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NEAR EXHAUSTION OF MEDICAL TREATMENT OPTIONS AVAILABLE FOR PRIAPISM IN A CHILD WITH SICKLE CELL ANAEMIA FROM YOLA, NIGERIA: A CASE REPORT

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Abstract: *Priapism could be described as painful penile erection unrelated to sexual stimulation, which may not subside without medical intervention. This report described a case of near exhaustion of medical treatment options available for priapism in a child with sickle cell anaemia from Yola, Nigeria. The diagnosis was based on non response to the different stages of medical intervention for the condition. He is a known sickle cell anaemia patient who presented with painful, tender and sustained penile erection of seven hours. Blood film showed numerous sickled red cells and packed cell volume of 28 %. Having ruled out other possibilities, a diagnosis of priapsm in a child with background sickle cell anaemia was made, and the patient was placed on antioxidants, sedation, analgesic and ice packs without response. Next was hydration which was also not successful. Aspiration of the copora cavernosum led to detumescence followed by a rebound penile erection. Intra-coporal injection of epinephrine was also not remarkable. Dexamethasone, a steroid was further used and the child had complete detumescence. He was subsequently discharged on follow-ups. Now that surgical intervention is becoming obsolete, current case suggests the need for more studies to investigate the role of steroids and other newer treatment options to further expand on the current medical interventions.*

Keywords: Priapism, Younger children, Sickle cell anaemia, Medical interventions, Steroids, Yola-Nigeria.

INTRODUCTION

The word priapism was coined out from the Greek god of fertility called Priapus (Cherian et al, 2006). Priapism is defined as inappropriate penile erection that does not return to its flaccid state, despite the absence of both physical and psychological stimulus (Cherian et al 2006, and Brian et al 2008). The condition is thought to be as a result of pooling of blood in the corpus cavernosum of the penis leading to prolonged erections. The duration of each episode of priapism as per the definition is still not clear because some authors have reported few hours, while several others have published a duration of up to a day (Cherian et al 2006, Brian et al 2008, and Emond et al 1980). Although the mechanisms leading to priapsm are poorly understood, complex neurologic and vascular factors were linked to it (Brian et al 2008). Haematological disorders like sickle-cell anaemia, leukemia and thalassemia; neurologic disorders such as spinal cord lesions and spinal cord trauma were associated with priapism. Because NADPH is a co-factor involved in the

formation of nitric oxide leading to priapism; Finley in 2008 reported that glucose-6-phosphate dehydrogenase deficiency might generate high levels of nitric oxide, and adenosine that are associated with vasodilation and increase penile blood flow leading to priapism. Medicaments associated with priapism include anti-hypertensives, antipsychotics, anticoagulants, and recreational drugs (Cherian et al 2006).

Apart from the established guidelines for managing priapism, other medical treatment options are still being studied (Cherian et al 2006). Difficult cases of priapism that could exhaust this guideline may be on the rise now, thereby, necessitating the need to expand the present medical options (Cherian et al 2006, Brian et al 2008, and Emond et al 1980). Other medical options under study include hydroxyurea, gabapentin, terbutaline, methylene blue and baclofen.

Most cases of priapism with background sickle cell anaemia were reported in adolescence and adult and majority of patients responded to simple analgesics, ice packs, sedation and or hydration (Cherian et al 2006, Brian et al 2008, and Emond et al 1980). There is a gap in knowledge of this condition especially among younger children. More so that almost all stages of medical interventions were employed in current case, a situation that is rarely heard off among clinicians. This paper reports the near exhaustion of medical treatment options available for priapism in a child with sickle cell anaemia from Yola, Nigeria.

