HIGH – FLOW PRIAPISM IN A PEDIATRIC PATIENT. A CASE REPORT

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Abstract. Priapism is a rare entity in the pediatric population and represents a persistent, usually painful erection that lasts for more than 4 hours and occurs without sexual stimulation in all age groups including newborns. However, it usually affects men between the ages of 5 to 10 years and 20 to 50 years. Priapism is caused by an imbalance between penile blood inflow and outflow. There are two types of priapism, low-flow due to venous occlusion and high-flow priapism due to uncontrolled arterial flow to the veins. High-flow priapism most frequently occurs as a result of penile or perineal trauma in which the intercavernosal artery disruption causes an arteriocavernosal fistula. Treatment ranges from expectant management to open surgical exploration with vessel ligation. The major chronic morbidity associated with all types of priapism is persistent erectile dysfunction, impotence and psychogenic sexual aversion. We report a successful treatment of high-flow priapism in a 5-year-old boy with superselective transcatheter embolization of the fistula with platinum microcoil using a coaxial microcatheter.

Keywords: priapism, high-flow, embolization, microcoil

CASE REPORT

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Introduction

Priapism is a pathological persistent erection that lasts more than 4 hours in which time the erect penis does not return to its flaccid state, despite the absence of sexual stimulation.[1] Priapism is caused by an imbalance between penile blood inflow and outflow. [2] It normally involves the corpus cavernosa while the ventral corpora spongiosum and glans remain flaccid. This illness was first reported in sickle-cell disease patients in 1934. [3]

The overall incidence of priapism is 1.5 cases per 100,000 person-years which increases to 2.9 cases per 100,000 person-years for males older than 40 years.[4] Although priapism may occur at any age from infancy through old age, a bimodal distribution, with a peak between 5-10 years in children and another between 20-50 years in adults, is noted.[2,5] Priapism in younger groups is more often associated with medical conditions while, in older groups, it tends to be secondary to pharmacologic agents. No racial predilection exists.[6] Priapism is primarily a disease of the male species, priapism of the clitoris has been reported, but is extremely rare.[7]

The classification of priapism as high-flow or low-flow appeared in the 1980s when it became essential to suggest causality and to determine prognosis and treatment.[8]

Low-flow priapism is usually due to full and unremitting corporeal veno-occlusion where venous stasis and deoxygenated blood pools within the cavernous tissue. The situation is an emergency, patients present with a painful erection and should be resolved within 6 hours from the onset of the episode in order to minimize the sequelae. Prolonged veno-occlusive priapism results in fibrosis of the penis and a loss of the ability to achieve an erection. Significant changes at the cellular level are noted within 24 hours in veno-occlusive priapism. It is a true urologic emergency that may lead to permanent erectile dysfunction and penile necrosis if left untreated. Corporal fibrosis due to persistent priapism can result in deep tissue infections of the penis.[9] The major chronic morbidity associated with all types of priapism is persistent erectile dysfunction and impotence. The duration of symptoms is the most important factor affecting the outcome. 92% of patients with priapism for less than 24 hours remained potent, while only 22% of patients with priapism that lasted longer than 7 days remained potent.[10]
High-flow priapism is characterized by an increase of arterial supply to the corpus cavernosa, usually secondary to a rupture of a cavernous artery with unregulated flow into the lacunar spaces and normal venous drainage. [11,12] This rare type of priapism is usually less painful or even painless and is not considered an emergency, the onset may be delayed after the acute injury; the delay may be due to an initial vessel spasm or to the formation of a clot that is generally reabsorbed over a period of days.[6] Arterial high-flow priapism occurs with less tumsence of the erect penis compared with venous priapism, has better prognosis and complication as impotence is rare (less than 20%).[8] In children, high-flow priapism is typically caused by post traumatic arteriocavernosal fistula (after penile, blunt perineal injury or pelvic trauma), and is generally manifested several days after the trauma.[13]

Priapism is associated with a number of important medical conditions and pharmacologic agents, trauma or activities that may result in the formation of an arterial-venous fistula, especially in children.

