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Case Report

Advanced Primary Urethral Cancer Presenting as a Perineal Mass in Male

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ABSTRACT

Background: Primary urethral cancer is a rare cancer entity, and the main body of literature consists mainly of case reports, resulting in a limitation in clinicians' experience. Our patient was diagnosed with primary urethral cancer by cystoscopy and biopsy. He underwent total penectomy with B/L Orchiectomy with excision of perineal mass and adjuvant radiotherapy. Due to its rarity and lack of literature, there are no definite guidelines for managing primary urethral cancer. Surgical methods and postoperative adjuvant treatments vary in different hospitals, leading to diverse outcomes. We aimed to report an interesting PUC case managed by our department and provide a review of the available literature.

Case presentation: A 45-year-old male patient had painless gross hematuria and passing tissue bits during micturition for three months and painless perineal mass for two months. HPE of perurethral tissue bits revealed-Infiltrating Urothelial Carcinoma. The patient underwent total penectomy with B/L Orchiectomy with excision of perineal mass. Postoperative HPE revealed high-grade papillary urothelial carcinoma of the bulbar urethra with extensive necrosis. The patient was given adjuvant radiotherapy. Post-treatment follow up at nine months shows no local recurrence.

Conclusion: The prognosis in advanced stages is low, and those with proximal or whole urethral involvement require radical surgical procedures with neoadjuvant/adjuvant treatment. Distal urethral cancer has a relatively good prognosis and can be treated with surgery or radiotherapy. Advanced urethral cancer requires multimodal therapy and further clinical studies for determining optimal management guidelines. This case highlights the importance of reporting new cases and optimizing primary urethral cancer diagnosis to obtain an effective treatment.

Keywords: Primary urethral cancer; Surgery; Adjuvant radiotherapy

INTRODUCTION

Primary Urethral Cancer (PUC) is a rare malignancy and makes <1% of all genitourinary malignancies. PUC is three times more common in males than in females, and its incidence rises with increasing age >75 yr old [1-4]. Etiologies for PUC are chronic inflammation secondary to infection, radiation, urethral diverticula, chronic irritation of the urethra due to catheterization, and strictures [5-7]. Genital lichen sclerosis is reported as a potential risk factor for squamous cell carcinoma [8]. PUC has several histological subtypes arising from different cell types. On pathological examination, the predominant histological type is urothelial carcinoma 54% to 65%, squamous cell carcinoma 16% to 22%, and adenocarcinoma 10% to 16%. Other, rarer histological types include sarcoma or melanoma. Between male and female, subtypes are not evenly distributed and have different origins, which can be explained by different urethral anatomy [1,3]. The differences in origin and distribution have led to recent proposals for a new histological classification. In the future, the diagnosis, therapy, and follow-up will depend on the histological classification and anatomic specifications of each tumor. Most publications on PUC were based on small patient cohorts or case reports only. These results in limited knowledge on the optimal management of PUC compared with other malignancies. However, in recent years, larger, population based, and multicentric studies were published, providing new insights into this rare malignancy with a variable natural history. We aim to overview the current literature in context with the established knowledge on PUC to help guide current, optimized diagnosis and clinical management [8].

CASE STUDY

Case

A 45 yr male came with chief complaints of painless gross hematuria and passing tissue bits in urine for three months and painless perineal swelling since 2 months, rapidly progressive. Associated with dribbling of urine:

- No h/o DM/HTN/TB/ASTHMA/CAD/CVA
- H/o circumcision present, Trocar SPC+
- H/o Smoking and Alcoholism present
- P/A-soft, no tenderness, no palpable mass, BS+, all hernial orifices intact
- SPC+ in situ, draining clear urine

Local examination: Circumcised penis, Pinpoint meatus+, BXO changes present over glans penis, ventral shaft skin, and scrotum; Scrotum-B/L Testis: normal; Single Perineal Swelling of size approximately 20 ×10cm, no local rise of temp, no tenderness, hard to soft in consistency, mobility restricted in both planes, about 2 cm

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anterior to the anal verge, scrotal skin adherent to swelling on the left side; DRE-Anal tone: normal, no growth, anteriorly mass can be felt, Mucosa – normal, no other mass, no blood staining of the finger; No palpable B/L Inguinal Lymphadenopathy (Figure 1).

