Malignant refractory priapism: An urologist’s nightmare

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Abstract

Malignant priapism is described as persistent, nonsexual erections caused by invasion or metastasis from a primary neoplasm. We present two cases of malignant priapism with different etiologies and the respective management strategies. A 75-year-old patient had undergone radical cystectomy for a high-grade bladder tumor 5 months ago and came with priapism. The patient persisted to have partial penile tumescence with low-grade pain even after intervention. Another 66-year-old patient came to emergency with persistent painful priapism who had been diagnosed to have Multiple Myeloma. He required a corporotomy and open drainage as a last resort which finally relieved him of pain but with loss of erection. The treatment needs to be individualized based on the clinical course of the patient.

Keywords: Cavernotomy, malignancy, priapism, shunt

INTRODUCTION

Priapism is a full or partial erection that continues more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation. Malignant priapism is a term first used by Peacock[1] in 1938 to describe persistent, nonsexual erections caused by invasion or metastasis from a primary neoplasm.

CASE REPORTS

Case 1
A 75 year old patient was referred to us with 15 days’ history of painful priapism. He denied any use of intracavernosal agents or sexual activity. He had undergone radical cystectomy and ileal conduit 6 months back for high-grade transitional cell carcinoma T2N0M0. He is a hypertensive since 25 years. His medications included calcium citrate malate, metoprolol, telmisartan, aspirin, atorvastatin, and levetiracetam. On examination, he had an ileal conduit, priapism with engorged corpora cavernosa but no signs of ischemia. Magnetic resonance imaging of the penis done elsewhere 10 days before did not show any evidence of penile metastasis or arteriolar–sinusoidal fistula.

He had anemia with hemoglobin of 6 g/dl and hypocalcemia with corrected calcium of 11.8 mg/dl during biochemical analysis of his blood sample. Peripheral smear revealed microcytic hypochromic anemia. Penile Doppler showed corporal artery peak systolic velocities of 11.9 cm/s on the right [Figure 1a] and 7.3 cm/s in the left corporal body [Figure 1b], which was suggestive of ischemic priapism.

The patient was immediately wheeled into the operation theater. Cavernosal blood gas analysis showed pH 7.34,
pO₂ 64.3 kPa, and pCO₂ 41.2 kPa. This was neither consistent with classical low-flow nor with high-flow priapism. Percutaneous aspiration of corporal blood and irrigation was done using a pediatric intravenous cannula. 0.9% normal saline solution in combination with intracavernous injection of adrenaline (2 mL of 1/100,000 adrenaline solution) was repeated five to six times over a period of 20 min. Only partial detumescence was achieved, and his pain which was better initially again started increasing. The next day, he was taken up for shunt surgeries. Bilateral proximal cavernospongiosal (Quackles shunt) was performed through a perineal incision in lithotomy position [Figure 2b]. Clutton’s metallic sound was used to evacuate the dark clotted blood from corporal bodies [Figure 2a]. Corpus spongiosum and urethra were indurated, and hence, an incisional biopsy was sent. Detumescence was not complete even after shunt surgery. The patient continued to have recurrent painful erections but no signs of ischemia during subsequent inpatient stay, which was managed with analgesics. The histopathology of the biopsy showed metastatic urothelial cancer. Subsequently, the patient underwent total penectomy.

Case 2
A 66-year-old patient came to emergency with persistent painful priapism [Figure 3] since a day. He was undergoing chemotherapy for multiple myeloma stage-3. The first cycle of chemotherapy had been administered comprising of Bortezomib 2 mg and Thalidomide 100 mg. He was admitted and immediately taken up for intervention. Corporal wash was given using an 18 gauge needle into the base of the penis. A volume of 200 mL of dark blood was drained out. Blood gas analysis showed hypoxia and hypercapnia suggestive of ischemic priapism. Detumescence was noted initially. He developed recurrence of painful erection after around 6 hours. We, therefore decided to do a shunt procedure. In view of the history of hematological malignancy and prolonged duration of priapism, a combination of the distal Ebbehoj shunt and a proximal Quackles caverno-spongiosal shunt were created [Figure 4a]. This again provided temporary relief till next morning only. As a last resort, cavernotomy was planned. The patient and his attendants were explained about the need for radiotherapy to corporal bodies if there is recurrence of priapism and loss of erection in the post operative period. His hemoglobin by then had dropped to 6.1 gm%, and he needed two units of blood.

