Clitoral Priapism with No Known Risk Factors

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Clitoral priapism is a rare condition that is associated with an extended duration of clitoral erection due to local engorgement of clitoral tissue resulting in pain. Although the pathophysiology is not completely understood, it has been associated with specific classes of medications, diseases that alter clitoral blood flow or others associated with small to large vessel disease. We present a case report of a 26-year-old patient who developed clitoral priapism without a clear medication or disease related etiology. The patient was treated with opiates, imipramine, non-steroidal anti-inflammatory medication, and local ice packs. She recovered uneventfully.


INTRODUCTION

Clitoral priapism is a rare cause of clitoral pain. It is thought to be a result of alterations in local hemodynamics leading to incomplete venous or lymphatic drainage of the clitoral tissue, resulting in edema and tissue swelling with concomitant pain.\(^1\) Due to the increasing number of medications being prescribed that can cause priapism as well as the resultant deficiency in blood flow to the clitoral region, it is paramount for the emergency physician (EP) to diagnose and treat appropriately. The prolonged erection is not associated with sexual stimulation, can last from minutes to days, and is frequently uncomfortable.\(^1,2\) Embarrassment can preclude patients from describing the exact nature of their symptoms, emphasizing the importance of a complete physical exam that includes an evaluation of the clitoris.

Clitoral priapism has been associated with medications producing alpha blockade, inhibition of serotonin reuptake, and non-SSRI antidepressants.\(^1,3\) It has also been associated with transitional cell carcinoma obstructing venous and lymphatic clitoral outflow.\(^4,5\) Since cases are rare, there is inadequate evidence to determine the incidence of this condition or the likelihood of a particular cause. However, it has been shown that discontinuing the offending agent can result in a therapeutic resolution within 24-72 hours.\(^1,6\) Other treatment modalities include imipramine, non-steroidal anti-inflammatory medications (NSAIDs), ice packs, opiates, and, rarely, intracavernous injection of adrenaline.\(^2,6-8\) Presented here is a case of clitoral priapism of unknown etiology and no known risk factors.

CASE REPORT

A 26-year-old gravida 4 para 2 TAB 2 African-American patient presented to the emergency department (ED) with a two-day history of pain following wiping of her urethra. The patient described her pain as constant, aching and sharp, and feared she cut herself near her urethra while wiping. She has not been sexually active for several months and denied insertion of foreign bodies. She described swelling in the region of her clitoris and attempted to alleviate the symptoms with several hot baths with minimal relief. She denied a history of trauma, vaginal pain and discharge, prior similar symptoms, and changes in hair pattern or voice. She also denied fever, dysuria, suprapubic pain, and gastrointestinal symptoms. She denied recent medication use or recreational drug use. The patient’s obstetrical and uro-gynecologic history was significant for two first trimester therapeutic abortions for unintended pregnancies without complication, several urinary tract infections treated effectively, and chlamydia vaginitis treated as an outpatient. She states that all annual pap smears and pelvic examinations prior to her presentation were otherwise normal. The patient also denied a family history of sickle cell disease and cancer.

Her vital signs included a temperature of 37 degrees Celsius, blood pressure of 129/70 mm Hg, pulse of 87 beats
per minute, and respiratory rate of 16 breaths per minute. She was alert, oriented, and in moderate discomfort lying supine on the gurney. She pointed to her clitoral region when asked to point to the area of maximal pain. Her pelvic exam revealed a firm, swollen and acutely tender clitoris and clitoral hood with minimal vulvar tenderness and swelling (Figure 1). Her external genitalia including labia were normal and without lesions, lacerations, or abrasions. She had no cervical motion tenderness, os was closed, and no masses or discharge were present. Uterus was midline and non-tender. Her head and neck, cardiopulmonary, abdominal, lymphatic, and extremity exams were unremarkable.

Laboratory tests included an unremarkable urinalysis and a negative urine pregnancy test. The patient was given vicodin for pain control with minimal relief. A gynecology consultation recommended NSAIDS, imipramine, and ice packs. The patient was referred to her primary gynecologist and instructed to follow up within two days with precautions to return if loss of sensation, increased swelling, or persistent or worsening pain after 48 hours. Her follow-up was uneventful.

