Priapism in paediatric patients with sickle cell disease - a report of three cases at the University of Port Harcourt Teaching Hospital

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Abstract

Background: Priapism is an abnormal persistent erection of the penis which may be painful or not painful. Approximately two-thirds of all paediatric patients who have priapism also have sickle cell disease (SCD). The aim of this study is to review all the cases of priapism admitted into the paediatric emergency room of the University of Port Harcourt Teaching Hospital (UPTH) over a 5-year period and to determine the prevalence, presentation, precipitating factors and management of cases seen.

Method: The admission register in the Children's emergency ward was reviewed from January 2001 to December 2005. All the cases of sickle cell disease were extracted. The folders of the patients with priapism were studied and data analysed.

Results: Of the 185 SCD cases, three (1.6%) had priapism. They were adolescents aged 17 years, 11 years and 10 years 9 months respectively. Two patients had never attended a sickle cell clinic, never been on routine drugs nor received advice on oral fluids intake. One patient had stuttering priapism, 24 hours before the event. Two of them had glanulo-cavernous shunts, with early detumescence on Day 3 and 4 respectively, while the patient who was managed conservatively had detumescence after 12 days.

Conclusion: Priapism is an uncommon presentation of SCD in our environment. With prolonged duration, priapism has severe sequelae of impotence and erectile dysfunction, thus, its presentation and management should be known by patients and parents or care givers. Surgical intervention is recommended as it is more effective and results in shorter hospital stay.

Key words: Priapism, Sickle cell disease, Glanulo-cavernous shunt, Conservative management

Introduction

Priapism is a persistent and abnormal erection of the penis which is involuntary and prolonged a result from pooling of blood in the corpora cavernosa leading to obstruction of venous or arterial outflow. There are three basic patterns seen with priapism-stuttering priapism seen especially in children with multiple short episodes over a period of hours, days or weeks, acute severe priapism characterised by erection for hours and days and chronic relapsing priapism.

Two types of priapism are described. Arterial high-flow priapism, which is non painful, usually secondary to acute penetrating penile injury and the veno-occlusive painful priapism which is due to full and unremitting corporeal veno-occlusion.

Patients with sickle cell disease are more prone to veno-occlusive priapism. Approximately two-thirds of paediatric patients presenting with priapism have sickle cell disease.

Priapism may be unrelated to sexual stimulation and unrelieved by ejaculation. It is a urologic emergency and early intervention allows for functional recovery. Priapism has been described in all age groups from infancy through old age. In sickle cell disease, the peak incidence occurs between 19-21 years. The major morbidity associated with priapism is persistent erectile dysfunction and impotence. The duration of symptoms is the most important factor affecting outcome. A study reported that 92% of patients with priapism lasting less than 24 hours remained potent while only 22% of those having symptoms lasting more than 24 hours remained potent.
lasting longer than 7 days remained potent.

While some report priapism as rare in sickle cell disease, others report a high frequency. This contrasting picture and the long term sequelae of impotence, informed a review of all the cases of admission into the paediatric emergency room over a five-year period with a view to determining the prevalence, presentation, precipitating factors, clinical course and management of cases seen.

Methodology

The admission register in the Children's emergency ward was reviewed over a 5-year period from January 2001-December 2005. All the cases of sickle cell disease and their reasons for admission were extracted. The folders of the patients with priapism were studied and data such as age of patient, age at diagnosis of sickle cell status, history of stuttering, previous blood transfusion, regular fluid intake, routine drugs intake, duration of erection, home and hospital management including medical and surgical treatment were extracted. The educational background of parents was also recorded.

Results

There were 185 cases of sickle cell disease with complications admitted into the Children's emergency ward over the five-year period. Of these, only 1.6% presented with priapism.

Case 1

A 17-year-old, diagnosed with SCD at 2 years of age following 'hand and foot syndrome'. He had 5 episodes of vaso-occlusive crisis in the past year, one episode of acute chest syndrome three years earlier and had been transfused 4 times. He was regular at clinic follow up and received routine drugs with liberal oral fluids intake. He presented with persistent penile erection and pain of one day duration. This was the first episode with no previous history of stuttering. There was no precipitating factor such as playing football, sexual activity or masturbation, urethral discharge or urinary symptoms. His father was informed when he came back from work, though mother was at home with patient. He presented to hospital less than 18 hours after onset of symptoms. He had received paracetamol at home. He was the 5th of six children and in Class SS3. Father is an Accountant while mother is a nurse. Both parents have Genotype AS.

At the UPTH, he received intravenous fluids, oral pentazocine, ibuprofen and diazepam and a consult was promptly sent to the urologist on call. First review by the Urologist was 10 hours after admission and patient was treated conservatively with stilbesterol, aspirin, and ice pack to the phallus. Following no improvement, the patient had a glanulo-cavernous shunt operation on the 5th day of admission with detumescence 4 days later. He stayed in hospital for 16 days and has been followed up for one year with no repeat episodes.

Case 2

An 11-year-old, diagnosed at 1 year of age following haemolytic crisis at six months of age for which he received blood transfusion. Subsequently he was declared 'healed' by a pastor. He neither came for follow-up nor received routine drugs and liberal oral fluids. He presented with a first episode of penile erection and pain of 2 days with no previous stuttering. No precipitating factor was noted. He had received treatment in a chemist with ampiclox and novalgin. Father was the first to be informed. Father is an electrician and mother a trader. He was a primary 5 pupil and the second of 4 children. Genotype of parents was unknown. At the UPTH, he was treated with intravenous fluids and paracetamol and seen by the urologist unknown. 24 hours after admission, who managed him conservatively with sodium bicarbonate, stilbesterol, mist potassium citrate. He developed complications of sequestration crisis, heart failure and acute renal failure and received blood transfusion. He achieved detumescence after 12 days and was discharged on the 13th day of admission. Followup after one year showed no recurrence.
Case 3

A 10-year and 9 month old male was diagnosed on presentation with the first episode of penile erection and pain of one day duration. He had been transfused at 6 months of age for 'malaria'. He had played football three hours earlier. Father was informed. He was brought to UPTH within 12 hours after treatment in a private clinic with intramuscular diazepam, cold water enema and novalgin. He was seen by the urologist after 7 hours. He had glanulo-cavernous surgery within 3 hours of review by the urologists, achieved detumescence after 3 days and was discharged on the 6th day of admission. No repeat episode was noticed after one year of follow-up.

