Abstract

Neonatal priapism is a rare occurrence with uncertain etiology and unestablished management. Since adult priapism carries the risk of erectile dysfunction later in life, it can create significant anxiety for parents and caregivers. Historically, management of neonatal priapism has been mainly conservative. We present two cases of neonatal priapism in preterm infants, with one patient workup revealing mild polycythemia. We discuss their management and review the literature.

ABBREVIATIONS

CDU: Color Doppler Ultrasound

INTRODUCTION

Priapism is defined as a prolonged, persistent penile erection not associated with sexual stimulation or desire [1]. Multiple types of priapism in children have been described and include ischemic (veno-occlusive, painful), stuttering, non-ischemic (arterial, usually painless), and neonatal [2]. Neonatal priapism is defined as a prolonged and persistent erection lasting greater than or equal to four hours. It is relatively rare, with only 19 cases reported in the literature since 1876 [3,4]. In most cases, the exact cause is unclear. Idiopathic neonatal priapism is the most common etiology, occurring spontaneously and unassociated with stimulation [1]. In premature neonates, polycythemia is also a relatively common cause [2]. The condition usually resolves spontaneously, but there are currently no guidelines regarding the management of neonatal priapism.

CASE PRESENTATION

Case 1

A 20 day-old infant presented to the emergency room with a persistent erection. The patient was the product of a 32-week gestation. The parents did not recall when the erection started but the penis was erect for “several days.” It was noted during every diaper change that the infant had an erection. The boy was feeding and urinating regularly and without difficulty and was in no apparent pain or discomfort. Urologic consultation was obtained. On examination, the patient was uncircumcised, without any penile cyanosis or swelling, and with descended testes, bilaterally. The penis was tumescent. Both corpora were firm and the glans was nontender, soft, and non-cyanotic. Workup included a complete blood count, penile and scrotal duplex ultrasounds to assess arterial and venous blood flow (Figure 1), pelvic ultrasound to rule out masses, and a sickle cell preparation. Complete blood count revealed an elevated hematocrit of 57 g/L with a normal sickle cell panel.

Case 2

Urologic consultation was obtained for a day-old male in the neonatal intensive care unit. The child was the product of a 28-week gestation and had an erection that lasted over 12 hours prior to consultation. The infant was urinating regularly and without difficulty and was in no apparent pain or discomfort. On examination, he was uncircumcised, without any penile cyanosis or edema. The penis was tumescent, both corpora were erect, and the glans was soft. Workup also included a complete blood count, penile and scrotal duplex ultrasound (Figure 2), pelvic ultrasound, and a sickle cell preparation. In this case the complete blood count revealed a normal hematocrit of 41.9 g/L. The sickle cell panel was negative.

Both patient’s erections resolved after approximately two days with careful observation and without any treatment or intervention. They were seen in follow-up approximately three weeks later without any history of recurrent prolonged erection since discharge. On examination, the penis was normal and uncircumcised without an erection in both cases.

DISCUSSION

Priapism is considered a urological emergency with
management directed at prevention of penile disfigurement and preservation of future erectile function. Classification of priapism includes both ischemic and non-ischemic. Priapism in childhood is usually ischemic and a result of multiple etiologies as shown in Table 1 [2].

Non-ischemic priapism is rare in children and is usually idiopathic, with subclinical perineal birth canal trauma hypothesized to be the cause resulting in a benign, non-ischemic erection [2]. Other causes of primary neonatal priapism include polycythemia, infection, respiratory distress syndrome, and umbilical artery catheterization [2,3]. Although sickle cell disease in the most common cause of childhood priapism, the high levels of fetal hemoglobin in the newborn are protective against vaso-occlusion and resultant priapism in newborns with sickle cell disease [4].

Since it is common for healthy newborns to have an erection, neonatal priapism must be differentiated from prolonged physiological erections, which are benign and last less than four hours. Neonatal priapism is a clinical diagnosis, with a persistent erection lasting greater than or equal to four hours without any scrotal or penile discoloration, e.g. cyanosis, and no apparent pain. The duration has reportedly ranged from two to twelve days and does not seem to be associated with any long-term sexual dysfunction [1].