He underwent bilateral cavernotomy [Figure 4b]. Turgid erection persisted even on opening the corpora bilaterally.
due to fibrosis. The hard spongy tissue finally had to be curetted using metallic sounds to achieve detumescence. Suction drains were placed and removed after 3 days. He completed chemotherapy for multiple myeloma and is asymptomatic but has severe erectile dysfunction. Hewas not willing for prosthesis though.

**DISCUSSION**

Priapism is a full or partial erection that continues more than 4 h beyond sexual stimulation and orgasm or is unrelated to sexual stimulation. It has an incidence of 1.5 per 100 000 and can occur in all age groups from newborn to elderly. There is a bimodal peak of incidence, between 5–10 years in children and 20–50 years in adults. Sickle cell disease is the most common etiology in childhood while pharmacological agents are responsible for most cases of priapism in adults.

Malignancy leading to penile metastases is also one of the rare causes of priapism. Secondary malignancy of the penis from metastatic disease was first described by Eberth in 1870. Of these, 63.9% were metastatic lesions arising from the genitourinary tract (bladder: 28.6%, prostate: 27.9%, kidney: 6.9%, and ureter: 0.5%), with colorectal adenocarcinoma, carcinoma of the respiratory tract, melanoma, and hematologic diseases comprising the rest. Clinical manifestations of penile metastases may include indurated nodules, penile masses, skin lesions, or commonly, malignant priapism which is estimated to occur in 20%–50% of patients. The most frequent sign of penile metastasis is priapism.

Few hypotheses have been proposed to explain the mechanism of malignant priapism, but the actual cause remains unknown. The most accepted current theory is the spread of renal neoplastic cells to the penis by retrograde venous and lymphatic flow and/or antegrade arterial dissemination. The other route of spread may be by direct extension.

Priapism is due to the derangement of penile hemodynamics affecting the arterial component or the veno-occlusive mechanism. Hence, both high and low flow priapism can occur in penile metastases. The most important investigation is an intracavernosal blood gas analysis, as it permits differentiation between low flow and high flow priapism [Table 1]. However, it can occur in combination leading to refractory priapism and permanent loss of erectile function. The stepwise treatment of ischemic priapism is well summarized in EAU guidelines 2018 [Table 2].

There is paucity in the literature to favor one shunt procedure over another. In general, the type of shunt procedure performed is based on the surgeon’s preference and familiarity with the procedure. It is conventional for distal shunt procedures to be tried before considering proximal shunting. Cavernosal smooth muscle biopsy has been used to diagnose smooth muscle necrosis suggesting that shunting is likely to fail which helps decision making and patient counseling, particularly if they are being considered for an acute prosthesis. When the corporal aspiration failed in our case, we went ahead with proximal shunt surgery without attempting distal shunt in the first case as we assumed that erectile function should not be an issue in the patient considering his old age, comorbid condition, and refractory priapism. However, subsequently, we followed the conventional method of distal shunt first in the second case. Nevertheless, any of our intervention or shunt procedures failed to achieve complete detumescence finally ending up in cavernotomy. We did not initiate on anticoagulation therapy (although still controversial) to prevent shunt obstruction. The summary data generated by the AUA 2003 Panel show resolution rates of 74% for Al-Ghorab, 73% for Ebbehøj, 66% for Winter, 77% for Quackels/Sacher, and 76% for Grayhack procedures.

**CONCLUSION**

Metastatic lesions of the penis causing priapism can be managed by local excision, partial/tota1 penectomy, radiotherapy, chemotherapy, or conservative therapy. It indicates advanced disease with a poor prognosis. Life expectancy is <1 year. Malignant priapism acts as a forerunner of poor prognosis and treatment is usually palliative with low success rate. Whatever the cause of this malignant priapism, the routine evaluation, differentiation into high or low flow priapism and the classical textbook pathway of management might not be practical in the presence of malignancy.

Corporal blood gas analysis and penile Doppler may not always give classical findings of low flow/high flow priapism in patients with longstanding history (15 days’ duration). Because priapism is rare and usually unpredictable, the

**Table 1: Cavernosal blood gas analyses in different types of priapism**

| Source                                      | pO\(_2\) (mm Hg) | pCO\(_2\) (mm Hg) | pH    |
|---------------------------------------------|-----------------|------------------|-------|
| Normal arterial blood (room air)            | >90             | <40              | 7.40  |
| Normal mixed venous blood (room air)        | 40              | 50               | 7.35  |
| Ischemic priapism (first corporal aspirate) | <30             | >60              | <7.25 |
literature related to its management is neither voluminous nor rigorous, comprising mostly case reports and small case series rather than controlled trials. As a result, the relative efficacy and safety of different treatments are not clear.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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