Case Report **OPEN ACCESS**

Clear cell adenocarcinoma of a female urethra: A case report and review of the literature

Amel Trabelsi, MD[1], Soumaya Ben Abdelkrim, MD[1], Soumaya Rammeh, MD[1], Wided Stita, MD[1], Nabil Ben Sorba, MD[2] Moncef Mokni, MD[1], Slim Ben Ahmed, MD[3], Sadok Korbi, MD[1]

Departments of Pathology[1] and Oncology[3], Farhat Hached Hospital, Sousse, Tunisia.
Department of Urology[2], Sahloul Hospital, Sousse, Tunisia.

Citation: Trabelsi A, Ben Abdelkrim S, Rammeh S, Stita W, Ben Sorba N, Mokni M, Ben Ahmed S, Korbi S. Clear cell adenocarcinoma of a female urethra: A case report and review of the literature. **North Am J Med Sci** 2009; 1: 321-323.
Availability: www.najms.org
ISSN: 1947 – 2714

Abstract

Context: Clear cell adenocarcinoma of the urethra is an extremely rare tumour. Its histogenetic derivation remains controversial. **Case report**: We report a new case of clear cell adenocarcinoma of the proximal urethra in a 56-year-old woman who presented with grossly hematuria. Urethral cystoscopy revealed a tumour protruding from the posterior urethral wall at the bladder neck. Treatment consisted of urethrocystectomy with pelvic lymph node dissection. Histologically, the neoplasm consisted of clear cell adenocarcinoma of the urethra. **Conclusion**: It appears that female urethral adenocarcinoma has more than one tissue of origin.

Keywords: Clear cell carcinoma, histopathology, urethra.

Correspondence to: Soumaya Ben Abdelkrim, Department of Pathology, Farhat Hached Hospital, Sousse, Tunisia. Tel.: 21699326546, Fax: 21673226702. Email: benabdelkrims@voila.fr

Introduction

Clear cell adenocarcinoma (CCA) of the female urethra is very rare; most information has been gained from single case reports and small case series [1]. We report a new case in a 57-year-old woman and discuss the clinico-pathologic pattern.

Case Report

A 56-year-old woman presented with gross hematuria. On physical examination, bleeding from the urethra meatus was seen. Urethral cystoscopy revealed a tumour protruding from the posterior urethral wall at the bladder neck. Computed tomography scan of the pelvis revealed a severe thickening of the bladder wall (Fig.1). The patient underwent transurethral biopsy of the tumour that showed an invasive poorly differentiated carcinoma of the urethra.

A total urethrocystectomy was performed including anterior vaginal wall and pelvic lymph node dissection. An ileal conduit was chosen for urinary diversion. Grossly, the tumour was measuring 2.5x2x2cm and invaded all the urethral layers. The bladder mucosa was not involved.

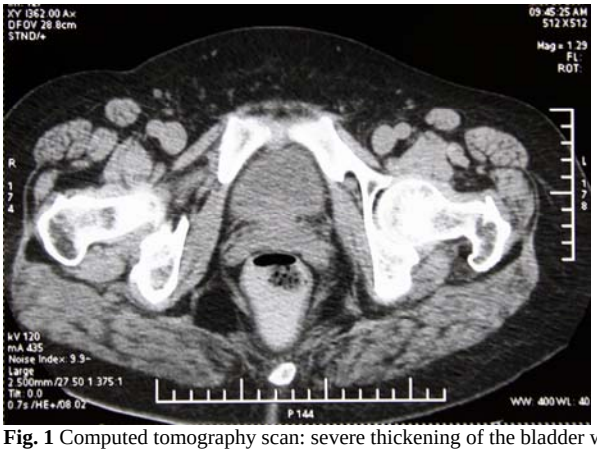

Fig. 1 Computed tomography scan: severe thickening of the bladder wall

Histological examination revealed a tumour composed of

nests and papillary structures (Fig.2) that were lined with cells having clearly cytoplasm with hobnail cells in some areas of the tumour (Fig 3); these cells showed severe cytologic atypia and high mitotic rate; tumour cells invaded all the urethral layers, but didn't involve the bladder.

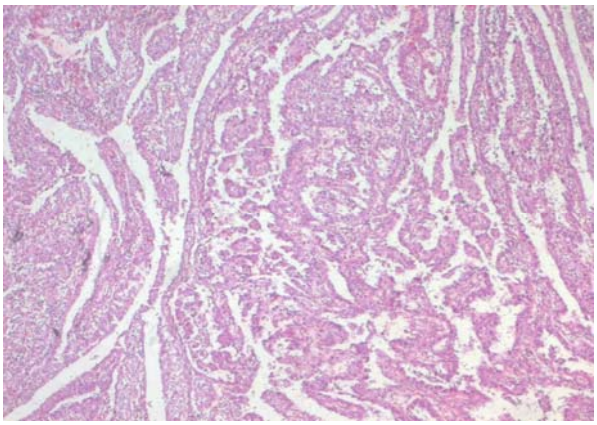

Fig. 2 Clear cell carcinoma composed of nests and papillary structures (HE x 40)

