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Malignant Mixed Mullerian Tumor of the Urinary Bladder in a 73 Year Old Woman

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Abstract
A giant tumor of the urinary bladder in a 73 year old female patient consisting of mesenchymal and epithelial parts turned out to be the first case of a Malignant Mixed Muellerian tumor of the urinary bladder.

Introduction

Non epthelial tumors of the urinary tract account for less than five percent of all vesical tumors [1]. Carcinosarcomas account for less than 0.1 %.

Carcinosarcoma and Malignant Mixed Muellerian Tumor (MMMT) are terms which are very often used interchangibly, but carcinosarcomas as highly malignant tumors contain malignant mesenchymal, chondrosarcoma or osteosarcoma, and epithelial elements which may be transitional cell carcinoma, squamous cell carcinoma, or adenocarcinoma [2]. These tumors are rare, usually occurring in middle-aged men, commonly presenting with gross, painless hematuria. The prognosis is uniformly poor despite aggressive treatment with cystectomy, radiation, and/or chemotherapy [1].

Malignant Mixed Mullerian Tumors (MMMT) are biphasic malignant tumors which enbody a mesenchymal and an epithelial part, originating from endometriosis. Their most common sites of manifestation are the uterus, ovaries, tubes and peritoneum. It has been reported that endometrioid carcinomas can be stimulated by estrogen or estrogen-agonistic therapy, e.g. treatment with tamoxifen [4]. We report what we think is the first report on MMMT in the urinary bladder.

Case Report

A 73 year- old caucasian woman presented to our hospital with abdominal pain and urinary retention. On physical examination we found swelling of the lower abdomen and edema of both legs. The abdomen and flanks were painfull to superficial and deep palpation. In the lower abdomen a huge mass was palpable and seemed to arise from the pelvis. She gave no history of haematuria or hormonal drug intake and had two uneventful pregnancies. Serum Creatinin level was elevated to 13 mg/dl.

Computed tomography (Fig.1) revealed a solid tumor of the urinary bladder, measurering 25 cm in diameter and hydronephrosis of both kidneys, rupture of the calyceal system on the right side and a thrombosis of the lower part of the inferior vena cava and

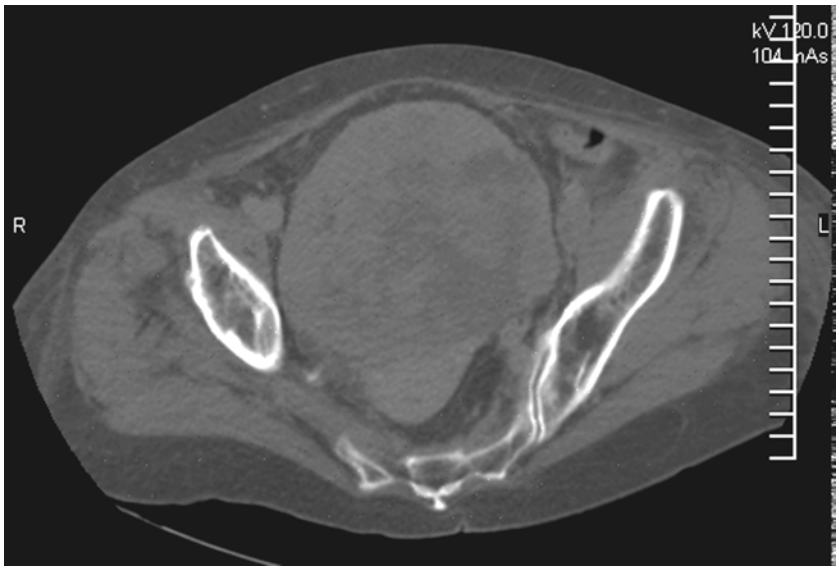

Fig. 1. Computed tomography revealed a solid tumor of the urinary bladder, measuring 25 cm in diameter. The uterus was visibly compressed dorsal of the tumor.

