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Urethral clear cell carcinoma – Case report and review of litrature

Ali Al-Gonaim [a,*], Omar B. Alfraidi [b], Tariq Alotaibi [c], Moayad Saleh alqazlan [d], Turki Al-Hussain [d], Sultan S. Alkhateeb [e]

[a] Surgery Department, Prince Sattam Bin Abdulaziz University, Alkharj, Saudi Arabia
[b] King Abdulaziz Medical City, NGHA, Riyadh, Saudi Arabia
[c] King Faisal Specialist Hospital and Research Center, Urology Department, Riyadh, Saudi Arabia
[d] Department of Pathology and Laboratory Medicine, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia
[e] King Faisal Specialist Hospital and Research Center, College of Medicine, Alfaisal University, Riyadh, Saudi Arabia

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ABSTRACT

Urethral clear cell carcinoma is very rare disease affecting both sexes, however it is mostly described in female urethra. The origin of this cancer is yet to be discovered.

We report a 57 years old lady who presented to our clinic with obstructive lower urinary tract symptoms and found to have a urethral diverticulum containing a soft tissue lesion found to be a clear cell carcinoma after excision.

Having high suspicion and early detection of these cases leads to a better outcome.

Introduction

Primary female urethral malignancies are not common, accounting for less than 1% of genitourinary malignancies. Urethral clear cell carcinoma is actually a more rare disease affecting both sexes, however it is mostly described in female urethra on ratio of 1:4.[1] The origin of this cancer is yet to be discovered however few hypothesis were suggested in the literature including.

Mullerian origin, diverticular origin, or glandular differentiation of urothelium or urothelial carcinoma.[2] Here we report a 57 years old lady with obstructive lower urinary tract symptoms and weight loss which was diagnosed with urethral clear cell carcinoma.

Case

A 57 years old lady known case of hypertension and hypothyroidism. The patient was on her regular health until five months prior to her presentation to our clinic when she underwent dilatation and curettage for thickened endometrium outside our hospital and during the procedure there was a cyst in the posterior vaginal wall which was biopsied at that time and histopathology showed clear cell carcinoma, Mullerian-type tumor. The patient presented to our clinic complaining of obstructive voiding symptoms in the form of incomplete bladder emptying, frequency, hesitancy and history of urine retention once. The patient reported a history of weight loss, about 5 kg in the last 2 months. There was no history of hematuria, dysuria, and suprapubic or flank pain. Patient has no self or family history of cancer. Urinalysis were positive for +3 red blood cells and urine culture was negative. MRI abdomen and pelvic was done and was showing picture highly suggestive of a urethral diverticulum containing multiple stones and debris with suspicious soft tissue thickening and nodularity (Fig. 1). Due to the rarity of this disease the case was discussed in our tumor board and the decision was made to go for cystoscopy and CT chest, abdomen and pelvis (CAP) for staging. Bimanual examination revealed a large mobile lesion around 3–4 cm from the urethral meatus and extending into the bladder. Cystourethroscopy showed no obvious lesions seen in the bladder nor the urethra. Urine cytology was taken at time of cystourethroscopy which showed atypical urothelial cells keeping with carcinoma. CT CAP done which showed a periurethral cystic structure with soft tissue component, no evidence of locoregional or distant metastasis. The patient was counseled about the rarity of this disease and the lack of evidence in the management of such cases. She agreed to go for anterior pelvic exenteration, pelvic lymphadenectomy and ileal conduit creation. Histopathology resulted as clear cell carcinoma of the urethra, 3.8 cm in size, invading the anterior vagina, margins were negative and no lymphovascular invasion noted. Bladder, uterus, cervix, both ovaries, fallopian tubes and all lymph nodes were negative for malignancy (Figs. 2 and 3). Patient was followed up after 3 months with CT CAP which

* Corresponding author.
E-mail address: ali.algonaim@gmail.com (A. Al-Gonaim).

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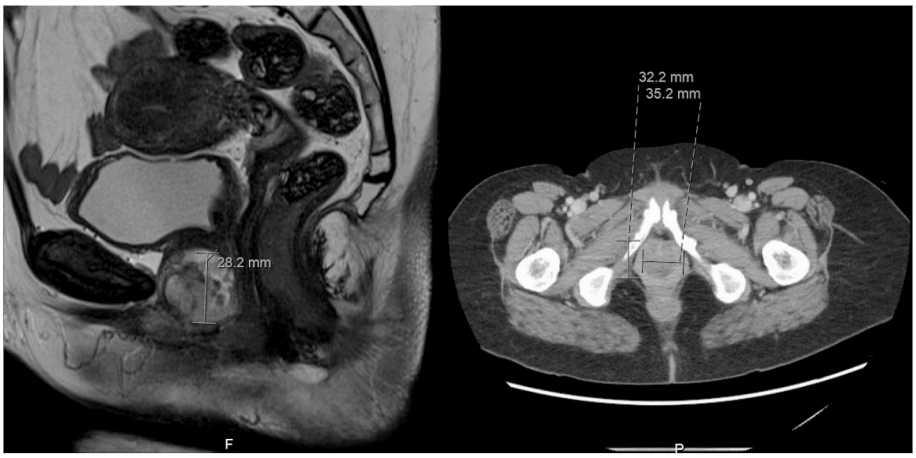

Fig. 1. Showing urethral diverticulum containing query multiple stones and debris with suspicious soft tissue thickening and nodularity.

