Case Report

Malignant priapism: case report and update on management protocols

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Background: Malignant priapism, a rare disease with only about 500 reported cases to date, consists of persistent erection secondary to invasion or metastasis of a primary neoplasm. While treatment guidelines for priapism in non-malignant cases have been established, there is currently no guideline for treating malignant priapism. Herein, we describe three cases of malignant priapism and suggest a step-by-step approach for clinical management.

Case Description: This study reports three cases of malignant priapism resulting from advanced genitourinary cancers. All patients experienced a sub-acute progression of penile pain and ultimately underwent palliative penectomy, resulting in sustained symptom relief.

Conclusions: Treatment of malignant priapism needs to be individualized to the needs of the patient. No matter the primary or secondary nature of the disease, current data suggest that malignant priapism is associated with poor outcomes and emphasis should be put on palliative care. Similar to previous cases, our cases died shortly after the diagnosis of malignant priapism. Conventional procedures such as shunting may not necessarily provide symptom relief in these patients. Although new radiation techniques have shown favorable outcomes, penectomy should be considered the last resort in clinical management. Revisions to the existing management guidelines for priapism are necessary to address its occurrence in malignant contexts.

Keywords: Priapism; malignant priapism; urogenital neoplasm; genitourinary neoplasm; case report

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Introduction

Malignant priapism, defined as primary penile involvement of malignancy or metastasis to the penis causing clinical priapism, is a rare disease with approximately 500 reported cases to date (1). Penile metastases arise from the genitourinary tract 64% of the time (bladder: 28.6%, prostate: 27.9%, kidney: 6.9%, ureter: 0.5%) with colorectal adenocarcinoma, lung cancer, melanoma, and hematologic diseases comprising the remainder. Malignant priapism is reported to occur in 20–53% of patients with penile metastasis (1).

The mechanism of malignant priapism remains unknown. The most accepted theory is an invasion of the corpus cavernosum as well as associated venous systems

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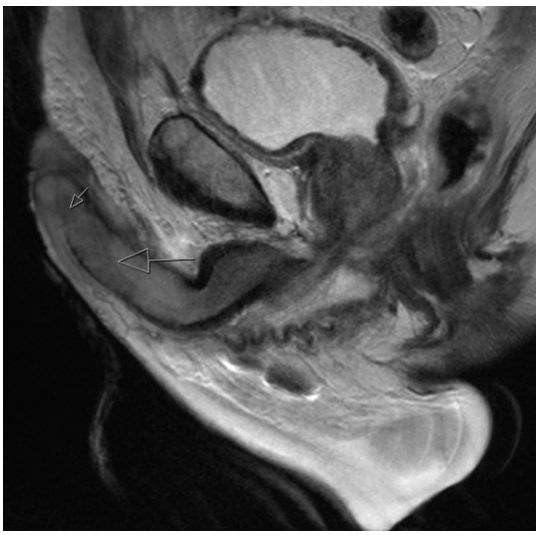

Figure 1 T2 sagittal view MRI of the pelvis; arrows indicate borders of metastatic corporal involvement. MRI, magnetic resonance imaging.

with malignant cells. Priapism can occur as a result of either hematogenous or lymphatic spread from the source tumor, leading to a derangement of arterial or veno-occlusive mechanisms. Both ischemic and non-ischemic priapism can occur (2).

Malignant priapism is indicative of stage 4 cancer and is associated with a poor prognosis. Life expectancy is less than one year (3).

Herein, we describe three cases with advanced genitourinary cancer, which presented with metastatic lesions to the penile corpora and subsequently underwent penectomy as palliative care for low-flow malignant priapism. We present this article in accordance with the CARE reporting checklist (available at https://tau.amegroups.com/article/view/10.21037/tau-23-327/rc).

Highlight box

Key findings
- Malignant priapism is associated with poor prognosis.
- Therapeutic approach using current guidelines may not result in favorable outcomes.

What is known and what is new?
- Penectomy has shown definite pain relief.
- New palliative radiation techniques may yield favorable results.

What is the implication, and what should change now?
- Malignant priapism requires a distinct management protocol.
- Management of priapism in malignant cases should be personalized.

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patients for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Case 1

Patient 1 was a 59-year-old male with a history of urothelial carcinoma of the bladder who underwent a radical cystoprostatectomy with ileal conduit and pelvic lymph node dissection after neoadjuvant chemotherapy (final pathology T3a N2). His postoperative course was complicated by a small bowel obstruction 1-month post-op that resolved with conservative management. Two months after the cystectomy he presented with a two-week history of penile pain and burning with a rigid penis concerning for priapism. His exam was notable for a rigid penis consistent with ischemic priapism and there were no palpable masses.