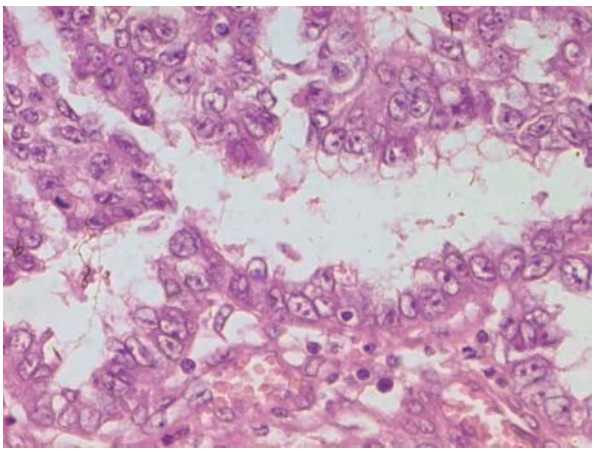

Fig. 3 papillary structures lined by cells with clearly cytoplasm and pleomorphic nuclei (HE x 400)

Immunohistochemical staining, using the two-step indirect imunoperoxydase technique with antibodies to prostate-specific antigen (PSA; DAKO, L-1838) showed no cytoplasmic reaction in the tumour cell. No lymph node metastasis was detected. The patient didn't receive any adjuvant therapy. She was free of disease three months after surgery.

Discussion

CCA of the urethra is an extremely rare tumour [2]. Most information has been gained from single case reports and small case series [1, 2, 3]. It mainly affects women and up to half of the cases develop in the context of a urethral diverticule [4, 5].

The histogenesis of CCA of the female urethra remains controversial [6]. Konnack [7] reported the first case in 1973, using the term "mesonephric carcinoma", and suggested that the tumour probably arises from the mesonephric duct or intermediate mesodermal vestiges. However, some authors [6] insisted on the mullerian origin of this tumour. In 1984, Pollen and Dreilinger [8] strongly supported the homogeneity between the female paraurethral duct and male prostate gland on finding positive immunohistochemical staining using antibodies to PSA (prostate-specific antigenin) and PAP (prostatic acid phosphatase). They have advocated that the tumour arises from the female para-urethral duct. In our case, tumour cells were negative for PSA. More recently, Zaviaci et al [9] reported a neoplasma with similar histologic appearance and immunohistochemical characteristics as adenocarcinoma of Skene's paraurethral glands and ducts.

The present findings support the theory that the female clear cell adenocarcinoma arises from the paraurethral duct [4]. However, it appears that female urethral adenocarcinoma has more than one tissue of origin with minority arising from the Skene's glands [10]. Morphologically, CCA of the urethral must be differentiated from nephrogenic adenoma of the urethra especially on biopsy. The predominance of clear cells, severe cytological atypia, high mitotic rate and necrosis favoured the diagnosis of CCA.

Because of the rarity of CCA in the urethra, the optimal treatment is unknown [2, 11]. It seems to be based on the localisation of the primary tumour and the presence of metastasis. Radical cystourethrectomy with or without irradiation was performed in most cases [11]. The response to chemotherapy is also unclear [11, 12]. In our case, the patient didn't receive any adjuvant therapy.

References

1. Drew PA, Murphy WM, Civantos F, Speights VO. The histogenesis of clear cell adenocarcinoma of the lower urinay tract: case series and review of the literature. Hum Path 1996; 27: 248-252.
2. Gögus C, Baltaci S, Orhan D, Yaman O. Clear cell adenocarcinoma of the male urethra. Int J Urol 2003; 10: 348-349.
3. Olivia EO, Young RH. Clear cell adenocarcinoma of the urethra. A clinicopathologic analysis of 19 cases. Mod Pathol 1996; 9: 513-520.
4. Abascal Junquera JM, Cecchini Rosell L, Martos Calvo R, et al. Presentation of a new case of clear cells urethral adenocarcinoma and its surgical management. Actas Urol Esp 2007; 31: 411-416.
5. Seballos RM, Rich RR. Clear cell adenocarcinoma arising from a urethral divertculum. J Urol 1995; 153: 1914-1915.
6. Kawano K, Yano M, Kitahara S, Yasuda K. Clear cell adenocarcinoma of the female urethra showing strong immunostaining for prostate-specific antigen. BJU int 2001; 87: 412-413.
7. Konnack JW. Mesonephric carcinoma involving the urethra. J Urol 1973; 110: 76-78.
8. Pollen JJ, Dreilinger A. Immunohistochemical identification of prostatic acid phosphatase and prostate-specific antigenin female periurethral glands. Urology 1984; 23: 303-304.

9. Zaviacic M, Sidlo J, Borovsk M. Prostate specific antigen and prostate specific acid phosphatase in adenocarcinoma of Skene's paraurethral glands and ducts. Virchows Arch A Pathol Anat Histopathol 1993; 423: 503-505.
10. Dodson MK, Cliby WA, Pettavel PP, Keeney GL, Podratz KC. Female urethral adenocarcinoma: evidence for more than one tissue of origin? Gynecol Oncol 1995; 59: 352-357.
11. Ebisumo S, Miyai M, Nagareda T. Clear cell adenocarcinoma of the female urethra showing positive staining with antibodies to prostate specific antigen and prostatic acid phosphatase. Urology 1995; 45: 682-685.
12. Maier U, Dorfinger K, Susani M. Clear cell adenocarcinoma of the female urethra. J Urol 1998; 160: 492-493.