Physical examination finds an erect or semierect penis with glans and corpus spongiosum rarely rigid. [14] It is a must to examine the patient carefully for any evidence of trauma or unreported injections sites to the genital region and for an underlying condition that may predispose to priapism.[6]

Laboratory studies – in patients with no known predisposing factors, a complete blood count is appropriate in order to identify leukemia; patients with sickle cell disease have a complete blood count and a reticulocyte count. If sickle cell status is unknown, a hemoglobin S determination may be useful. An arterial blood gas examination of the cavernous blood aspirate is useful in differentiating between high and low flow disease. [15] Values similar to venous blood suggest a low flow etiology, while values similar to arterial blood suggest high flow priapism. Other useful analyses are: a coagulation profile, a platelet count and urinalysis.

Imaging studies – colour flow penile Doppler is currently the study of choice to identify both high and low flow priapism. In patients with high flow priapism, selective penile angiography may be required in order to identify the site of the fistula.[15,16]

With respect to the treatment, any patient who has an erection for longer than 4 hours, especially if he has a predisposing illness, should probably receive therapy for priapism. Most cases seen early enough in their course respond to conservative measures, meaning use of ice packs to the perineum and penis or asking patient to walk up stairs.[17] Treatment for priapism secondary to sickle cell disease includes hydration, alkalinization, analgesia and oxygenation. Hypertransfusion and/or exchange transfusions may be required to decrease hemoglobin S to less than 30% (this method has a high rate of success, but may produce serious neurological side effects).[6]

Treatment for low-flow (vaso-occlusive) priapism include the use of terbutaline orally, at a dose of 5-10 mg, followed by another 5-10 mg, 15 minutes later, if required (produces resolution in about 1/3 of patients). Oral pseudoephedrine 60-120 mg is a potential therapy, due to its alpha agonist effect. If oral therapy fails, aspiration of the corpus cavernosum and intracavernous injection of alpha adrenergic agents or methylene blue instilled into the corpus cavernosa is the next line of therapy. If initial aspiration of the corpus cavernosum reveals bright red blood rather than dark venous blood, we could consider an arterial cause for priapism. Compression therapy may be successful in certain cases, especially in children. If the procedure fails, selective angiography with subsequent embolization of the offending vessel has been shown to be effective.[5] Patients who are not responding may benefit from surgical ligation of the fistula.[18]

Regarding the prognosis, most patients respond to therapeutic measures. In high flow priapism patients may require surgical intervention to correct the problem. Deaths in these patients are usually related to complications from the underlying condition (leukemia, sickle cell disease, malignancy).[6]

Case report

A 5-year-old boy was referred to the surgical department in June 2012 with a 10 days history of painless priapism. He sustained a straddle injury to his genital region while attempting to descend from an engine toy, at home. Priapism developed the following day. He was referred to another surgical department and started treatment with ice pads. After 10 days of painless priapism, we examined the patient and we discovered a non-tender erect penis without scrotal or genital bruising and no other complains.

Hematological investigations included a normal full blood count and negative sickle-cell screen, coagulation status, inflammatory and metabolic tests. Despite the history of trauma, it was important for us to rule out other contributory diseases such as sickle-cell disease and leukaemia. Doppler ultrasound was performed, which demonstrated a different flow in the two corpora cavernosa. The left side appeared to show an arterio-venous communication at the base.

We performed blood aspiration from the cavernous bodies. First we performed a penile nerve block with 1% lidocaine without epinephrine; after anaesthesia was ensured, we used a 19-gauge needle attached to a large syringe and punctured the corpus cavernosum; we used a lateral point of
insertion, through the shaft of the penis, to avoid the corpus spongiosum and urethra ventrally and the neurovascular bundle dorsally. We then aspirated 20-30 ml of blood from the 2-o’clock position while milking the shaft; because of multiple communications that exist from one corpus to the other, we did the aspiration only on one side. We aspirated bright red blood, demonstrating an increased concentration of arterial blood.