**DISCUSSION**

Priapism is defined as a persistent and painful erection of penile or clitoral tissue with a duration of more than six hours and is not associated with sexual arousal. Clitoral priapism is an infrequent cause of clitoral or vulvar pain and can result in unnecessary and extensive laboratory and radiographic testing if not diagnosed appropriately through the history and clinical examination. Although only infrequently associated with etiologies that can cause long-term morbidity, the episodes themselves can be incapacitating and can require multiple hospital or clinic visits for pain control, according to sporadic case reports in the literature. Often a vague history is obtained as a function of embarrassment and frequently the episode has abated by the time the clinician evaluates the patient, making it difficult to diagnose. The details of specific clitoral swelling followed by pain with concomitant erection of the clitoral tissue can be suggestive of the diagnosis. Often, patients will describe an episode of perceived vulvar or labial swelling or pain that leads to clitoral swelling, erection, and extensive pain. Others will describe symptoms of dysuria, anorgasmia, intestinal complaints, and even cramping pain in the pelvic or lower abdominal region within the episode of clitoral priapism. A search for antecedent or proximate causes should ensue, and would likely include a complete recent medication and illicit drug history, a complete physical and gynecological examination, relevant urine and pregnancy studies, and adjunct testing depending upon the physical exam and history findings.

The mechanism of clitoral priapism is theorized to be similar to the pathophysiology of male priapism, which involves altered circulation of the corpora cavernosa and increased clitoral intracavernous pressure resulting in erection of the glans. Decreasing venous outflow results in increased blood volume and pressure in the corpora. Although in men an increase in arterial inflow is an additional mechanism for producing priapism, this does not appear to be a relevant etiology in women. Corporeal venous outflow obstruction is likely a result of the following causes: 1) alpha sympathomimetic blockade resulting in extended relaxation of corporal smooth muscle, 2) physical obstruction or sinus compression, and 3) venous drainage occlusion.

Several groups of medications have been associated with clitoral priapism. Although the exact mechanism for producing clitoral engorgement and pain is unknown. Due to the increasing use of these agents, the condition may become more prevalent, which is why EPs should evaluate for this condition. Alpha adrenergic blocking agents resulting in smooth muscle relaxation and venous stasis in the corpus cavernosa may cause priapism. Priapism is also a known adverse effect of psychotropic medications and is a rare reported adverse effect of antidepressant treatment. Other medications associated with priapism include but are not limited to: Trazodone, bupropion, citalopram, fluoxetine, paroxetine, bromocriptine, olanzapine, and nefazodone have all been implicated as agents contributing to or responsible for episodes of clitoral priapism. Potent 5H2 receptor antagonists have also been reported to cause priapism, although the mechanism is unclear since the majority of 5H2 antagonists have limited alpha blocking effects. Bromocriptine may act through an increase in oxytocin or spinal dopaminergic pathways, although clear evidence is lacking.

Additionally, authors have provided evidence to support the idea that, rarely, clitoral priapism is the result of a physical obstruction of venous or lymphatic drainage and have included transitional cell carcinoma as a cause. Sickle cell disease and spinal cord injury are known causes of priapism, with sickle cell far more common as a cause in males. Although this patient was of African-American decent,
she had no family history of sickle cell disease and it is not thought to be the cause in her situation.

If an anatomic source exists, often excision or other therapies need to be considered with the appropriate specialist. The complete workup must exclude other common causes of clitoral or vulvar pain including urinary tract or pelvic infections, local trauma or abrasions, vulvovaginitis, contact dermatitis, and other causes of pelvic or abdominal pain.

No specific treatment regimen has been proven to be effective in managing clitoral priapism. However, caus-specific treatment frequently reverses the condition and has responded well, in case reports, to the usage of NSAIDS, cooling pads, and opiates, although their effectiveness is inconclusive.\textsuperscript{2,7,15,18} When the etiology in unknown, the course of treatment becomes more confused and uncertain. While several case reports have discussed the use of imipramine as the initial drug of choice, its effect is poorly understood.\textsuperscript{2,6,7} More likely, the patient’s condition resolved because the offending agent was removed. Intracavernous administration of alpha agonists similar to the treatment of penile priapism has also been reported with success.\textsuperscript{8} Therefore, the accepted management involves removal of the offending agent, replacement with an agent from another therapeutic class, and coordinated treatment with imipramine for symptomatic episodes.\textsuperscript{7}

Some authors have advocated for the use of alpha adrenergic agonists in the acute episodes, including phenylpropanolamine or phenylephrine. These medications have similar alpha agonist activity when compared to imipramine but do not have the anti-cholinergic effects. One case highlights scorpion venom as a possible treatment modality.\textsuperscript{19} Urologic and/or gynecologic consultation may be required if symptoms cannot be abated with the above remedies.

CONCLUSION

Clitoral priapism is a rare cause of vulvar pain and can be excruciating to the patient. Diagnosis may be difficult given embarrassment, vague history and inconclusive physical exam. Several causes has been linked to clitoral priapism including, alpha blocking agents, antidepressants, psychotropics, and 5H2 blockers. Other causes, such as cancer, have also been implicated. Treatment involves alpha agonists for acute attacks, imipramine, and symptomatic relief with NSAIDS and ice packs. Gynecology follow up is necessary for further evaluation.

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\textbf{REFERENCES}