



Case report

A case report: Female primary urethral carcinoma presenting as vulval mass

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ABSTRACT

Introduction: Primary urethral carcinoma is a rare disease with overall poorer outcomes in the past. It is relatively more uncommon in female sex.

Clinical presentation: We present a case of a female with primary urethral carcinoma that presented atypically as a vulval mass without any features of storage, voiding lower urinary tract symptoms, or gross haematuria.

Discussion: As primary urethral carcinoma is rare in occurrence, prospective studies are limited so as the recommendations to guide treatment options. Treatment recommendations are still on development from different small-scale studies as well as from data in higher volume centers. Management options depends on location, extent, histology of the lesion as well as on sex of the patient and fitness of the patient.

Conclusion: Early diagnosis and treatment with multidisciplinary consult and multimodality will improve the overall survival rate and quality of life of the patients.

1. Introduction

Primary urethral carcinomas are rare tumors that account for less than 1 % of all genitourinary malignancies, less than 1 % of female malignancies but usually of diagnostically challenging scenario with a poor prognosis. It is more common in men than in women [1]. It usually present with lower urinary tract symptom and haematuria. Moreover it rarely reported that urethral carcinoma presenting as vulval mass. The annual incidence rate increased with age to a peak in their 75-to-84-year age group. The histological types identified are transitional cell carcinoma which is the most common (55 %), squamous cell carcinoma (21.5 %) and adenocarcinoma (16.4 %) [4]. Risk factors include chronic inflammation, sexually transmitted disease, urethral stricture, history of radiation therapy, human papilloma virus and for female, urethral diverticula [2].

The average length of female urethra is about four centimeters. Except for its uppermost end, the urethra is embedded within the anterior vaginal wall [3]. Most of the lymphatics passes to the internal iliac nodes and some to external iliac nodes while the most distal parts can also drain to bilateral inguinal lymph nodes.

Cystourethroscopy and biopsy are diagnostic. If cystourethroscopy cannot be performed, other investigations such as retrograde

urethrography, urine cytology, and transurethral biopsies can be done. Cross-sectional imaging, such as computed tomography or magnetic resonance imaging, is needed to stage the disease. Management options are surgery or radiotherapy or chemotherapy or combination of these. This report was drafted in line with the SCARE 2020 criteria [5].

2. Case presentation

A 76-year-old previously apparently healthy patient presented with a vulval mass, dysuria, and whitish discharge for four months. On examination a hard mass around 3 × 3 cm with irregular surface and margin with purulent discharge was found surrounding the urethral orifice (Fig. 3B). Bimanual palpation, abdominal examination, bilateral inguinal lymph nodes, and other assessments yielded unremarkable findings.

The patient underwent a contrast-enhanced pelvic magnetic resonance imaging (MRI), which revealed a large enhancing circumferential mass measuring 3.8 × 4.7 × 6.4 cm within the urethra, causing a narrowed urethral orifice (Fig. 1). The mass exhibited intermediate signal intensity in both T1-weighted and T2-weighted images and showed infiltration into the base of the bladder. Additionally, multiple enlarged bilateral inguinal lymph nodes and a right iliac lymph node were

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detected.

A flexible cystourethroscopy was performed, identifying a tumor extending throughout the urethra up to the bladder neck (Fig. 2). A biopsy taken from the mass confirmed the presence of urothelial carcinoma with squamous metaplasia (Fig. 3A). A multidisciplinary team meeting involving urologists, radiologists, oncologists, and gynecologists was arranged and it was decided to proceed with local radiotherapy as a palliative care. Patient was then transferred to a specialized hospital for further management.

3. Discussion

Urethral cancer represents an exceedingly rare malignant condition, unique in its occurrence more frequently in females than in males within the genitourinary realm [6]. However, recent articles have indicated an upsurge in primary urethral carcinoma cases among males [7]. Since our case report focuses specifically on female urethral carcinoma, our subsequent discussion will center on this subject. Female urethral carcinoma exhibits a prevalence rate of a mere 0.02 % among all malignant tumors and comprises a mere 5 % of urological tumors affecting women [8]. The highest incidence is recorded within the population aged over 75 years [7], while urethral carcinomas are uncommon, they are associated with dismal outcomes. This can be attributed, in large part, to the anatomical characteristics of the female urethra, which is thin and short, thereby facilitating the local infiltration of malignant cells.

According to the esteemed RARECARE project, the 1- and 5-year overall survival (OS) rates of patients with primary urethral carcinoma in Europe stand at 71 % and 54 %, respectively [9]. The majority of histological classifications involve urothelial carcinoma, followed by squamous cell carcinoma, adenocarcinoma, and other subtypes [7]. Given the scarcity of cases within our population, comprehensive knowledge and management guidelines remain lacking.