Pelvic magnetic resonance imaging (MRI) confirmed metastatic disease in the distal right penile corpora with associated malignant priapism (*Figure 1*). The priapism was resolved initially with conservative management by treating the patient's pain with a patient-controlled analgesia (PCA) pump. He was transitioned to oral pain medications and discharged. He had two hospitalizations shortly thereafter for uncontrolled penile pain treated with dorsal penile nerve penile blocks.

Two days after his most recent discharge, the patient presented for follow-up in the urology clinic and had an increasingly firm and tender penis on exam. The decision was made to perform a palliative penectomy at that time. His hospital course was uncomplicated and he was discharged on post-op day 3. His pain was much improved and he was satisfied with the pain relief.

Following his penectomy, the patient had a number of subsequent admissions related to his metastatic disease, but unrelated to his penectomy. He ultimately passed away about two months after his penectomy.

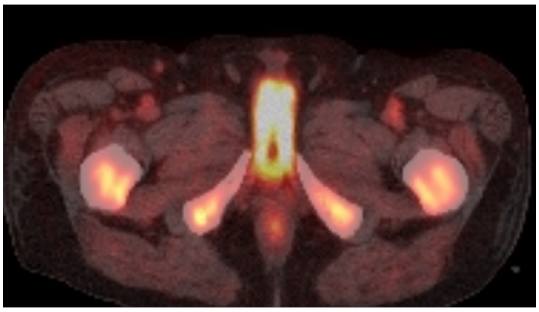

Figure 2 PET scan demonstrating the bilateral extension of prostatic tumor into the corpora cavernosa. PET, positron emission tomography.

Case 2

Patient 2 was a 71-year-old male with a history of metastatic urothelial carcinoma of the bladder treated with a palliative cystoprostatectomy due to refractory hematuria (final pathology T4 N2). This was followed by 4 cycles of adjuvant gemcitabine and cisplatin, and palliative radiation to the pubic rami for pain. Seven months after surgery, he presented to his medical oncologist with penile and perineal pain and was thought to have radiation dermatitis. His pain worsened despite treatment with topical steroids and he presented to urology two months later with a sustained erection and tenderness to palpation. His erection had been rigid for several days prior to presentation. There were no palpable masses.

He was admitted and an MRI was performed, showing a 1.2 cm lesion in the bulb of the corpus spongiosum. Ultrasound with Doppler also supported the diagnosis of corpus spongiosum metastasis but revealed patent cavernosal arteries and the dorsal vein of the penis. Initial penile blood aspirate revealed dark blood consistent with ischemic change. Penile blood gas was significant for pH of 7.32, pCO_2 of 57, and pO_2 of 46, consistent with ischemic priapism. He underwent a penile biopsy and distal shunt procedure. Pathology confirmed metastatic urothelial carcinoma in the glans and corpus spongiosum.

His glans started to become necrotic post-op day 1 and he returned to the operation room (OR) on post-op day 3 for a penectomy. Final pathology was significant for metastatic urothelial carcinoma with lymphovascular invasion and positive margins. His immediate hospital course was uncomplicated, and he was discharged on post-op day 5 with much improved pain. He was placed on hospice care about a month later and did not receive any further radiation or chemotherapy. He passed away shortly thereafter, approximately 2 months after his penectomy.

Case 3

Patient 3 was a 77-year-old male with a history of locally advanced prostate cancer treated with external beam radiation therapy in 1996. He developed a biochemical recurrence over 20 years after his initial radiation. Subsequent prostate MRI demonstrated the prostate was replaced by tumor spreading into the seminal vesicles and base of the bladder.

Shortly thereafter he was admitted several times with recalcitrant hematuria. This culminated with him undergoing bilateral nephrostomy tube placement, prostate artery embolization, and multiple fulgurations of the prostate in the operating room. After resolution of his hematuria he presented with a rigid erection for several days. A positron emission tomography/computed tomography (PET/CT) scan demonstrated a prostate tumor extending the length of the bilateral corpora cavernosa.

His priapism was initially managed with penile ring blocks, though he continued to have significant pain. After a lengthy discussion of options the patient then pursued a combined radical cystoprostatectomy, radical penectomy, bilateral orchiectomy, and ileal conduit urinary diversion for management of his recurrent hematuria and malignant priapism. The pain attributed to malignant priapism resolved following the penectomy.

