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Case Report

An unusual case of painless haematuria- a rare case and review of literature

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ABSTRACT

Urachal carcinoma is a rare and aggressive cancer arising from a persistent urachus which is a remnant of the allantois that extends from blader to umblicus. This is a case report of urachal carcinoma in a 37year-old male who presented with gross painless hematuria. On radiological investigation he was found to have a tumor involving dome of bladder and urachal remnant. Cystoscoic biopsy suggested malignancy. Partial cystectomy with enblock removal of urachus and umblicus was done. On cutting the specimen through umblicus the urachal remnant was identified very well with a tumor from distal urachus infiltrating into the bladder. Histologicaly this tumor was identified as adenocarcinoma-enteric type involving, urachus, bladder and perivesical fat.

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INTRODUCTION

Painless hematuria can be a manifestation of both neoplastic and non-neoplastic conditions. Among the neoplasms, urachal cancer is a well known but unusual cause for painless hematuria. It is a malignant epithelial neoplasm arising from the urachus which is the persistent embryological remnant of the urogenital sinus and allantois wich usually undergoes involution before birth. The neoplasms arising from this structure are predominantly adenocarcinomas with aggressive behavior presenting as mass lesions involving the

bladder dome or anterior abdominal wall.

CASE REPORT

Our patient, a 37 year-old male presented with history of gross painless hematuria and passing blood clots in urine since three days. No comorbidities and no relevant past history. He had no symptoms relating to any other organs in the body. Physical examination appeared within normal limits. All routine blood investigations were also within normal limits. Urine examination revealed gross and microscopic hematuria. Detailed radiological examinations were done. Ultrasound examination showed a hypoechoic solid lesion in urinary bladder suggestive of a neoplasm. CT scan revealed a heterogeneously enhancing soft tissue lesion in midline aspect of dome of urinary bladder suggesting a primary bladder malignancy. On further imaging with MRI, an irregular heterogenous polypoidal mass lesion of size 3.3x3.2x3.2 was noted arising from antero-superior wall of urinary bladder at the urachal attachment. Mild surrounding fat stranding was noted indicating perivesical fat involvement. All other organs appeared normal with USG, CT & MRI.

With this findings a cystocopy was done which showed a growth at the dome of bladder projecting over the mucosal surface and the same was biopsied. Histopathology report suggested a diagnosis of adenocarcinoma. A partial cystectomy with 2cm margin and enbloc dissection of the umbilicus was done (fig. 1) The whole specimen measured 10x6x2 cm and on cutting through the umbilicus was seen a linear whitish fibrous cord like structure(urachal remnant) extending from below the umbilicus through the subcutaneous tissue and reaching up to the dome of the bladder and a whitish, firm, solid, granular growth measuring 3.5x 3x 2 cm involving the dome of bladder with extension to the adjacent urachal remnant for a length of 1.5 cm.(fig.2) On serial sectioning the tumor was seen involving the entire thicknecss of the bladder wall with infiltration to the surrounding fat as seen in the specimen cut surface. Histology (fig. 3A-D) showed a moderately differentiated adenocarcinoma – enteric type. A mucinous component was seen in focal areas.

The neoplasm was seen infiltrating the urachal segment, perivesical fat, and the superficial and deep muscularis propria of the bladder. Adjacent mucosal epithelium of the bladder appeared normal. Umbilicus, periumblical skin, and subcutaneous tissue underneath and around the umbilicus was unremarkable. With this morphology, clinical and radiological findings and in the absence of any lesion in any other organs of the body a diagnosis of primary urachal adenocarcinoma-enteric type with a mucinous component was made.

DISCUSSION

The urachus is a musculofibrous band of tissue with a canal that connects the allantois to the fetal bladder during early fetal life. Failure of total obliteration of urachus which may occur in one third of adults can result in various non neoplastic and neoplastic lesions of the urachus in later life. Urachal carcinoma is the most important of these lesons.[1,2] We here report such a case in which a 37 year -old apparently healthy male who presented with a short history gross painless hematuria and finally proved to be urachal adenocarcinoma. It is a very rare malignant epithelial neoplasm with adverse outcome and constitute 0.01% of all adult cancers.[3] The main reason for the poor prognosis are related to the late diagnosis due to lack of early symptoms and the tendency for early local and distant spread.

In our case, the patient was apparently healthy at the time of presentation. Radiological investigation of our patient revealed the tumor infiltrating the bladder wall and projecting into the mucosal surface. This had resulted in hematuria which was the only clinical manifestation in our patient. Other symptoms reported in patients with urachal carcinoma are irritative voiding, mucus like discharge from umbilicus and abdominal symptoms due to mass effect or umbilical pain. It may even be detected incidentally when imaging is done for other purposes.[4] Very rarely patients can present with metastatic disease. Lee W[5] has reported a urachal carcinoma metastatic to the ovaries which resembled a primary ovarian mucinous carcinoma. Such cases can be differentiated using immunohistochemical (IHC) studies. In our case there was no clinical, radiological or gross morphology differential diagnosis and IHC was not done.

Diagnosis of urachal carcinoma is mainly based on typical radiological findings and cystoscopy followed by biopsy correlation as has happened in our case. MRI will be of great help in assessing local invasive features, regional lymph node involvement and also distant metastasis.

Various histological types of urachal carcinoma are intestinal, mucinous, signet ring, unspecified adenocarcinoma and mixed histological type. 94% of urachal carcinomas are adenocarcinomas.[6] Our case was reported as grade 2 adenocarcinoma of enteric type with a mucinous component.

Regarding management strategy, the main therapeutic option is surgical for localized disease .Partial cystectomy with enbloc dissection or radical surgery appears to be the standard protocol. In our case partial cystectomy with enbloc dissection was done. Chemotherapy is preferred usually in metastatic setting.

Recent treatment modalities based on cytogenetic studies consists of targeted therapy using drugs like as cetuximab, and atezolizumab, with satisfactory responses, suggesting that personalized treatment could be the most suitable option for urachal carcinoma.[7]

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