Our patient presented with a vulval mass, dysuria and discharge. It is worth noting that presenting symptoms often lack specificity and can manifest as urethral bleeding or spotting, urinary obstructive symptoms, increased urinary frequency, palpable mass, urethral discharge, dyspareunia, recurrent urinary tract infections, and hematuria [6,10].

The lymphatic drainage of the proximal urethra typically proceeds towards the obturator and internal iliac nodes, while the distal urethra drains towards the inguinal lymph nodes [11]. In our case, no palpable inguinal nodes were detected; however, imaging revealed involvement of the lymph nodes. Early stages of the disease often exhibit local invasions, while distant metastases remain exceedingly uncommon. The most effective means of obtaining a pathological diagnosis involves acquiring a tissue sample through urethroscopy, a procedure that also aids in visualizing the tumor's extent. Management strategies are contingent upon the staging of the disease, as determined by the TNM (Tumor, Nodes, Metastasis) system, with overall survival outcomes

stratified according to the primary T stage. MRI with contrast enhancement stands as the preeminent imaging modality for loco regional staging, while contrast-enhanced CT scans of the chest and abdomen are employed to assess distant metastasis [12].

Treatment options are tailored based on staging and the available facilities. Viable treatment modalities encompass surgical resection, chemotherapy, radiotherapy (including external beam and brachytherapy) or combinations thereof, contingent upon tumor location and extent. In the case of distal urethral tumors, surgical interventions are typically more suitable for men, while radiation therapy represents a reasonable alternative for women. Radiotherapy emerges as an effective treatment modality for female urethral carcinoma, preserving normal anatomical structures and functionality. Brachytherapy has shown promise in enhancing local tumor control, possibly attributable to the delivery of higher radiation doses that can be safely administered. In one study, the addition of brachytherapy to external beam radiation therapy reduced the risk of local recurrence by a factor of 4.2 [13]. Routine inguinal or ilioinguinal lymphadenectomy may be considered for higher-stage tumors; however, the scarcity of data derived from primary urethral cancer treatment poses challenges in supporting this practice. Multimodal therapy is typically reserved for advanced stages, employed in conjunction with surgery to improve overall survival [14].

Surgical options range from transurethral resection to pelvic exenteration, entailing en bloc removal of the pubis and genitourinary diaphragm, with or without lymphadenectomy. When contemplating surgery, achieving a 2 cm disease-free margin is preferred, albeit challenging given the risk of encountering urinary incontinence, as the length of the female urethra spans a mere 3–4 cm [15,16]. In our patient's case, with the tumor extending throughout the urethra up to the bladder neck, local radiotherapy was recommended following a comprehensive MDT meeting comprising urologists, radiologists, oncologists, and gynecologists, in order to circumvent potential surgical complications. Follow-up care holds paramount importance in managing patients with primary urethral carcinoma and should be individualized based on specific risk factors. Regrettably, no definitive guidelines have systematically explored surveillance regimens. The European Association of Urology guidelines have merely recommended more extensive follow-up for those who undergo urethra-sparing surgery [17]. In one study, postoperative surveillance for primary urethral carcinoma encompassed outpatient visits, urinary cytology, urethroscopy, uroflow studies, and cross-sectional imaging at 3 to 6 months post-surgery, followed by subsequent evaluations every 6 months for a minimum of 2 years [18]. In summary, the rarity of this disease renders it challenging to acquire comprehensive knowledge regarding its characteristics and establish a definitive treatment strategy. Despite advancements in modern medicine, the recurrence rate for urethral carcinoma stands at a staggering 55 % despite treatment, and the 10-year survival rate remains a mere 60 % [19].

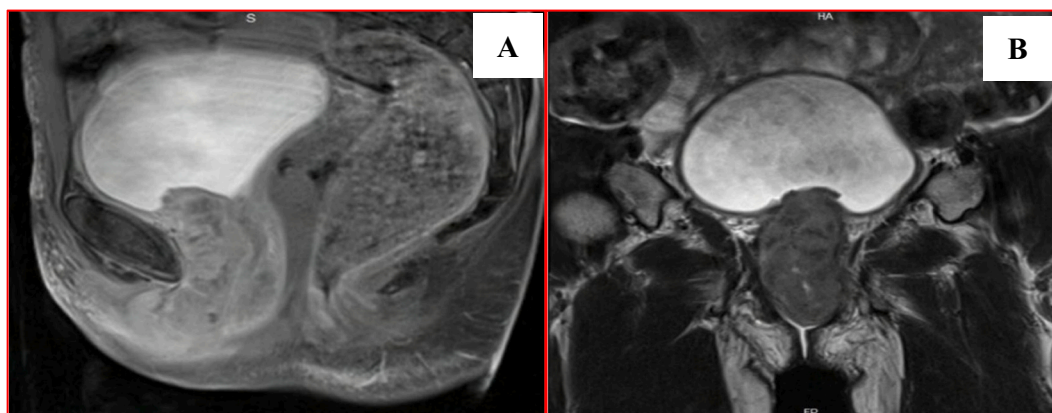


Fig. 1. MRI shows (A) sagittal section of urethral carcinoma extending throughout the urethra to the neck of the bladder (B) the coronal section of the lesion.