Pathology demonstrated residual prostate adenocarcinoma extending to involve bilateral seminal vesicles, bladder, periureteral soft tissue, periurethral soft tissue, and corpus cavernosum (*Figure 2*). Shortly after surgery developed metastatic disease to his bony pelvis. Over a period of months the patient slowly declined. He ultimately decided to go under hospice care and passed away shortly thereafter (*Figure 3*).

Discussion

Priapism is defined as any type of sustained erection erections that do not arise from sexual stimulation and do not respond to conventional abortive procedures after four hours. Conventionally, priapism is classified into three distinct groups namely high-flow (non-ischemic), low-flow (ischemic), and stuttering priapism (4). However, the pathophysiology which different malignancies may result in priapism varies. For instance, conditions with excessive

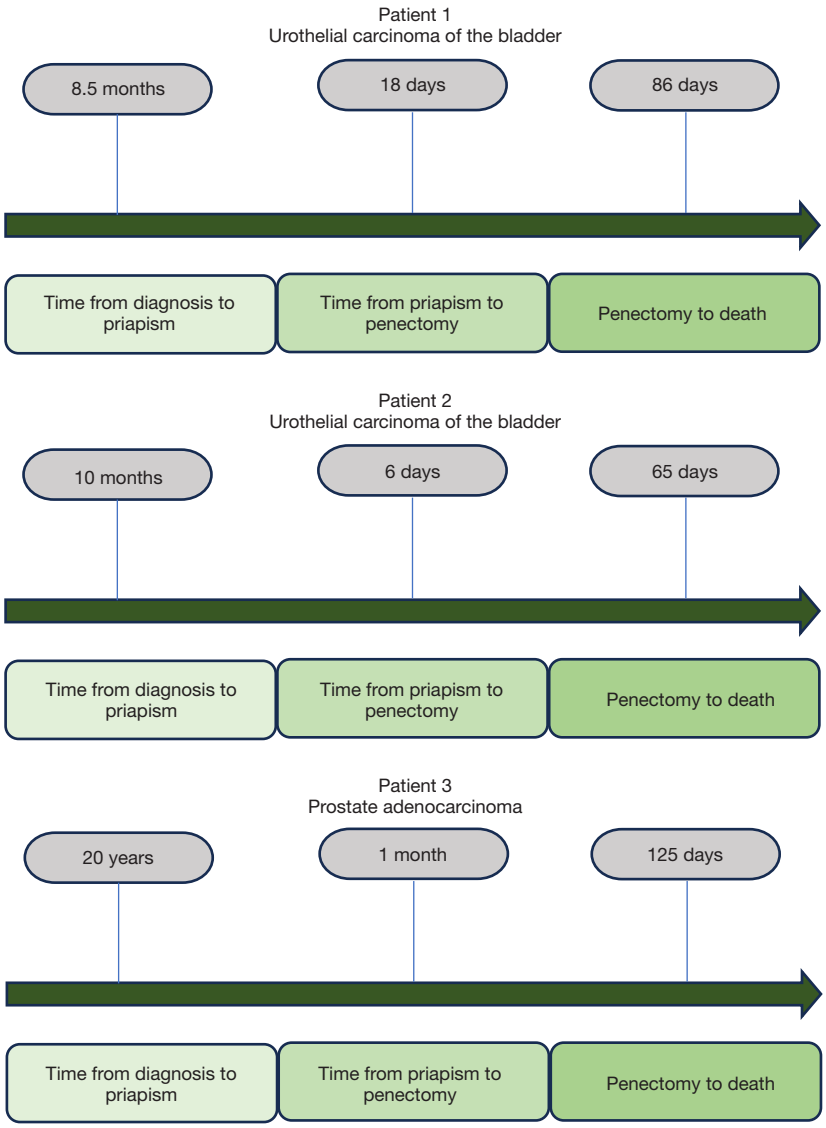

Figure 3 Timeline of disease progression; note that the schematic has been simplified to portray the duration of each period.

