A RARE UNDERLYING CAUSE OF PRIAPISM

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ABSTRACT Priapism is an uncommon, potentially devastating urological emergency. Malignant priapism is a rare subtype caused by tumour invasion of the corpora cavernosa. Penile malignancy itself has a low incidence and is most often of metastatic origin, while primary urethral malignancy accounts for a minority of cases described. Management of malignant priapism is difficult, and the prognosis is poor. We present the case of an 80-year-old man with malignant priapism as the presenting sign of primary penile carcinoma.

KEYWORDS malignant priapism, penile carcinoma, priapism, urology

Introduction
Priapism is a urological emergency characterized by an erection lasting greater than 4 hours, unrelated to sexual desire, persisting past ejaculation, and that ultimately culminates in compartment syndrome and fibrosis of the penis if not treated in a timely manner [1,2]. The Ebers Papyrus, an ancient Egyptian document from 1550 BC, contains the first written evidence of priapism and describes a remedy of watermelon, flax, pine, and Hycosamus flowers [3]. In the 3rd century, a Greek physician named Galen of Pergamon described priapism as an “unwanted erection unrelated to sexual desire” and believed that it was caused by dilated arteries in the penile shaft[3]. However, it was not until 1824 that Thomas Callaway became the first surgeon to incise the corpora cavernosum of a patient suffering from priapism[3].

Priapism carries an incidence of 0.5 - 0.9 cases per 100,000 in the general population[2]. It can develop at any age but is associated with a bimodal age distribution, usually occurring between 5 to 10 and 20 to 50 years old[4]. While 50% of cases are idiopathic, the most well-known precipitants are sickle cell disease and drugs such as trazodone and vasoactive erectile agents[5]. There are 3 distinct subtypes of priapism: ischemic, nonischemic, and stuttering or intermittent. Ischemic priapism accounts for most cases (95%) and is characterized by painful erection with rigidity in the corpora cavernosa and a soft glans[2]. Ischemic priapism leads to cavernosal acidosis, hypoxia, necrosis, fibrosis, and erectile dysfunction if detumescence is not achieved within 12 to 24 hours[2]. Stuttering priapism, classically seen in patients with sickle cell disease, is a subtype of ischemic priapism consisting of self-limiting but recurrent episodes of painful erection. Nonischemic, also known as “high-flow” or “arterial” priapism, is typically painless and arises from an arterio-cavernosal fistula. Nonischemic priapism almost universally occurs secondary to traumatic spinal injuries, and management is conservative6.

Case report
We present the case of an 80-year-old man (DS) who presented to our emergency department with a complaint of one week of persistent painful erection. The patient had no known past medical history and took no medications or illicit drugs. On physical exam, the patient’s penis was erect with soft glans, and there were no visible lesions or regional lymphadenopathy. We performed a dorsal penile nerve block with lidocaine 1% without epinephrine, followed by corporal aspiration at the 10 o’clock position with an 18-gauge needle. The blood flow stopped after only 5 mL, and the following removal of the needle from the patient’s penis, we noted purulent drainage from the insertion site. We immediately halted the procedure and initiated intravenous Vancomycin, Zosyn, and Ceftriaxone. A subsequent MRI of the abdomen and pelvis with IV contrast (figures 1-2) revealed a 3.9 x 3.6 x 4.4 cm abnormal enhancing mass involving corpora cavernosum and corpus spongiosum suspicious for tumour “likely urethral in origin,” as well as multiple abscesses in the midshaft penis. We communicated these findings with Urology, who took the patient to the operating room for incision and drainage of the penile abscesses. The patient was taken back to the operating room for pancystourethroscopy, which showed nodular-appearing lesions in the penile urethra from
the 3 o’clock to 9 o’clock positions in the proximal, mid, and distal penile urethra. The prostate and bladder appeared normal. Biopsies were consistent with high-grade urothelial carcinoma. The urologists offered the patient a penectomy, but the patient ultimately was lost to follow-up.

Discussion
Malignancy is an exceedingly rare cause of priapism and has seldom been described as the presenting sign of tumour invasion. For example, Gong et al. found only 4 cases of priapism as the presenting sign of primary penile lymphoma. However, after a thorough review of the literature, we have concluded that this is the first case of priapism as the presenting sign of a primary urethral carcinoma[10]. Malignant priapism arises from primary, metastatic, or hematologic malignancy. Peacock et al. first coined the term in 1938, and as of 2016, only 512 cases have been detailed, not including our case report[8]. A proposed mechanism of malignant priapism is direct neoplastic cell invasion into the penile cavernous sinuses, causing an outflow obstruction[9].

Penile malignancy itself is uncommon, with primary penile malignancies carrying a much lower incidence than those from metastatic disease[2]. Penile carcinoma typically presents as nodular lesions, but other manifestations include ulcerations, hematuria, and obstructive urinary symptoms[9]. Even in metastatic disease, only 24% of cases present as priapism[9]. While a vast majority of malignant priapism is ischemic, a few cases are nonischemic, so clinicians should obtain a penile blood gas prior to intervention. Unfortunately, management of malignant priapism is difficult and mortality is high and challenging mortality, so a multidisciplinary team approach including oncology, urology, and possibly palliative care should be undertaken to determine appropriate care. The prognosis of malignant priapism is poor, with a median survival time of 14.5 months[11].

Conlusion
Priapism has been described since ancient times and is a perplexing disorder even in the present day. Irreversible smooth muscle dysfunction may occur as early as 4 hours after onset of ischemia, and up to 50% of men do not regain erectile function even when properly treated[12]. Malignant priapism is an especially rare and aggressive form of the disease, and only about 500 cases have been documented in the literature. Prognosis is poor, and management usually requires a multidisciplinary team approach. Priapism is seldom the presenting sign of penile malignancy, which often manifests as nodular or ulcerative lesions. We believe that this is the first case of primary urethral carcinoma presenting with malignant priapism.

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Conflict of interest
There are no conflicts of interest to declare by any of the authors of this study.

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