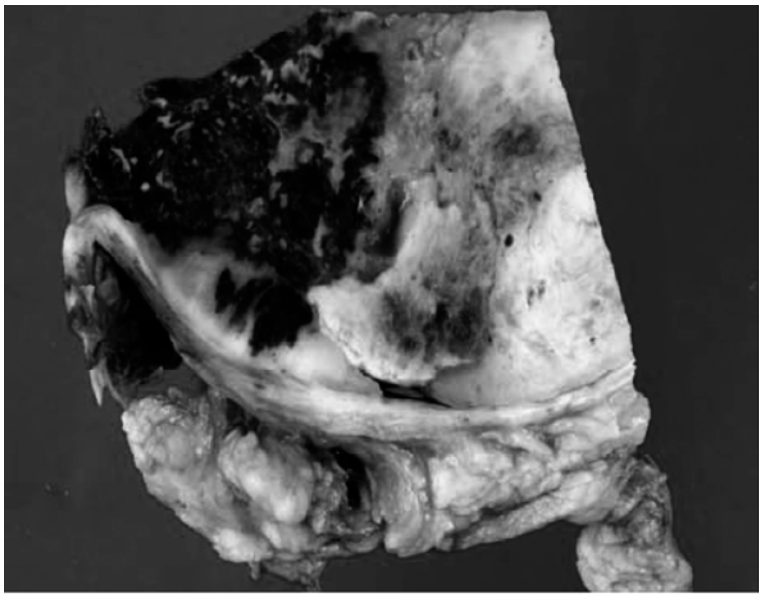

Fig. 2. Surgical specimen with solid tumor mass after cystectomy.

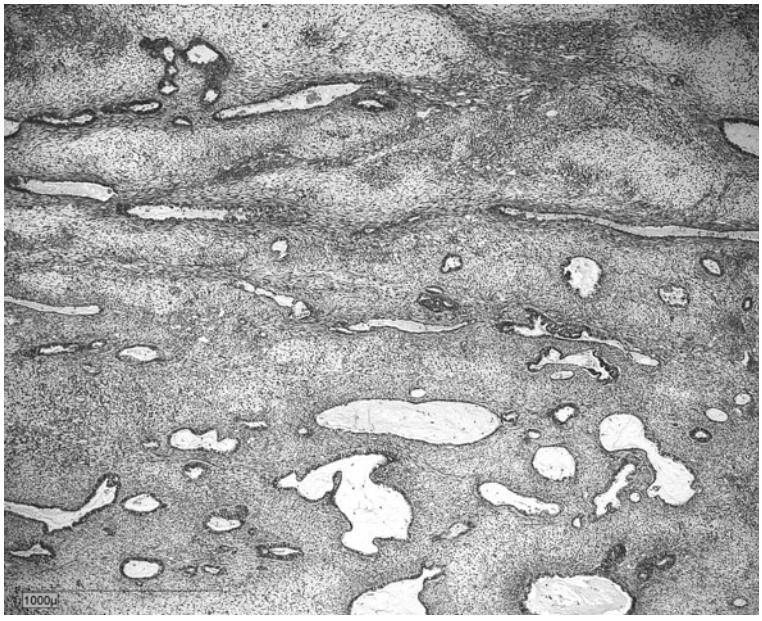

Fig. 3. Biphasic malignant tumor consisting of mucin forming neoplastic glands and mesenchymal component with spindle cells, rhabdomyoblasts and rhabdomyoblst-like tumor cells. The transitional cell epithelium was nearly completely replaced by a ciliated epithelium resembling Mullerian differentiation.

pelvic veins. The uterus was visibly compressed dorsal of the tumor. Uric cytology revealed inflammatory, but no malignant cells.

A percutaneous nephrostomy was placed into the right kidney and within ten days the serum creatinin values returned to normal values.

Explorative laparotomy revealed a large, but well defined tumor not infiltrating surrounding structures and a radical cystectomy and hysterectomy combined with an ileal conduit was performed. The postoperative period was uneventful.

The histological examination revealed a mitotically highly active, biphasic malignant tumor consisting of mucin forming neoplastic glands and a malignant mesenchymal component with

spindel cells, rhabdomyoblasts and rhabdomyoblast-like tumor cells. The transitional cell epithelium was nearly completely replaced by a ciliated epithelium resembling Mullerian differentiation (Fig. 3), which was, combined with the expression of CD 10 [5], considered diagnostic for MMMT. Residual endometriosis was not noted in the bladder wall, which might be due to destructive tumor growth. Histology of the uterus revealed a proliferated endometrium and myomas. The tumor was not penetrating beyond the superficial muscle layer of the bladder wall and a margin negative resection was achieved.

Comment

Since MMMTs are exceedingly rare, no standardized treatment has been defined. A comprehensive review of MMMT of the uterus, suggests that radical surgery followed by radiotherapy is the therapy of choice [3], increasing survival. The most common site of local recurrence is the pelvic peritoneum and paraaortic lymphatic nodes, the sites of distant recurrence are the abdomen, lungs and supradiaphragmatic lymphatic

nodes. A study from MD Anderson [3] showed that adjuvant radiotherapy of the pelvis can prolong the mean time to any distant relapse (17.3 vs. 7.0 months) in patients suffering from MMMT of the uterus, though the five year survival rates still remain poor and range from 33% to 39%. In chemotherapeutic trial favorable effects have been discribed for a liposomal doxorubicin and platinum based treatment of ovarian MMMT [4]. As urologists we should adopt these strategies, e.g. radical cystectomy and adjuvant radiotherapy, to offer our patients the best therapy available.

In our case the patients age, general state of health and personal preferences led us to agree on a watchful waiting strategy.

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