white blood cell content, hematologic dyscrasias and leukemias may lead to priapism due to hyperviscosity of blood within the corpus cavernosum and accentuation of the cavernous outflow (5). Malignant priapism, first described by Peacock in 1938, collectively refers to the priapism arising from solid tumor infiltration to the cavernosal system (6). The majority of cases are ischemic in nature secondary to primary or metastatic tumor involvement (7,8). Although rare, high flow non-ischemic etiologies could also be found in malignant priapism (9). It is imperative to know that malignant priapism is often associated with end stage cancer and a short life expectancy. Similar to other studies, our patients died shortly after initial presentation of malignant priapism. A literature review of 400 cases has shown that the average life expectancy of malignant priapism patients is 9 months (10). However, life expectancy should be judged mostly based on the etiology of cancer, namely primary or metastatic carcinoma (11,12). Primary penile cancer has shown a 5-year life expectancy rate of 67.7% and 65.67% between 2000 and 2014 (13). Despite these numbers, case reports have shown that malignant priapism in the case of primary penile cancer is extremely rare and is associated with poor outcomes and a short life expectancy (14). In one case with primary penile lymphoma

and malignant priapism, pain control was achieved using one episode of E-CHOP regimen (cyclophosphamide, vincristine, prednisone, epirubicin and etoposide) (15). Unal *et al.* resorted to penectomy for pain alleviation in a case of primary penile squamous cell cancer-related malignant priapism (7).

Reports indicate that malignant priapism in the setting of metastatic carcinoma has been controlled with palliative radiation therapy, chemotherapy and palliative penectomy. However, penectomy was seen as the last resort in numerous cases. One report indicated that a patient with prostate cancer achieved symptom relief using volumetric modulated arc therapy (VMAT) (16). Hormonal therapies including antiandrogens, gonadotropin-releasing hormone (GnRH) agonists and 5α-reductase inhibitors are also an emerging modality of treatment in priapism patients. However, their effectiveness in such advanced cases of malignant priapism is yet unknown (17).

Considering the advanced age at initial presentation and poor life expectancy in most of these patients, we suggest that the management of malignant priapism should be decided on a case-by-case basis and focus on end-of-life care and pain reduction rather than preserving erectile function. The management protocols regarding malignant priapism should be based on the pathology's ischemic (low flow) or non-ischemic (high flow) nature. Priapism due to hematologic cancers could be treated using local and systemic therapies. However, the management of ischemic malignant priapism is a topic for which we believe an update is needed for better clinical care.

An algorithm for the management of ischemic priapism has been developed by the American Urological Association. In the case of patients with underlying malignancy, as noted in the guidelines, it is important that systemic treatment should not be undertaken as the only treatment for the priapism. Priapism is typically treated in a stepwise manner with therapeutic aspiration, intracavernosal injection of sympathomimetic drugs, and ultimately shunting procedures (18). We have found that this algorithm is not as successful when the cause of priapism is malignancy. Similar to our experience, other reports have indicated the inefficacy of this step-wise approach in dealing with malignant priapism. It was observed in a patient with malignant melanoma and malignant priapism that treatment with narcotic analgesics did not provide sufficient pain relief. Also, surgical shunting of the cavernous system did not result in complete relief of tumescence. Therefore, neurolysis of the dorsal penile nerve was done under the guide of sonography which significantly decreased the pain. In line with our findings, the patient in the mentioned report died after two months of malignant priapism presentation (19). Our experience, however, shows that penile ring block may not achieve satisfactory and long-lasting pain reduction.

Because of the rarity of malignant priapism, the effectiveness of the above algorithm is not proven in this population. For this reason, treatment may need to be adjusted case by case based on clinical judgement. For priapism patients with a history of cancer, we recommend obtaining a pelvic MRI to assess for the presence of metastatic lesions obstructing the vascular drainage of the penis. We have also found PET/CT to be useful in demonstrating the distal extent of the disease in the penis. After confirming malignancy as the cause of the priapism, we have found conservative management with pain control, including PCA pumps as well as penile blocks were unsuccessful in controlling pain. We also found priapism to be inadequately treated with shunting procedures. This is likely because, unlike in traditional priapism cases, these treatments do not address the underlying cause of the priapism. The ineffectiveness of shunt procedures may be attributed to their failure to address the underlying cause of the problem. As the tumor continues to invade the corpora, the shunt is unlikely to succeed and may exacerbate the pain, as was observed in our case. One of our cases also developed penile necrosis following the shunt procedure which may be attributed to the rapid growth of neoplasm. Platelet dysfunction in malignancies may also lead to inadequate outcomes with penile shunts (20,21). However, there have been cases in which shunting resulted in temporary pain relief and incomplete resolution of erection that ultimately required total penectomy (2). Given the etiology of malignant priapism, we have found palliative penectomies to be the most helpful solution to achieve lasting symptomatic resolution.