Angiography – right femoral artery approach; 4F artery sheath; Cobra 2 catheter – 4F toggled in left penile artery; we discovered a major high-flow arterio-venous fistula; contrast filled the penile tissue fast; we used a 3X80 mm platinum microcoil and obstructed the fistula; angiographic control showed no evidence of a remaining fistula; no complications noted.

Our patient was reviewed on the 9th of June 2012. He was clinically well with no priapism and normal nocturnal erections reported by his mother.

Discussion

The arterial blood supply to the penis is derived from the internal pudendal artery, a branch from the anterior division of the internal iliac artery. The corpora cavernosa are supplied by the deep arteries of the penis while the corpus spongiosum and glans are supplied by the artery of the bulb. There is an additional dorsal artery of the penis.[19]

The patho-physiological process of high-flow priapism is somewhat more complex than simply an increased blood flow. It is thought that nitric oxide release from endothelial cells and activation of the guanylate cyclase enzyme causes relaxation of the smooth muscle tissue of the cavernous bodies and subsequent persistent erection. Nitric oxide release is stimulated by shear stress on endothelial surfaces, such as that produced by high turbulent flow adjacent to an arteriocavernous fistula and by high oxygen pressures.[6]

Priapism is frequently idiopathic in etiology, but is associated with a number of important medical conditions and pharmacologic agents: hematological disorders (sickle-cell disease – 38-42% of adult patients reported at least one episode, leukemia, thalassemia, multiple myeloma, Henoch-Schönlein purpura, thrombocytopenia, paroxysmal nocturnal
haemoglobinuria), neurological disorders (head or spinal cord lesions or trauma), metabolic disorders (Fabry’s syndrome, deficiency of glucose-6-phosphate dehydrogenase, diabetes, gout, amyloidosis, haemodialysis, hyperlipidemic parenetal nutrition), hyperlipidaemia, arterial blood hypertension, mental illness (bipolar, schizophrenia, depression), malignancy (primary or metastatic), achalasia, infections diseases (malaria, Mycoplasma Pneumoniae, HIV), trauma or activities that may result in the formation of an arterial-venous fistula or shunt such as bicycle riding or motorcycles and medications (for erectile dysfunction – papaverine, phentolamine, prostaglandine E1, selective cyclic guanosine monophosphate inhibitors such as sildenafil, testosterone, antihypertensives - hydralazine, antidepressants and antipsychotics – chlorpromazine, thioridazine, trazodone, quetiapine, and newer agents such as citalopram, anticoagulants – heparin and warfarin), other drugs such as omeprazole, metoclopramide, tamoxifen, androstendione and relaxing or illicit drugs (ethanol, ecstasy, marijuana, cocaine, heroin).

A diagnosis of high-flow priapism can be made from a typical history of trauma and painless erection, supported by clinical findings and investigations. As in our case, the most frequent history described in the literature is the blunt penile or perineal trauma of a straddle injury. The most commonly described example of trauma was falling onto bicycle handlebars. Time between injury and the onset of priapism is variable but is usually within 7 days. Delayed onset occurs because nocturnal erections cause vasodilatation rupturing the damaged artery resulting in unregulated high flow into the corporeal sinusoids. The resulting fistula bypasses normal regulation by the helicine arteries of the penile erectile tissue. In addition, after the injury, a clot may form occluding the defect which then undergoes lysis or is dislodged in response to nocturnal erection.

Case reports in the literature describe similar examination findings usually with a partially erect penis. The turgidity is confined to the corpora cavernosa sparing the corpus spongiosum and the glans, as their arterial supply is different. Also, tension of the corpora cavernosa is often less in the anterior third of the penis. Perineal compression with the thumb resulting in immediate detumescence with relapse after withdrawal of the thumb is known as Piesis clinical sign and indicates high-flow priapism. In high flow priapism, the diagnosis proposed the oxygen pressure exceed 30 mm Hg, carbon dioxide pressure less than 60 mm Hg and pH greater than 7.25. A diagnosis of low-flow priapism can be made with converse blood gas analysis.

Whilst aspiration and blood gas analysis is an excellent method of confirming a diagnosis of high-flow priapism if uncertainty exists, aspiration is not universally recommended and is thought to be unnecessary if the diagnosis is clear.