The process of tumescence includes a latent phase, with relaxation of arteries in the corpus cavernosum and sinusoidal smooth muscle leading to an increase in arterial influx and capacitance, respectively. The subsequent tumescent phase results from sinusoidal blood trapping. The full erection phase is caused by sub-tunica venous plexus compression leading to stretching of the tunica albuginea and resultant occlusion of emissary veins [5]. Lastly, the rigid erection phase results from contraction of the ischiocavernous muscles, which increases pressure in the corpus cavernosum to above systolic blood pressure for short periods [6]. Although the corpus spongiosum and glans experience increased arterial influx, they lack a tunica albuginea and therefore act mainly as an arteriovenous shunt [2]. An erection can be caused by stimulation including genital/ reflexogenic and central/psychogenic or central origination/nocturnal, i.e. androgen release during rapid eye movement sleep in adolescence [6]. Reflexogenic erections are physiologic in neonates and children and can occur during bathing, diaper changing, urethral catheterization, or with a full bladder; however, detumescence should occur once the stimulus is removed.

The postulated pathophysiology of priapism varies depending on the subtype. Ischemic priapism, according to Hinman’s classic theory, is a result of reduced blood flow and congestion leading to increased blood viscosity and the resultant dark, deoxygenated blood observed during corporal aspirations [7]. The ischemia leads to necrosis followed by fibrosis of the corpus cavernosum smooth muscle which causes erectile dysfunction and penile distortion [2]. The exact pathophysiology of stuttering priapism, or recurrent prolonged erections, is poorly understood. It is believed to have a similar etiology as ischemic priapism and is most commonly caused by sickle cell disease in children. Proposed mechanisms of stuttering priapism include over-responsiveness to androgenic or sexual stimulation resulting from corpus cavernosum endothelial nitrous oxide deficiency leading to reduced tonic corpus cavernosum smooth muscle tone, adrenoceptor impairment, transforming growth factor-beta upregulation, intracavernous venule scarring, and an abnormal central nervous system control mechanism [2,7]. Non-ischemic priapism is most commonly caused by an arterio-sinusoidal fistula. Penile, perineal, or pelvis trauma, usually from straddle or coital injury, leads to laceration of an artery or arteries, most commonly the cavernous arterioles in the crura or corporal bodies. This leads to formation of the arteriolar-sinusoidal fistula.

<table>
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<th>Table 1: Etiology and incidence of neonatal priapism.</th>
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<td><strong>Etiology of Childhood Priapism</strong></td>
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<tr>
<td>Sickle Cell Disease</td>
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The resultant high-flow state and sinusoidal blood pooling causes mechanical stimulation of the endothelial nitrous oxide synthase and smooth muscle relaxation. Although non-ischemic priapism without a high-flow hemodynamic state, e.g. fistula, may occur, the etiology is unknown [2].

All published cases of neonatal priapism have reported full functional recovery, independent of etiology, duration, or management. Seventy-five percent of cases managed by observation resolved spontaneously [3]. Although reported cases that have undergone a workup were found to be non-ischemic, the risk of untreated ischemic priapism is cavernous body fibrosis and resultant erectile dysfunction. For this reason, initial testing with color Doppler ultrasound (CDU) in neonatal priapism is recommended to confirm that it is the non-ischemic subtype before deciding on conservative management. CDU is a noninvasive, relatively inexpensive, and safe method of detecting scrotal and penile blood flow as well as rule out pelvic masses. Corporal aspiration and blood gas analysis should be reserved for cases without demonstrable arterial blood flow via CDU or in which ischemic priapism is otherwise suspected. A complete blood count, sickle cell panel, and C-reactive protein have been suggested in the literature and could be considered in the initial work-up, especially when there is concern for infection, polycythemia, sickle cell disease, or leukemia [1,3]. In clinical situations associated with polycythemia, such as this case, studies have suggested performing a red cell volume reduction (venesection) or phlebotomy and partial transfusion [1,4]. We managed our cases with close observation and noted spontaneous resolution without sequelae suggesting that not all cases of neonatal priapism with concomitant polycythemia require intervention. Furthermore, there may be a threshold at which an elevated hematocrit associated with neonatal priapism required intervention. Future work might aim to identify this threshold.

In this study we present two cases of non-ischemic neonatal priapism, one case associated with polycythemia, that both resolved within two days with minimal workout and without intervention. All published cases of neonatal priapism have reported full functional recovery, independent of etiology, duration, or management, suggesting conservative management of neonatal priapism is recommended. However, because the risk of untreated ischemic priapism is cavernous body fibrosis and resultant erectile dysfunction, we recommend initial testing with color CDU in neonatal priapism to confirm non-ischemic subtype before deciding on conservative management. Blood gas analysis of aspirated corporal blood is reserved for select cases.

REFERENCES