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Prolonged ischemic priapism in an adolescent with sickle cell anemia: Challenges of management

Akputa Aja Obasi, Wilson Egwu Sunday Omebe

ABSTRACT

Introduction: Persistent penile erection lasting more than 4 hours and unrelated to sexual interest or stimulation is called priapism. Priapism is uncommon in children. Of the three widely accepted types: ischemic, non-ischemic, and stuttering priapism; ischemic priapism is the predominant type seen in children. A common cause of ischemic priapism in children is sickle cell disease. Ischemic priapism of prolonged duration is associated with a higher risk of complications especially irreversible loss of erectile function. It is a difficult clinical condition to manage and there are no established guidelines for its management in children. Adequate surgical treatment does not guarantee against the risk of devastating long-term sequelae.

Case Report: We report a case of a 12-year-old boy with sickle cell anemia who presented after 48 hours of sustained painful penile erection. He required multiple surgical shunting procedures including distal corporoglanular shunt with tunneling of both corpora cavernosa with Hegar's dilator to achieve detumescence. The management challenges encountered are highlighted.

Conclusion: A potential role for anticoagulation and the use of low-dose hydroxyurea in the prevention of recurrent ischemic priapism in children with sickle cell anemia are advocated.

Keywords: Children, Ischemic, Priapism, Prolonged, Shunt, Sickle cell anemia

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INTRODUCTION

Persistent penile erection lasting more than 4 hours and unrelated to sexual interest or stimulation is called priapism [1]. Priapism is uncommon in children [2]. Of the three widely accepted types: ischemic (low flow), non-ischemic (high flow), and stuttering (recurrent) priapism; low flow (ischemic) priapism is the commonest type seen in most male subjects and predominantly in children [2]. Ischemic priapism is usually painful [2]. A common cause of ischemic priapism in children is sickle cell disease (SCD) [1–3]. Treatment within 6 hours of onset will result in the preservation of erectile function [3]. Ischemic priapism of prolonged duration (usually longer than 48 hours) is associated with a higher risk of erectile dysfunction (ED), corporal fibrosis with penile deformity, penile gangrene, and psychological sequelae regardless of adequate treatment [2]. Whereas the management of prolonged ischemic priapism in adults follows well-established guidelines, consensus on such guidelines is yet to be established for priapism in children [2, 4].

Prolonged ischemic priapism is a difficult clinical condition to manage [5, 6]. Thus, we report our experience with the management of prolonged ischemic priapism in an adolescent with sickle cell anemia (SCA) and discuss the management challenges encountered.

CASE REPORT

We present a 12-year-old male with SCA (HbSS genotype) who presented to the sickle cell clinic of our institution with a 48-hour history of sustained painful penile erection. This had not been provoked by sexual stimulation, drug ingestion, or the use of illicit substances. There was no prior perineal or penile trauma. For about a month before the current episode, he had been having painful nocturnal erections lasting four (4) or more hours. Cold compress and over-the-counter analgesics had offered relief until the current event. He had not been regular at the sickle cell clinic and had not been on hydroxyurea. He had an episode of ischemic priapism about a year previously which resolved with aspiration and saline irrigation.

He was promptly admitted to the children emergency room (CHER). Blood samples were taken and hydration with intravenous fluid was commenced. Parenteral opioid analgesics, intravenous antibiotics, and oxygen were given. In addition, an exchange blood transfusion was done but the erection persisted. This necessitated a consult to the pediatric surgeons.

The pediatric surgeon's review revealed a pale boy with a temperature of 38°C. The external genitalia showed a fully erect tender penis with rigid corpora cavernosa (CC) and flaccid glans penis. The skin of the phallus looked normal (Figure 1). There was no evidence of trauma noted around the penis and perineal area.

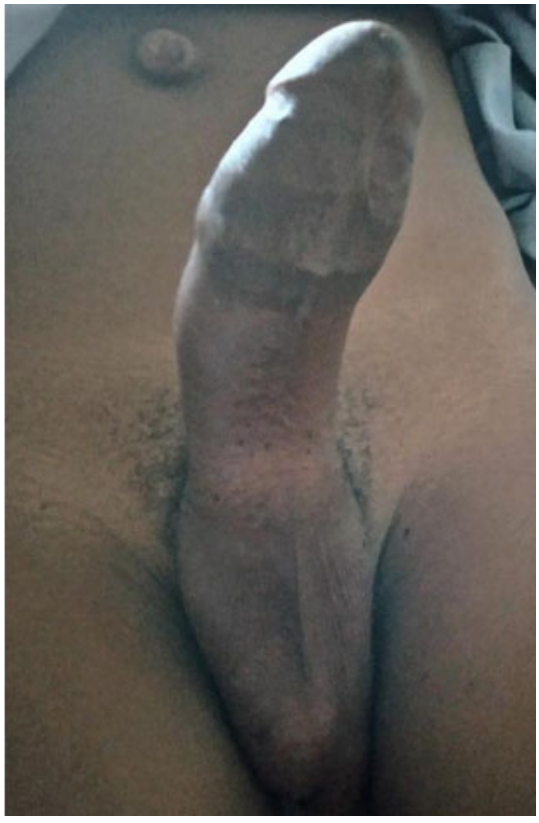

Figure 1: The fully erect phallus with normal penile skin.

