the cystic areas show lining epithelium resembling urachal cyst lining. Areas of necrosis, infarcted areas and dystrophic calcification areas are also noted. Perineural invasion is seen. However no evidence of lymphovascular emboli seen, no evidence of cystitis cystica or cystitis glandularis seen. Occasional lymphoid aggregates are also noted. The superior (upper) and right margin as indicated by the thread in the specimen show tumor infiltration of the mucosal margin. The left margin and inferior margin

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.

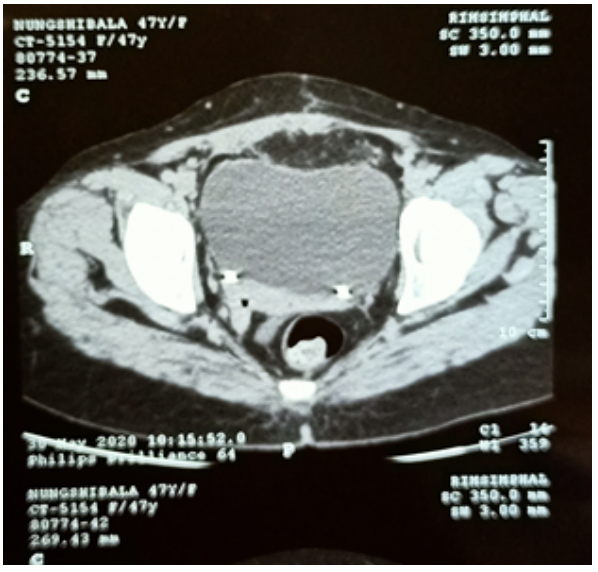


Fig.4

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 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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