Discussion

Only three patients with SCD presented with acute priapism and one of them with a history of stuttering, in our five-year study. This is one-third of the number seen in a Togolese study\(^1\) in which over a three-year period, eleven patients were seen, six of whom presented with acute priapism, four with stuttering priapism and one with acute priapism complicating stuttering priapism. Our low prevalence may be explained by patients not reporting in hospital because they are not previously counselled and may be embarrassed by the presentation of erect phallus and so, reluctant to inform parents. This also explains why only the fathers were informed. Priapism can be frightening, embarrassing and often associated with guilt\(^2\).

The onset of priapism may be in early childhood or adolescence\(^3\). The actuarial probability of young males with SCD experiencing priapism by 20 years of age was estimated at 89\(^6\). In our study, the age range of onset of priapism is 10-17 years which is comparable to another study with a range of 2.5-15 years\(^4\). All our patients were adolescents with only one probably in the sexually active age group. They all denied any sexual activity prior to symptoms. Priapism can be precipitated by prolonged intercourse or masturbation\(^2\).

Apart from the 17-year-old who was attending regular follow-up clinic at the sickle cell clinic, the two others had never been to the sickle cell clinic so were both not counselled on the preventive values of liberal oral fluids and were not on routine drugs. Priapism can result from idiopathic or secondary causes. The commonest cause in sickle cell disease is localized sickling and obstruction of venous drainage from the corpora cavernosa\(^4\). As with other vaso-occlusive crisis in sicklers, the precipitating factors include physical exertion, exposures to extreme weathers, fever, dehydration and emotional disturbance\(^2\). Two of our patients had symptoms few hours after playing football, so exercise and dehydration may have been predisposing factors.

Priapism is a urologic emergency and early intervention is the best chance for functional recovery. The duration of symptoms before treatment is the most important factor affecting outcome\(^1\). The patient who presented on the same day within 12 hours of onset of symptom, had surgery same day and achieved detumescence within 3 days, whereas the patient who was managed conservatively had sustained erection for 2 days at home and for another 10 days in hospital. This delay is being highlighted because it is reported that 92% of patients with priapism lasting less than 24 hours remained potent while only 22% of those with priapism lasting more than 7 days remained potent\(^1\).

Prehospital care of patients with priapism, especially in those with sickle cell disease include ice pack to the perineum and penis and asking the patient to walk up the staircase. The mechanism for the latter is an arterial steal phenomenon\(^2\). If these measures fail to achieve rapid detumescence, patients should be transferred to hospital immediately. These management techniques were unknown to our patients, including the patient on regular follow-up probably because priapism is not one of the symptoms discussed during counselling sessions in the clinic. Key steps in management of priapism in SCD include oxygenation, hydration,
alkanisation, analgesia, hyper transfusion and/or exchange blood transfusion to increase the haematocrit concentration to greater than 30% and decrease the haemoglobin-S value to less than 30%\textsuperscript{3}. Red cell transfusions may be required. All our patients received intravenous fluid, one had sodium bicarbonate and the third was transfused. Immediate emptying of bladder, warm sitz baths, oral rehydration and analgesics have been used successfully at the first onset of symptoms\textsuperscript{2}.

Pharmacological interventions include the use of alpha-agonists such as metaraminol bitartrate or methylene blue\textsuperscript{3}, which counteracts smooth muscle relaxation. Oral terbutaline is also used\textsuperscript{1}. Etilefrine has also been used successfully in children. Stilbesterol was used in two of our patients, one of whom had surgery after failed medical management and the other was managed conservatively. For the patient who did not have surgery, stilbesterol was started within 36 hours yet intumescence lasted 12 days, so the effectiveness of stilbesterol was doubtful.

The failure of conservative measures to cause rapid alteration of priapism encourages the use of surgical procedure. Intracavernous injection of an alpha-adrenergic agonist is said to be effective in childhood\textsuperscript{1}. The glanulo-cavernous shunt (Winter technique) is the first reasonable approach for refractory cases\textsuperscript{1}. This was done for the two patients who had surgery with good response. Other procedures include cavernous shunt (El-Ghorab procedure)\textsuperscript{1}, cavernosal-spongiosum shunt (Quackle)\textsuperscript{1} and cavernosal-saphenous vein shunt (Grayhack)\textsuperscript{1}.

Though surgical procedures are recommended if conservative measures fail to produce detumescence\textsuperscript{2}, the duration of detumescence in our report was shorter in the patients who had surgery (about four days), and prolonged (fourteen days) in the patient managed conservatively. Considering the pain, embarrassment and sequelae of erectile dysfunction and impotence, early surgical intervention is advocated. It also results in early resolution of symptoms, shorter hospital stay and less family dysfunction.

**Recommendation**

Priapism is an uncommon complication of sickle cell disease which if not treated promptly results in impotence. Counselling for prevention, early detection and prompt management should be part of the counselling session given to male patients with SCD and parents. Surgical intervention is more effective and thus advocated.

**References**