Based on our experience we have proposed a new algorithm for management of priapism specific to priapism caused by malignancy. First, a thorough history and physical exam would be needed. There needs to be a high index of suspicion for malignant priapism in patients with a known history of cancer, particularly of genitourinary. It would also be unusual for an older man without a previous history of priapism to have an unprovoked (not on erectile dysfunction medications) priapism. Although most malignant priapism is ischemic, there have been reports of high flow priapism secondary to malignancy (9,12). Because

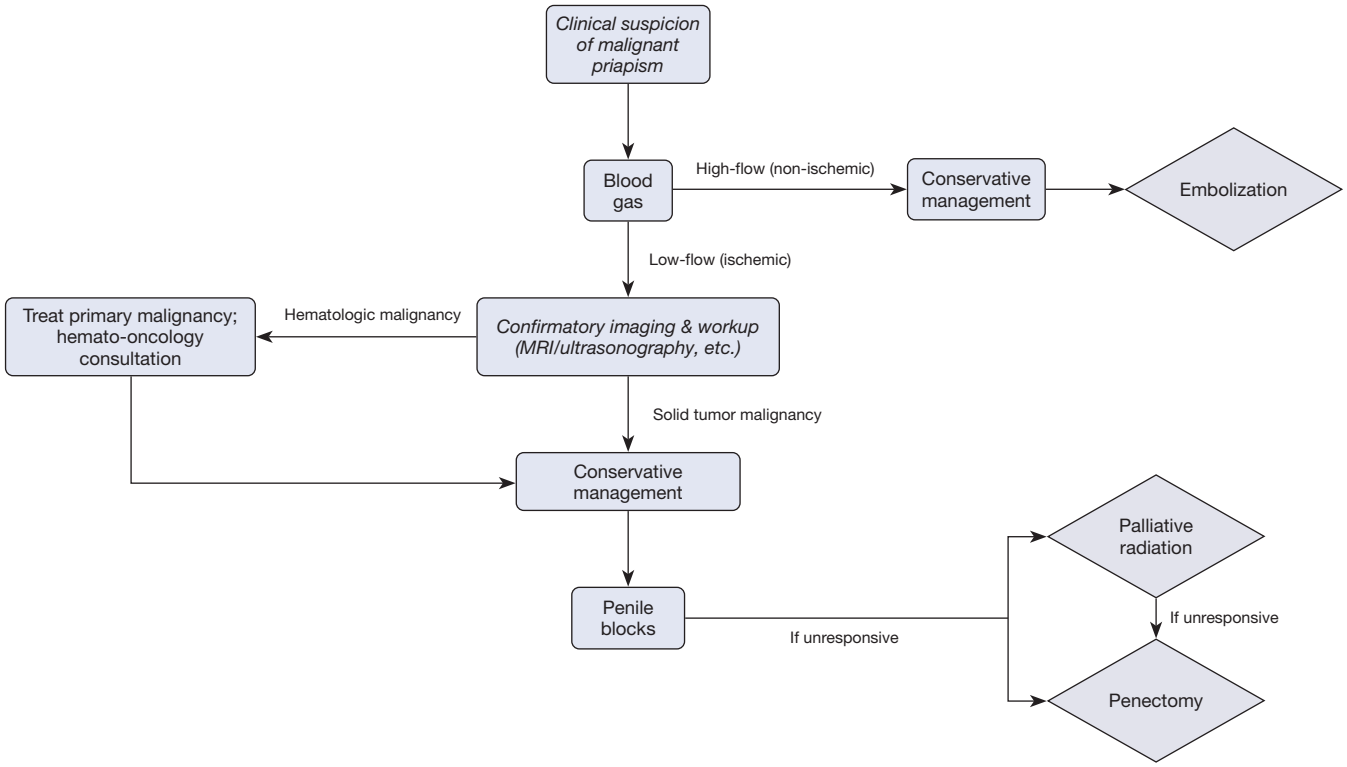

Figure 4 Suggested flowchart for treating malignant priapism.

of this, we recommend starting with penile blood gas, as is recommended in the standard priapism guidelines (*Figure 4*).

Conclusions

Metastatic lesions of the penis causing priapism can be managed through a number of different therapies, including conservative therapy, shunting, or penectomy. The presence of malignant priapism indicates advanced disease with a life expectancy of less than one year. The presence of malignant priapism may require a different algorithm for treatment than priapism from other causes. Due to the rare nature of malignant priapism, the literature related to its management consists of mostly case reports and case series rather than clinical trials. As a result, the optimal treatment pathway and relative efficacies of treatments are not clear (3). Treatment may need to be individualized based on the clinical course of the patient.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tau.amegroups.com/article/view/10.21037/tau-23-327/rc

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tau.amegroups.com/article/view/10.21037/tau-23-327/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained

from the patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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