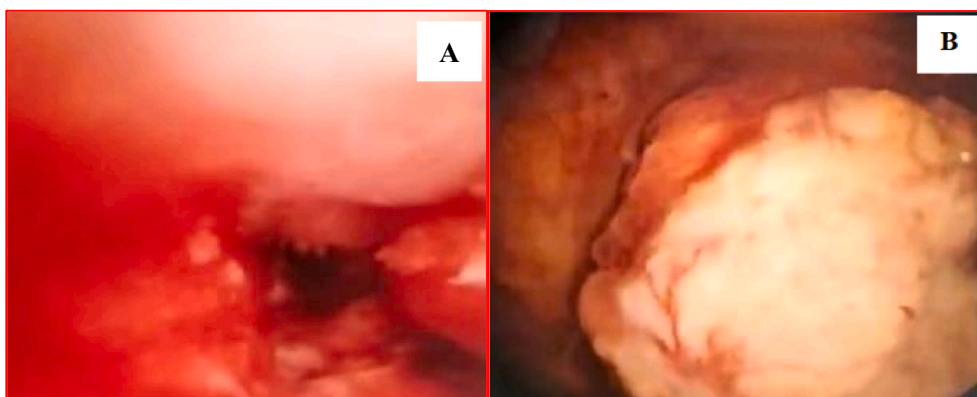


Fig. 2. (A) Cystourethroscopic view of abnormal urethral epithelium with near normal lumen size (B) intra-vesical view tumor extending protruding into the bladder.

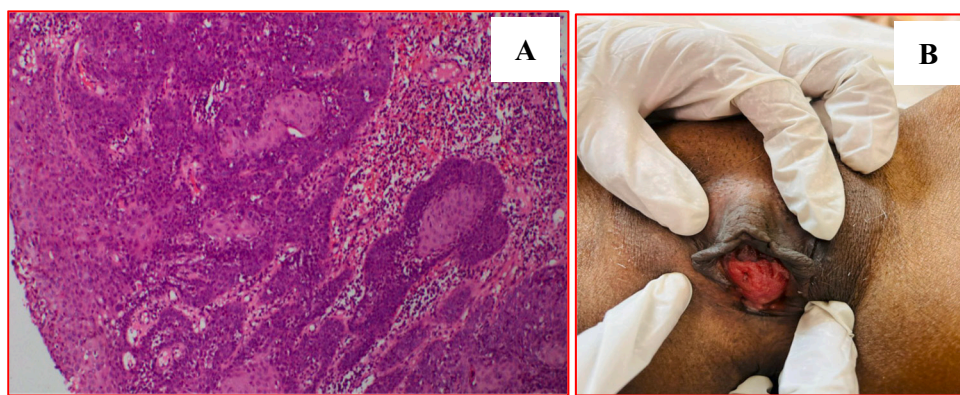


Fig. 3. (A) microscopic view of the biopsy from the lesion of transitional cell carcinoma with squamous differentiation (B) tumor surrounding the urethral orifice.

4. Conclusion

As primary urethral carcinoma is a rare entity especially in females a strong clinical suspicion of it should be raised whenever a patient presents with related symptoms and signs. Also, a valvular mass can be considered as one differential diagnosis in the case of urethral carcinoma. As it generally has poorer outcome and reduced quality of life following treatment early diagnosis and intervention will lead to improved survival rate and quality of life. We recommend further studies in the disease to improve the overall approach towards the pathology if encountered in the future.

CRedit authorship contribution statement

Study concept – Balagobi B.
Data collection – Nalini R, Theepan J and Chrishanthi R.
Data analysis – Abiharan P and Heerthikan K.

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None.

Ethical approval

The ethics clearance was not necessary to this study because our Institutional Review Board (IRB) does not require ethical approval for reporting individual cases or case series.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of this journal on request.

Guarantor

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Declaration of competing interest

There is no conflict of interest between the authors as everybody is aware of the work and participated actively and equally.