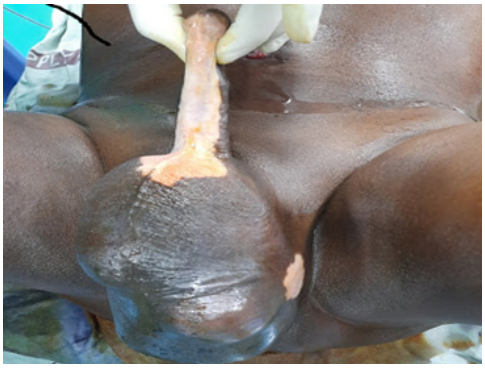

Figure 1: Clinical photograph.

Investigation

- Hb: 8.8gm%
- TLC: 10,000
- LFT: normal
- RFT-Serum creat: 1.3 mg/dl
- Serum electrolytes: Normal
- Coagulation profile:Inr-1.1
- Urine analysis (from SPC): No rbc
- Pus cells: 5-6/hpf
- Urine culture and Sensitivity: No growth
- Urine cytology: No malignant cells seen
- HPE of per urethral tissue bits: Infiltrating Urothelial Carcinoma
- USG Abdomen: B/L Kidneys: Normal; Urinary Bladder: SPC+ in situ; Liver and Spleen: Normal
- Chest Xray-normal

Cect abdomen and pelvis

Large heterogeneously enhancing lesion measuring 15.5 × 12.1 × 8.6 cm noted in the perineal region with loss of fat planes with posterior urethra and anterior rectal wall. The lesion is infiltrating the scrotal skin. B/L Testis: Normal. e/o few subcentimetric lymph nodes (Figures 2

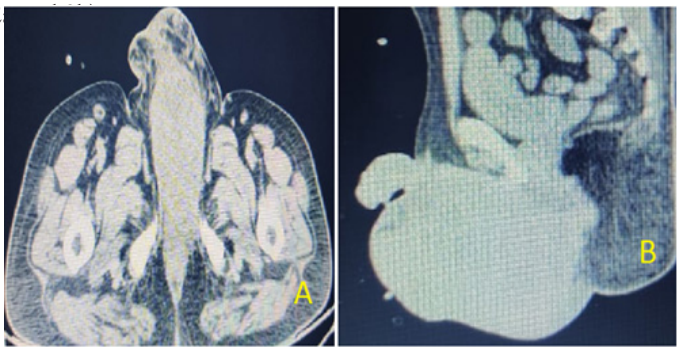

Figure 2: CECT Abdomen and pelvis –A: Axial view, B: Sagittal view shows large heterogeneously enhancing perineal mass with loss of fat planes with posterior urethra and anterior rectal wall.

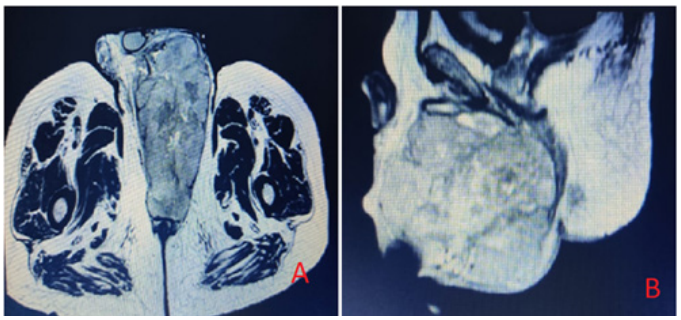

Figure 3: MRI of Abdomen and pelvis : A: Axial, B: Sagittal T2-weighted images demonstrate a T2-hyperintense soft tissue mass in the perineum with loss of fat plane between the lesion and anterior wall of the rectum.