CASE SUMMARY

A 6 year old known sickle cell anaemia child presented to our health facility with painful sustain penile erection of seven hours without prior trauma to the penis. There were no fever, diarrhea or vomiting. No known neurologic or spinal anomaly and child is not on any drugs apart from his routine folic acid and paludrine. Other than the sickle cell anaemia, features suggesting other common haematological conditions that could give similar presentation were absent. Physical examination revealed an erect tender penis (Fig 1 and 2). Vital signs were normal and review of systems was unremarkable. Blood film showed sickled red cells, but the percentage of sickled red cells in relation to total red cells was not estimated due to logistics reason. Full blood count and differentials and erythrocyte sedimentation rate were within normal limits. Packed cell volume was 28 %. Diagnosis of priapsm in a child with background sickle cell anaemia was made, and the patient was placed on antioxidants, sedation, analgesic and ice packs. He did not respond to this initial treatment; next was hydration which was also not successful. Aspiration of the copora cavernosum led to immediate flaccidity, then a rebound of penile erection. Intra-coporal injection of epinephrine was carried out with slight success. Dexamethasone, a steroid was further introduced and the child had complete detumescence, and he was subsequently discharged on follow-ups.

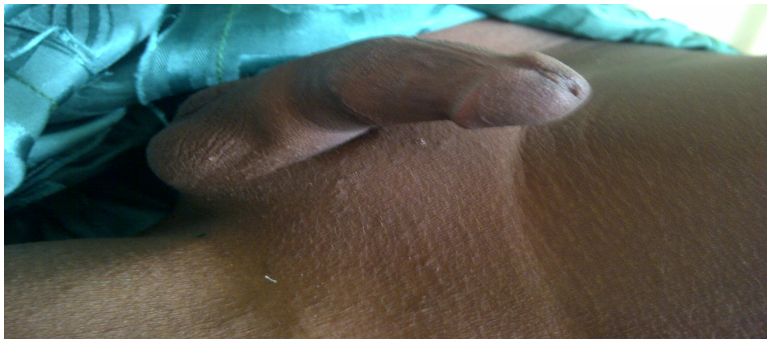

Fig 1: Lateral view of sustain penile erection (priapism)

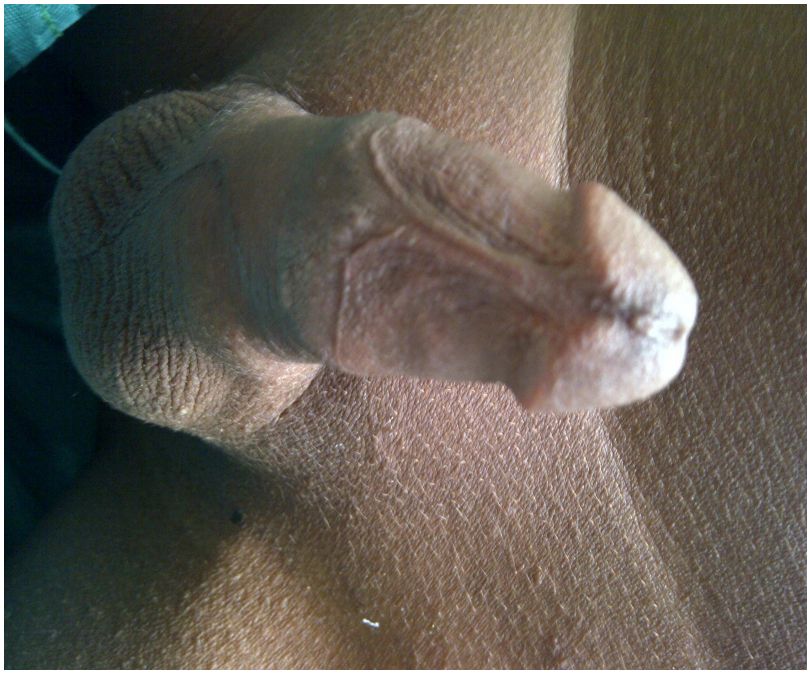

Fig 2: Dorsal view of sustain penile erection (priapism)

DISCUSSION

Penile erection is achieved when blood flow to the corpora cavernosum is increased and outflow reduced. Both sickled red cells and vascular endothelium contribute to pooling of blood and also reducing blood outflow from the penis. Sickled red cells microvascular thrombosis, endothelial hyperplasia and rouleaux formation had been associated with gradual stagnation of blood and reducing its out flow from the penis (Cherian et al 2006, and Brian et al 2008). This phenomenon is known as low flow priapism and is potentiated by vascular sialic acid anomaly and loss of membrane phospholipid from vascular endothelial cell damage (Brian et al, 2008).