References

- [1] Mia A. Swartz, et al., Incidence of primary urethral carcinoma in the United States, *Urology* 68 (6) (2006) 1164–1168.
- [2] Farrell, M Ryan et al. “Current perspectives on the diagnosis and management of primary urethral cancer: a systematic review.” *Res. Rep. Urol.* vol. 13 325–334. 1 Jun. 2021.
- [3] C.S. Sinnatamby, R.J. Last, *Last's Anatomy: Regional and Applied*, Churchill Livingstone/Elsevier, Edinburgh, 2011.
- [4] Manju Aron, et al., Primary female urethral carcinoma: proposed staging modifications based on assessment of female urethral histology and analysis of a

- large series of female urethral carcinomas, *Am. J. Surg. Pathol.* 44 (12) (2020) 1591–1601.
- [5] R.A. Agha, T. Franchi, C. Sohrab, G. Mathew, A. Kirwan, A. Thomas, et al., The SCARE 2020 guideline: updating consensus Surgical Case Report (SCARE) guidelines, *Int. J. Surg.* 84 (1) (2020) 226–230.
- [6] V. Srinivas, S. Khan, Female urethral cancer—an overview, *Int. Urol. Nephrol.* 19 (1987) 423–427, <https://doi.org/10.1007/BF02550360>.
- [7] M. Wenzel, L. Nocera, C. Collà Ruvolo, C. Würnschimmel, Z. Tian, S.F. Shariat, F. Saad, A. Briganti, D. Tilki, P. Mandel, A. Becker, L. Kluth, F.K. Chun, P. I. Karakiewicz, Incidence rates and contemporary trends in primary urethral cancer, *Cancer Causes Control* (2021) 32, <https://doi.org/10.1007/s10552-021-01416-2>.
- [8] J.D. Carvalho Neto, S.C. Leão, R. Fakhouri, et al., Adenocarcinoma of the female urethra: a case report, *J. Bras. Patol. Med Lab.* 52 (4) (2016) 266–269.
- [9] Gatta M. Surveillance of rare cancers in Europe. <http://www.eurocare.it>. [Accessed 28 May 2023].
- [10] Z. Cope, C. Anglin, A. Dryden, J. Hawthorne, K.B.Y. Choi, Female urethral disease: a contemporary review of presentation, diagnosis, and current management strategies, *J. Surg. Open Access.* (2021) 7, <https://doi.org/10.16966/2470-0991.228>.
- [11] Kochergin M, Fahmy O, Asimakopoulos AD, Gakis G. Role of inguinal and pelvic lymph node dissection for primary urethral carcinoma: a systematic review. *Curr. Opin. Urol.* 2023; <https://doi.org/10.1097/MOU.0000000000001101>. DOI: <https://doi.org/10.1097/MOU.0000000000001101>.
- [12] S. Gourtsoyianni, T. Hudolin, E. Sala, D. Goldman, B.H. Bochner, H. Hricak, MRI at the completion of chemoradiotherapy can accurately evaluate the extent of disease in women with advanced urethral carcinoma undergoing anterior pelvic exenteration, *Clin. Radiol.* 66 (11) (2011) 1072–1078, <https://doi.org/10.1016/J.CRAD.2011.07.039>.
- [13] M.F. Milosevic, P.R. Warde, D. Banerjee, M.K. Gospodarowicz, M. McLean, P. A. Catton, et al., Urethral carcinoma in women: results of treatment with primary radiotherapy, *Radiother. Oncol.* 56 (2000) 29–35.
- [14] S. Traboulsi, J.A. Witjes, W. Kassouf, Contemporary management of primary distal urethral cancer, *Urol. Clin. North Am.* 43 (2016) 493–503, <https://doi.org/10.1016/j.ucl.2016.06.010>.
- [15] P. Narayan, B. Konety, Surgical treatment of female urethral carcinoma, *Urol. Clin. North Am.* 19 (2) (1992) 373–382 (PMID: 1574827).
- [16] R.J. Karnes, R.H. Breau, D.J. Lightner, Surgery for urethral cancer, *Urol. Clin. North Am.* 37 (3) (2010) 445–457, <https://doi.org/10.1016/j.ucl.2010.04.011> (PMID: 20674699).
- [17] Gakis G, Bruins HM, Cathomas R, Compérat EM, Cowan NC, van der Heijden AG, et al. EAU guidelines on primary urethral carcinoma. <https://uroweb.org/guidelines/primary-urethral-carcinoma>. [Accessed 2 March 2021].
- [18] C.C. Peyton, M. Azizi, J. Chipollini, C. Ercole, M. Fishman, S.M. Gilbert, et al., Survival outcomes associated with female primary urethral carcinoma: review of a single institutional experience, *Clin Genitourin Cancer.* 16 (2018) e1003–e1013, <https://doi.org/10.1016/j.clgc.2018.05.012>.
- [19] L. Dell'Atti, A.B. Galosi, Female urethra adenocarcinoma, *Clin Genitourin Cancer.* 16 (2) (2018) e263–e267, <https://doi.org/10.1016/j.clgc.2017.08.003>.