Complete blood count showed anemia, leukocytosis with absolute neutrophilia, and thrombocytosis. Blood film was positive for *Plasmodium falciparum*. With a diagnosis of prolonged ischemic priapism, he was worked up for percutaneous distal corporoglanular shunt. Bilateral T-shunts using a size 10 blade scalpel were done 12 hours after admission and dark ischemic blood and clots were evacuated. Rapid complete detumescence was achieved for about one hour. However, before the patient left the operating room (OR), the penis had become semi-flaccid. By hospital day 2, the penis had almost assumed full erection status. Under regional anesthesia, a repeat T-shunt of both CC with tunneling using size 4 Hegar's dilator was done 24 hours after admission into the hospital.

This resulted in complete detumescence (Figure 2). He reported complete pain relief after the procedure. Postoperatively, he was placed on an anticoagulant (clopidogrel) for 7 days. He was discharged home on postoperative day 5.

Follow-up six weeks post-shunt revealed that the penis had remained flaccid but he was yet to have nocturnal erections. Color Doppler ultrasonography of the penis showed excellent blood flow in the corpora cavernosa without evidence of fibrosis. He was worked up for the commencement of hydroxyurea.

Figure 2: The phallus in the flaccid state following detumescence.

DISCUSSION

Priapism as a urological complication of SCD is often cited as a difficult clinical condition encountered by urologists [3]. Up to 65% of cases of priapism in children occur in boys with SCD [3, 6]. The mean age of occurrence

is 12 years of age [3]. Ischemic priapism presenting more than 24 hours after onset is a challenging emergency to manage [7].

The index patient presented very late with a 48-hour history of persistent priapism. He was 12 years old and was presenting with a second episode of prolonged ischemic priapism in as many years. His parents were not unaware of this complication of SCD. We believe the reason for the late presentation was embarrassment. Embarrassment has been reported as a causative factor for failure to seek medical attention and under-reporting of pediatric priapism [2].

Prolonged ischemic priapism is known to cause cavernosal smooth muscle necrosis, fibrosis, and subsequently erectile dysfunction (ED). Studies in adults report that 90% of cases presenting more than 24 hours after the onset of ischemic priapism develop ED [7]. Detumescence was eventually achieved 72 hours after the onset of priapism in our patient. Though there is a very high risk of ED in our patient, our optimism is guarded since some reports in adults have reported preservation of erectile function in adults presenting with prolonged ischemic priapism following treatment with T-shunt and intracavernous tunneling [7, 8]. In addition, some studies report good prognosis in prepubertal boys with priapism [3].

Another challenge of management we experienced was a paucity of equipment for diagnosis and prognostication. Analysis of blood gases (ABG) is not routinely done at our center. Insisting on such investigation would lead to further loss of time before surgical intervention. In addition, magnetic resonance imaging (MRI) with the ability to reliably provide information on the viability of the penile smooth muscles and therefore the potential for ED in future is not readily available in our setting. Moreover, the cost and the time it would take to undertake an MRI are further limitations in a patient presenting with prolonged ischemic priapism [7], who requires prompt diagnosis and expeditious treatment.

Timely treatment using an effective modality is crucial in priapism. Non-surgical treatment is unlikely to result in the resolution of prolonged ischemic priapism [7, 9]. Our patient received conventional medical treatment including exchange blood transfusion to no avail. This tends to suggest the lack of awareness that ischemic priapism of extended duration is refractory to non-surgical management. The knowledge that prolonged ischemic priapism is amenable to distal corporoglanular shunts and modifications thereof [3, 7, 9], needs to be disseminated to physicians who treat children with SCD.

A surprising challenge in managing this child was the failure of the percutaneous distal corporoglanular shunt. After the initial T-shunt, detumescence was transient with the patient almost regaining full erection status a few hours later. Plausible explanations could be that percutaneous shunts don't provide adequate drainage of the old, stagnated deoxygenated blood within the proximal corpora in prolonged ischemic priapism [7]. Alternatively, exposed collagen being a potent initiator of blood coagulation led to the formation of a clot leading to blockage of the distal corporoglanular shunt [10]. For this reason, we subsequently gave the patient clopidogrel to prevent shunt blockage by a clot.

Prevention of recurrence is an ever-present challenge in this child. He had unrecognized stuttering priapism. Stuttering priapism is the precursor of acute ischemic priapism [3, 7]. Current measures used to prevent stuttering priapism in SCA like androgen ablation therapy or intracavernous injection of type 5 phosphodiesterase (PDE5) inhibitors are neither suitable nor recommended in children [3, 10]. Though the use of hydroxyurea is controversial [3, 10], it has shown proven benefits in the prevention of stuttering priapism and reversal of chronic organ damage including restoration of erectile function lost as a result of prolonged priapism [3, 11–13]. Low-dose hydroxyurea may be beneficial in adolescent patients like ours.

CONCLUSION

Management of prolonged ischemic priapism is a real challenge in children with sickle cell anemia. To minimize the risk of long-term sequelae, the child with prolonged ischemic priapism should proceed to percutaneous distal shunt with tunneling or open distal shunt with or without tunneling. The role of anticoagulation needs to be evaluated with prospective studies involving children. Prevention of recurrence of ischemic priapism in children with SCD is still problematic and we recommend the use of hydroxyurea.

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Author Contributions

Akputa Aja Obasi – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be 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

Wilson Egwu Sunday Omebe – Conception of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be 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

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.