



## Oncology

## Clear Cell Adenocarcinoma of the Female Urethra, Mimicking Cystocele



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## ABSTRACT

Clear cell adenocarcinoma of the urethra is an extremely rare neoplasm mainly described in women. Anterior pelvic exenteration was the treatment performed in most reported cases. It seems to have poorer prognosis than urothelial carcinomas.

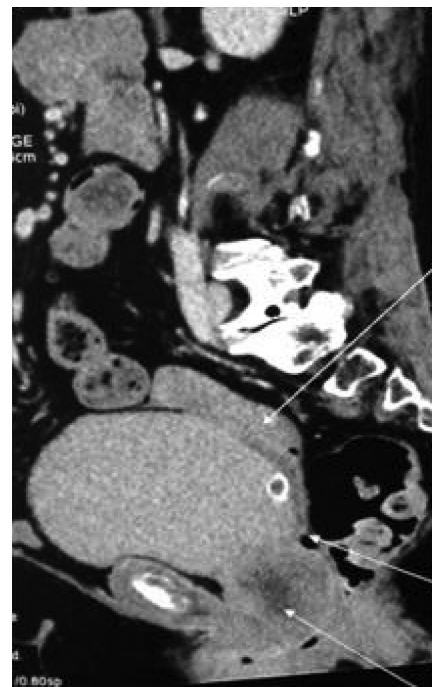
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## Introduction

Clear cell adenocarcinoma of the female urethra is poorly described in literature. In most cases, prognosis was poor even after pelvic exenteration. We report a new case in a 73 year old woman.

## Case presentation

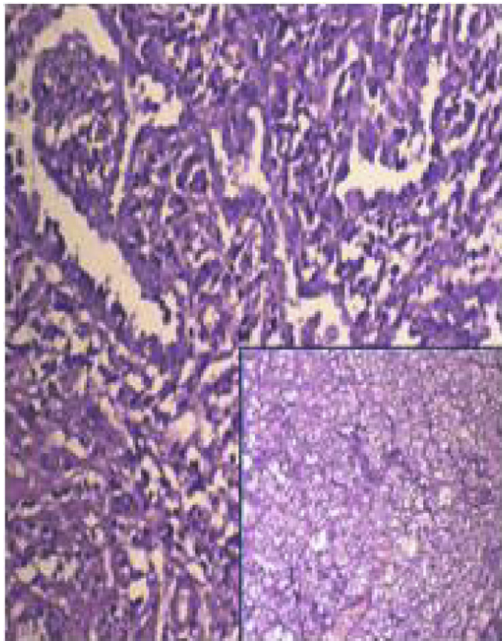
A 73-year-old woman, with a medical history of hypertension and diabetes, was referred for cystocele. She presented with dysuria and feeling of fullness in the vagina, with no gross hematuria. Physical examination revealed a hard palpable mass through the anterior vaginal wall. Urethrocystoscopy showed a circumferential tumor of the urethra, starting 2 cm from the external meatus and extending to the bladder neck. Transurethral resection of the tumor revealed a clear cell adenocarcinoma histology, penetrating the muscularis propria. Chest and abdomino-pelvic CT scan showed a localized circumferential tumor of the urethra, not invading the bladder, with no lymph node invasion or distant metastatic disease (Fig. 1).



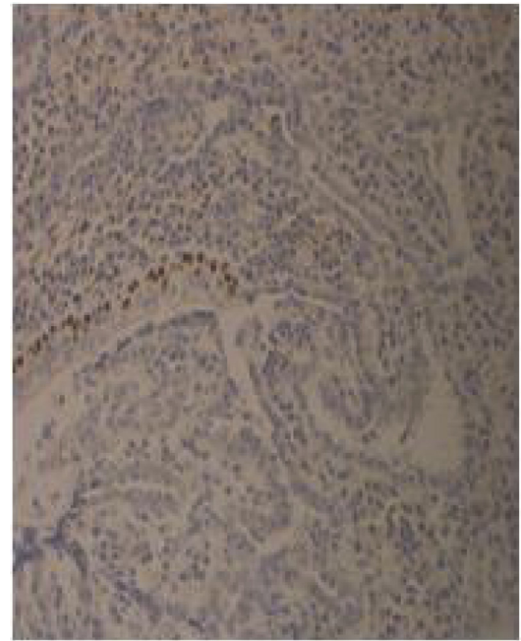
**Figure 1.** Sagittal CT scan showing circumferential and heterogeneous urethral tumor, not involving the bladder (arrows show uterus, vagina and urethral tumor).

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**Figure 2.** Definitive histology, H and E,  $\times 200$ , papillary and tubular tumor structures with epithelial lining with clear cells and focal hob-nail appearance. **Inset:** H and E,  $\times 400$ , solid areas were composed of clear cells.



**Figure 3.** Definitive histology with p63 immunohistochemical staining,  $\times 400$ , negative staining on tumor cells with a positive nuclear staining in the normal urothelial epithelium.

The patient underwent an anterior pelvic exenteration with an ileal conduit urinary diversion. Definitive histology, helped by negative p63 immunohistochemical staining, showed pT3 N0 clear cell adenocarcinoma of the urethra, with negative margins (Figs. 2, 3). The patient died from a massive pulmonary embolism on the fifteenth postoperative day.

## Discussion

Primary clear cell adenocarcinoma of the urethra is extremely rare, reported only in single case reports and small case series. It has been most commonly described in female urethra, with a mean age of 58 years.<sup>1</sup> Patients present with the same symptoms than the other urethral carcinomas, especially hematuria.<sup>1–3</sup> CT scan or MRI show a heterogeneous mass of the urethra, which may be localized in a diverticulum.<sup>4,5</sup> Diagnosis is made by urethroscopy with biopsies.<sup>5</sup> Microscopic examination reveals hobnail and flattened cells with abundant clear cytoplasm, moderate to marked nuclear pleomorphism with frequent mitotic figures.<sup>1,2</sup> Tumor cells may show positive immunohistochemical staining for PAX2, PAX8, cytokeratin 7, p16, p53, CA125, CAM5.2,

AE1/AE3.<sup>1,4</sup> In most reported cases, the treatment was anterior pelvic exenteration and pelvic lymph node dissection. According to the few cases reported, clear cell adenocarcinoma of the urethra seems to be an aggressive neoplasm with a survival rate less than 5 years in most cases.

## Conflicts of interest

None.

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