Mri pelvis and abdomen

E/o large lobulated perineal mass of size 15 × 13.2 × 7 altered signal intensity (Hyperintense on T2 with the tiny cystic area and linear fibrotic strands, hypointense on T1) with peripheral T2 disrupted hypointense rim. The lesion is infiltrating the bulbar urethra. Loss of fat plane between the lesion and anterior wall of the rectum-infiltration. f/s/0 Malignant neoplasm: likely soft tissue sarcoma with infiltration of adjacent structures (Figures 3a and 3b).

Retrograde cystoscopy

The cystoscopy can be passed up to the bulbar urethra, growth (Figure 4).:

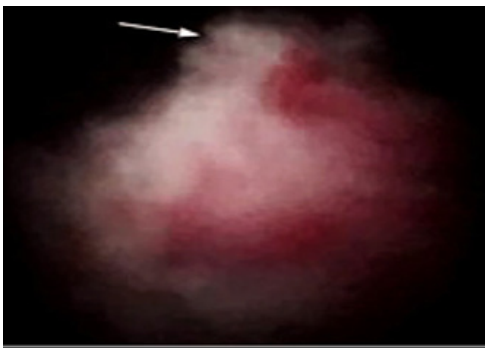

Figure 4: Retrograde cystoscopy: Showing growth in the bulbar urethra.

Antegrade cystoscopy

Urinary Bladder: Normal; Posterior urethra: Normal; Colonoscopy: Normal (Figure 5).

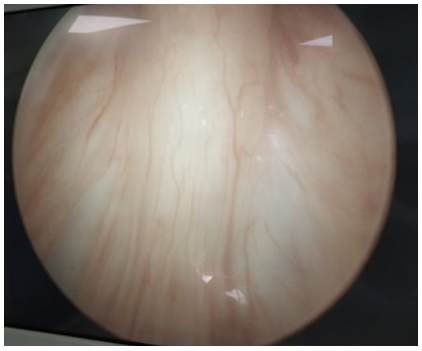

Figure 5: Antegrade cystoscopy: showing normal posterior urethra.

The patient was diagnosed as having stage T4N0M0 primary urothelial carcinoma of the urethra. The Patient underwent total penectomy with B/L Orchiectomy with excision of perineal mass. Primary closure of the wound was done. The post operational period was uneventful (Figures 6a and 6b).

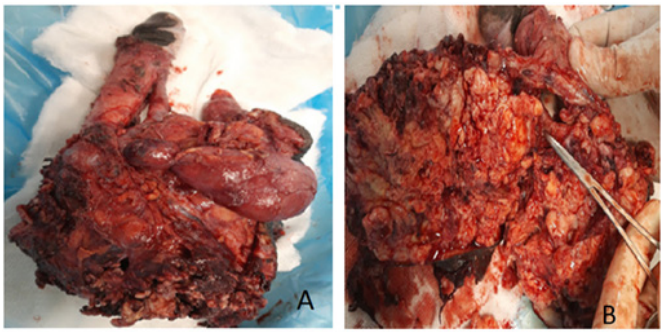

Figure 6: A: Complete surgical specimen, B: Cut section showing growth arising from the bulbar urethra.

Hpe of surgical specimen

Suggestive of high grade papillary urothelial carcinoma arising from the bulbar urethra. Adjuvant radiotherapy was given to the patient. The patient remains recurrence free at nine months follow up (Figures 7a and 7b).

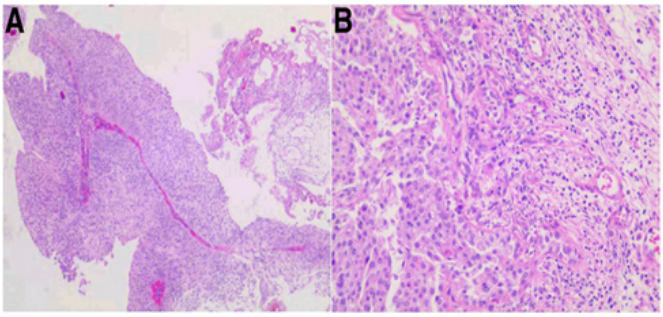

Figure 7: Postoperative HPE-low-power view of tumor, Hematoxylin and eosin staining, A: magnification,*40, B: magnification,*100.