For a successful management of priapism, treatment has been recommended in phases; starting with the use of oxygen, analgesics, ice packs and sedation (Farrer et al, 1961). These were used on current case without success. A similar observation was made by Krauss and Fitzpatrick in 1961. Nonetheless, however, improvement with the aforesaid treatments had been documented by other authors (Cherian et al 2006, Brian et al 2008, and Farrer et al 1961). The patient was then commenced on hydration, alkalization and pseudoephedrine. All treatment modalities above were aimed at reducing pain, blood viscosity, prevent sickled red cells from being formed and to enhance outflow of blood from the penis. Yet, penile tumescence did not resolved, which led to aspiration of copora cavernosum of the penis. Success achieved with aspiration of copora cavernosum was however short lived. Divergent reports on aspiration of copora cavernosum have been documented, while some had recorded success, other clinicians were unsuccessful (Cherian et al 2006, and Farrer et al 1961). Intra-coporal injection of epinephrine to constrict the vessels thereby reducing in flow of blood but at the same time promoting blood outflow from the penis was carried out with slight success also. Similar finding was made by Bos and Buys in 1994; however, complete detumescence with this treatment modality was published by other clinicians (Cherian et al 2006, Brian et al 2008 and Montague et al 2003). Constant hemodynamic monitoring is required with the use of phenylepinephrine because the drug can cause hypertension, tachycardia, and arrhythmias. Gladly, these did not occur in the index case.

Other medical treatment options include exchange blood transfusion or hyper-transfusion and steroid, a newer medical option. Detumescence was fully achieved in present case with steroid administration. This treatment modality is relatively new (Abern et al, 2009). Not only is success rate of steroid good, it also has an added advantage of preventing reoccurrence of priapism (Abern et al, 2009). Exchange blood transfusion or hyper-transfusion are usually left as last resort after all medical management options have failed. The index case was being prepared for hyper-transfusion which is aimed at keeping his packed cell volume at least 30% (Cherian et al 2006, Brian et al 2008 and Montague et al 2003). By doing so, it is hoped that the amount of sickled red cells in circulation will be less than 30%. Fortunately, he responded to steroid therapy. Of note is the controversy that exists with the use of anti-androgens like oestrogens and gonadotrophins (Montague et al 2003, and Levine et al 1993). In a nutshell, many workers are of the opinion that the use of anti-androgens and gonadotrophins will benefit mainly adolescent that had sexually matured, and better still if priapism is of idiopathic origin (Cherian et al 2006, Brian et al 2008, Emond et al 1980, Farrer et al 1961, Montague et al 2003, and Levine et al 1993).

Surgical management of praipism (shunts) is now becoming old fashion because most cases respond to one or combination of medical treatments. The use of shunts attempt to reverse blood flow from the rigid corpora cavernosum into the corpus spongiosum, which contains the glans and the urethra. The result of this approach was generally poor. Not only does the surgery often fail to resolve the priapism, but the procedure itself risks inducing impotence (Cherian et al 2006, Brian et al 2008, and Emond et al 1980). Proximal shunts of Quackel's, distal shunt of winter’s, corpora cavernosa and saphenous vein shunt of Grayhack are some of the surgical treatment of

priapsm (Cherian et al 2006). Penile prosthesis can be implanted where dense fibrosis had occurred as a complication of prolonged priapism.

Considering that clinicians are faced with challenges while managing cases of priapism, this report provided information on the contributory role of steroid in treating cases of childhood priapism. Secondly, this information could have public health relevance to policy development and program implementation, as it relates to the overall management of childhood priapism.

CONCLUSION

Sickle cell anaemia is an important causative factor of priapism, a urological emergency of public health importance. Prompt intervention in straight forward cases permits full recovery; however, there are exhaustive cases akin to index case being encountered in clinical practice. In this regard, there is need for more studies to investigate the role of steroids and other newer treatment modalities in order to expand the current medical interventions. Especially now that surgical intervention is becoming obsolete.

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