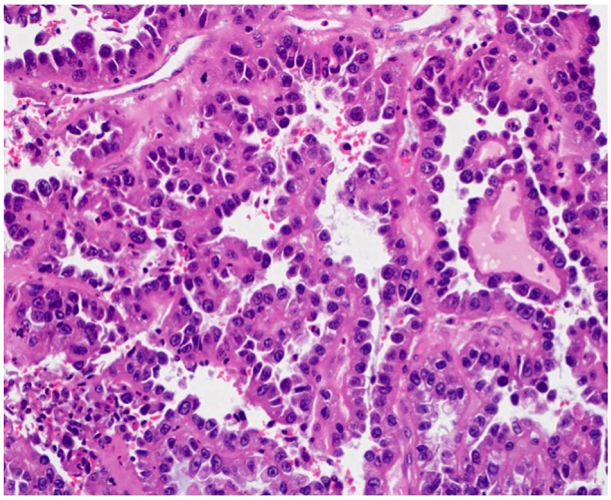

Fig. 2. Clear cell carcinoma with papillary growth pattern. Papillae with hyalinized fibrovascular cores are lined by hobnail cells with hyperchromatic nuclei (H&E, original magnification x20).

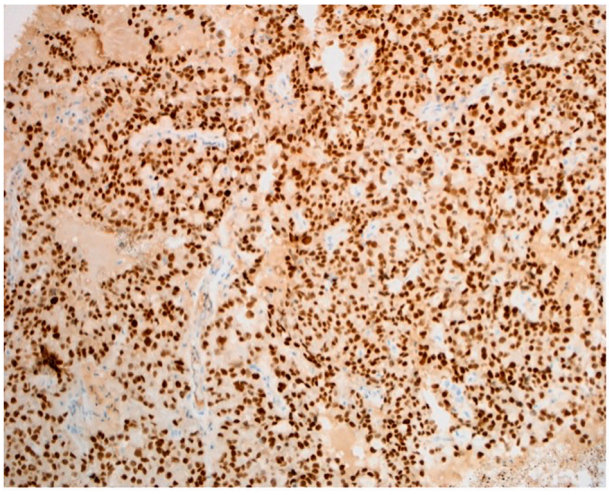

Fig. 3. PAX-8 is positive in the nuclei of tumor cells (Immunohistochemistry, original magnification 10x).

showed no signs of local recurrence or distant metastasis.

Discussion

Primary female urethral malignancies are not common, accounting for less than 1% of genitourinary malignancy with squamous cell carcinoma being the most common type histologically.[1] Urethral clear cell carcinoma is a rare type that occurs more frequently in female with female to male ratio of 4:1, furthermore, it approximately accounts for 0.003% of female genitourinary malignancy, with average age of occurrence 58 years.[3] There are no specific symptoms or signs for such cases, obstructive lower urinary tract symptoms with non-specific lower abdominal pain with or without hematuria should rise the suspicion of urethral carcinoma. Workup of such cases is not yet standardize, however Staging CT CAP to exclude metastasis and cystourethroscopy should be done, and MRI can be considered if CT was not diagnostic. Approximately only 40 cases have been reported since 1951.[4] The occurrence of urethral carcinoma is highly associated with urethral diverticulum disease whereas other types appears to be less common.[3] Clayton M et al. reported a 46–56% of urethral diverticular associated carcinoma were carcinoma and about 15–18% were squamous cell carcinoma.[5] The histological origin of primary urethral clear cell carcinoma is yet to be discovered, however multiple theories were suggested including Mullerian origin, diverticular origin, or glandular differentiation of urothelium or urothelial carcinoma.[2] In our case the patient was a lady who is 57 years old, which supports the epidemiological findings in the literature. Tumor was positive for PAX-8, CK7, CAM5.2, and P53 and negative for ER, PR, CK5/6, P63, and Napsin A, and urethral diverticulum was seen in the CT and MRI images although cystourethroscopy was negative for any lesions which supports the findings in the studies that urethral clear cell carcinoma arises more commonly in a diverticulum.

Conclusion

Urethral clear cell carcinoma is a very rare disease with no specific symptoms or signs, which may lead to delay in the diagnosis. The prognosis of this disease is bad in advanced cases and early detection and intervention leads to better outcome.

References

1. Drew PA, Murphy WM, Civantos F, Speights andVO. The histogenesis of clear cell adenocarcinoma of the lower urinary tract: case series and review of the literature. *Hum Pathol*. 1996;27(3):248–252.
2. Kato H, Ogihara S, Kobayashi Y, Toguri AG, Igawa Y, Nishizawa O. Carcinoembryonic antigen positive adenocarcinoma of a female urethral diverticulum: case report and review of the literature. *Int J Urol*. 1998;5(3):291–293.

3. Venyo AK-G. Clear cell adenocarcinoma of the urethra: review of the literature. *International Journal of Surgical Oncology*. 2015;2015:790235.
4. Cocco AE, MacLennan GT. Unusual female suburethral mass lesions. *J Urol*. 2005; 174:1106.
5. Clayton M, Siami P, Guinan P. Urethral diverticular carcinoma. *Cancer*. 1992;70: 665–670.