DISCUSSION

Primary Carcinoma of the male urethra is uncommon, and no large scale experience with such cases has been published. The male to female ratio with PUC is 2.4:1 [9]. The risk factors of male and female PUC are different. PUC is mainly associated with the long-term repeated insertion of urethral catheter, urethral stricture, radiotherapy, and Human Papilloma Virus 16 infection in male patients. Main clinical symptoms in male are hematuria or urethral secretions (62%), dysuria (41%-48%), urethral stricture (76%), lower urinary tract irritation (20%), urethra cutaneous fistula (10%), and abscess (5%) [10]. In female, PUC is associated with factors such as recurrent urinary tract infection, urethral diverticulum, chronic infection of human papillomavirus, and other factors [11]. Clinical symptoms in female patients are urinary irritation, dyspareunia (70%), urinary tract infection, dysuria and hematuria [12]. During the physical examination, examine the external genitalia and perform a digital rectal examination. In female patients, do pelvic examination also. Examine bilateral inguinal lymph nodes, which can provide evidence of disease progression. The gold standard investigation for diagnosis is cystourethroscopy with biopsy of growth. With cystourethroscopy, note the location, size, shape, and invasion by tumor and concomitant bladder tumor presence. In PUC, the prognosis of patients depends on the tumor (T) and lymph node (N) staging and location of the primary growth [13]. The staging is based on the Tumor Node Metastasis (TNM) staging system. Tumors in the prostatic urethra have different staging. In the USA, the patients with PUC have survival rates of 46% and 29% of 5-10 years, compared with 68% and 60% for overall cancer 5-10 year survival rates [14]. In Europe, 1-5 year survival rates are 71% and 54% [15]. Various treatment modalities are surgery, radiation therapy, chemotherapy, and neoadjuvant chemotherapy. Monotherapy has a low therapeutic effect and a high recurrence rate, and multimodality treatment appears to be superior with distinct advantages. According to TNM staging, if a patient's tumor is in the stage of Ta or Tis, the patient can undergo chemotherapy. For chemotherapy, various drugs are platinum, hydroxycamptothecin, and some other drugs. Different histologic types of cancer have different chemotherapeutic infusion drugs. The recurrence rate after treatment is 28%-30%, which needs radical surgery and pelvic lymph node dissection, and urethral reconstruction [16,17]. Postoperatively, local radiotherapy can also be used, but it has complications, such as tissue necrosis, fistula, urethral stricture, and edema. The therapeutic effect is better if cancer is discovered early. Definitive treatment guidelines for advanced PUC are lacking, but most suggest multimodal therapy with a combination of surgery, chemotherapy, and radiotherapy, but this requires further clinical studies.

Recently, the role of immunohistochemistry in clinical practice is increasing, and it helps in: The diagnosis and identification of malignant neoplasm; to determine the tissue source of malignant neoplasm; to make the histopathological type of a tumor more exactly; to find a small metastatic focus or remnant of cancer; to determine the patient's prognosis; to provide a treatment plan for management. Medical development has its limits, and many diseases Immune Histo Chemistry (IHC) are not studied thoroughly. An online search in database revealed that no such specific antigen had been found for PUC. For the bladder urothelial cancer pathway, the expression of β-catenin was significantly increased, and it was involved in the occurrence and development of UC [18]. B-catenin overexpression may indicate that the UC has a poor prognosis, and so it can facilitate the development of targeted therapy [19]. Currently, NANOG and GATA3 are considered sensitive markers for UC and might be a potential biomarker for the early diagnosis of UC. However, further research is needed, and in the near future, it is believed that a PUC-specific marker will be found.

CONCLUSION

For advanced PUC, currently, multimodal therapy is used, which requires further clinical research. In our patient, a radical cystoprostatectomy with pelvic lymphadenectomy with the urinary diversion with or without total penectomy should have been done, but the patient chooses total penectomy with B/L Orchiectomy with excision of perineal mass and urinary bladder preservation with permanent suprapubic catheter followed by adjuvant radiotherapy. Although this treatment may not be curative for him, it did provide symptomatic relief for the patient, and he remains recurrence free at nine months of follow-up.

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