The most commonly performed imaging studies is colour flow Doppler ultrasound (Duplex) and angiography, which can both demonstrate a pathological increased arterial flow. Doppler ultrasonography must be performed with the patient supine with his legs bent allowing access to the perineum. The transducer is placed in the perineal region to locate the cavernous artery laceration. Ultrasound scanning of the cavernous bodies shows high, turbulent flow and peak systolic velocities that can be measured and compared on each side. The advantages are numerous. It is a non-invasive technique with no radiation exposure, no contrast administration and thus no allergic response and is widely available. Angiography, whilst invasive, has a much higher rate of fistula detection and allows embolization to be performed.

There is no clear consensus as present as to the gold standard of management in the paediatric population. Expectant management or “watching and waiting” is an acceptable and effective management. It is safe as, despite the increased influx of arterial blood into the corpus cavernosum, venous outflow is not compromised; therefore there should be no damage to the corporal bodies and no development of compartment syndrome. There is no clear indication of the acceptable safe duration of “watching and waiting”. It is probable that older children and adults are less likely to tolerate the embarassment of priapism and may seek early definitive management. We propose that the conservative management is highly appropriate in children and that for non-resolving cases, angiography is the gold standard investigation that allows embolization treatment, if necessary.

Aspiration/injection of the corpus cavernosum – by using a needle attached to a large syringe, puncture the corpus cavernosum; then aspirate 20-30 ml of blood from either the 2-o’clock or 10-o’clock position while milking the shaft; if aspiration or injection is successful in producing detumescence, an elastic bandage can be placed around the shaft of the penis to ensure continued emptying of the corpora and to compress the puncture site. Aspiration alone has a success rate around 30%. If this procedure is not successful, phenylephrine, epinephrine or methylene blue may be instilled into the corpus cavernosa under constant hemodynamic monitoring concerning about severe hypertension, bradycardia, tachycardia and arrhythmia. We may use alpha adrenergic agonists such as phenylephrine or pseudoephedrine, beta adrenergic agonists (selective beta-2-adrenergic...
agonists such as terbutaline) and guanylate cyclase inhibitors (inhibitory effect affecting smooth muscle relaxation such as methylene blue). Phenylephrine is an alpha agonist very effective in some cases. The exact mechanism is not clear. It is a strong postsynaptic alpha receptor stimulant with little beta-adrenergic activity that produces vasoconstriction of arterioles and increases peripheral venous return. The drug is best administered in a diluted solution, 10 mg (1 ml) of phenylephrine added to 499 ml of saline 0.9%. Pseudoephedrine (sudafed) stimulates vasoconstriction by directly stimulating alpha adrenergic receptors. Consider to use of oral alpha adrenergic agonists for 3-5 days to help prevent recurrent episodes. It is useful to note the successful use of ketoconazole and prednisone for treatment of recurrent priapism.[21]

If expectant management and the procedure of aspiration/injection fails, selective angiography with subsequent embolization has been shown to be effective. Patients who are not responding may benefit from surgical ligation of the fistula, despite the potential complication of this procedure. [18]

Conclusion

Priapism caused by trauma to the perineum is rare in young subjects. High-flow priapism is a rare occurrence and thus the majority of clinicians have limited experience in such cases. Color Doppler sonography is a useful pre-angiography study for localization of the causative lesion and it is currently considered the imaging modality of choice for the diagnosis of high-flow priapism because it is sensitive, noninvasive and widely available. In patients who have failed with conservative treatment, superselective transcatheter embolization of the fistula with microcoil using a coaxial microcatheter must be attempted.

We report the successful treatment of high-flow priapism in a 5-year-old boy. Conservative management, highly appropriate in children, has failed in our case; we did superselective transcatheter embolization of the fistula with platinum microcoil using a coaxial microcatheter. A rare entity was successfully managed by a mixed team including paediatric surgeon and radiologist. The case also highlights the importance of warning both parents and surgeons about children who have had even minor perineal trauma about the possibility of developing delayed high-flow priapism.

References

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