

Pediatrics

Idiopathic non-ischemic priapism in an infant: A case report

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ABSTRACT

Priapism is a pathologic erection lasting >4 hours that is unrelated to or persists beyond sexual stimulation. Priapism is rare in children, and rarely reported in infants. Although a small number of cases have been reported in neonates, even fewer have been reported in infants outside of the neonatal period. Due to its rarity and poorly understood pathophysiology, the diagnosis and management of priapism in infants is challenging. We report a rare case of idiopathic non-ischemic priapism in a 9-month-old highlighting some of the challenges in diagnosis and management of priapism in this age group.

Introduction

Priapism is typically defined as a prolonged erection lasting >4 hours unrelated to or persisting beyond sexual stimulation, and is rare in pediatric patients outside of the sickle cell population. Priapism is classically divided into 2 categories: ischemic (low-flow, veno-occlusive), or non-ischemic (high-flow, arterial). Ischemic priapism is the most common type in children and usually presents with pain. Emergent intervention is needed in order to prevent long-term damage to erectile tissue. Non-ischemic priapism is a partial erection caused by unregulated cavernosal arterial flow, is typically painless and frequently associated with perineal trauma. Neonatal priapism presenting as a non-painful prolonged erection in the first 28 days of life has also been described, with fewer than 20 cases reported in the literature.¹ The pathophysiology of neonatal priapism is poorly understood, but is thought to represent a form of non-ischemic priapism. Our current understanding of the etiology and management is based on a limited number of case reports. Outside of the neonatal period, few cases of priapism have been reported in infants. We present a rare case of non-ischemic priapism in a 9-month old.

Case presentation

A 9-month-old white male with no prior medical or surgical history presented to the emergency room with a persistent painless erection for 8 hours. He was not taking any medication. There was no associated penile or perineal trauma. According to the parent, he had been

behaving normally throughout the day and had been voiding normally. The patient has no prior history of priapism. Physical exam revealed a normal circumcised erect phallus. The corpora cavernosa were rigid with a flaccid glans. Laboratory evaluation including a complete blood count with differential and a basic metabolic panel were normal with the exception of an elevated white blood cell count of 14.76 thousand. Penile color doppler ultrasound showed an erect penis with patent cavernosal arteries bilaterally and dorsal penile artery with no sign of an arteriovenous fistula (Fig. 1). At the conclusion of the ultrasound, the penis had detumesced and remained flaccid. On a follow-up phone call the following day, mom reported normal erections with no recurrence of the priapism.

Discussion

Priapism is the presence of a prolonged erection unrelated to or persisting beyond sexual stimulation. There are two categories of priapism, ischemic and non-ischemic priapism. Priapism is rare in children, but ischemic priapism is commonest among pediatric patients. The diagnosis of priapism can be made with history and physical exam. Cavernosal blood gas can be used to distinguish ischemic from non-ischemic priapism. However, this requires aspiration of blood from the cavernosal bodies, and may be difficult to perform in an infant without sedation or anesthesia. Penile Doppler ultrasonography can also be used to distinguish ischemic from non-ischemic priapism, but can also be difficult to perform and to interpret because it is infrequently utilized in children and infants. Neonatal priapism thought to be a form of non-

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ischemic priapism in infants within the first 28 days of life has been reported rarely in the literature.¹ Although the exact etiology of neonatal priapism is poorly understood, reported cases in the literature were not associated with pain.¹ The majority of cases had no identified underlying causes, and resolved spontaneously with observation alone and with no long-term consequences.¹ Polycythemia was noted as a possible etiology in 4 cases and was treated with observation, phlebotomy, or exchange transfusions.¹ While the incidence etiology, and management of priapism in children have been extensively reviewed, there is a paucity of documented cases of priapism in infants outside of the neonatal period.¹ We report a rare case of non-ischemic priapism in a 9-month-old male.

There are few reports of non-ischemic priapism in infants and young children outside of the neonatal period. Griffin et al. report a similar case of a 4-month-old male with non-painful priapism lasting 1 day that resolved spontaneously. Laboratory exams were normal, but no additional evaluation was done.² Hammond et al. reported three cases of priapism associated with transverse myelitis in 5-month old, 7-month old, and 12-month old infants. Evaluation with corporal blood gas or penile ultrasonography was not pursued.³ Ozbek et al. described a case of non-ischemic priapism in a 2-year old with history of perineal trauma. He was found to have a dermal sinus tumor at the base of the penis causing the priapism which resolved with treatment of the tumor.⁴ Ozturk et al. reported a case of non-ischemic priapism in a 2-year old with history of perineal/penile trauma. In this case, angiography was performed, showing pseudoaneurysms from cavernosal/bulbourethral artery with fistulation to the corpora cavernosa. The priapism resolved with embolization.⁵

The main goal of treatment is preservation of erectile function.

Untreated ischemic and recurrent priapism can lead to erectile dysfunction, which has been reported in children and adults. Although follow-up for infants with priapism is limited, there are currently no reports of erectile dysfunction or other long-term complications following episodes of non-ischemic priapism.

Most cases of non-ischemic priapism in infants reported in the literature are idiopathic, and did not require treatment. For those with a cause, priapism resolved with treatment of that underlying cause. While observation alone is sufficient for the majority of cases, additional treatments to consider are phlebotomy and/or partial exchange transfusion for patients with polycythemia, and ketamine infusion. Surgical intervention could be considered as a treatment option, although there are no reports of surgical treatment for infants with idiopathic non-ischemic priapism.¹

There are few reports of non-ischemic priapism in an infant outside of the neonatal period in the literature. Similar to the findings in neonatal priapism, our patient presented with non-ischemic priapism that resolved spontaneously without intervention and with no clear underlying cause. This case highlights the challenges that exist in differentiating ischemic and non-ischemic priapism in infants, and supports the approach of initial observation.

Conclusion

Priapism in infants outside of the neonatal period is exceedingly rare. We report a case of a 9-month-old male with idiopathic non-ischemic priapism diagnosed using penile Doppler ultrasound that resolved spontaneously.

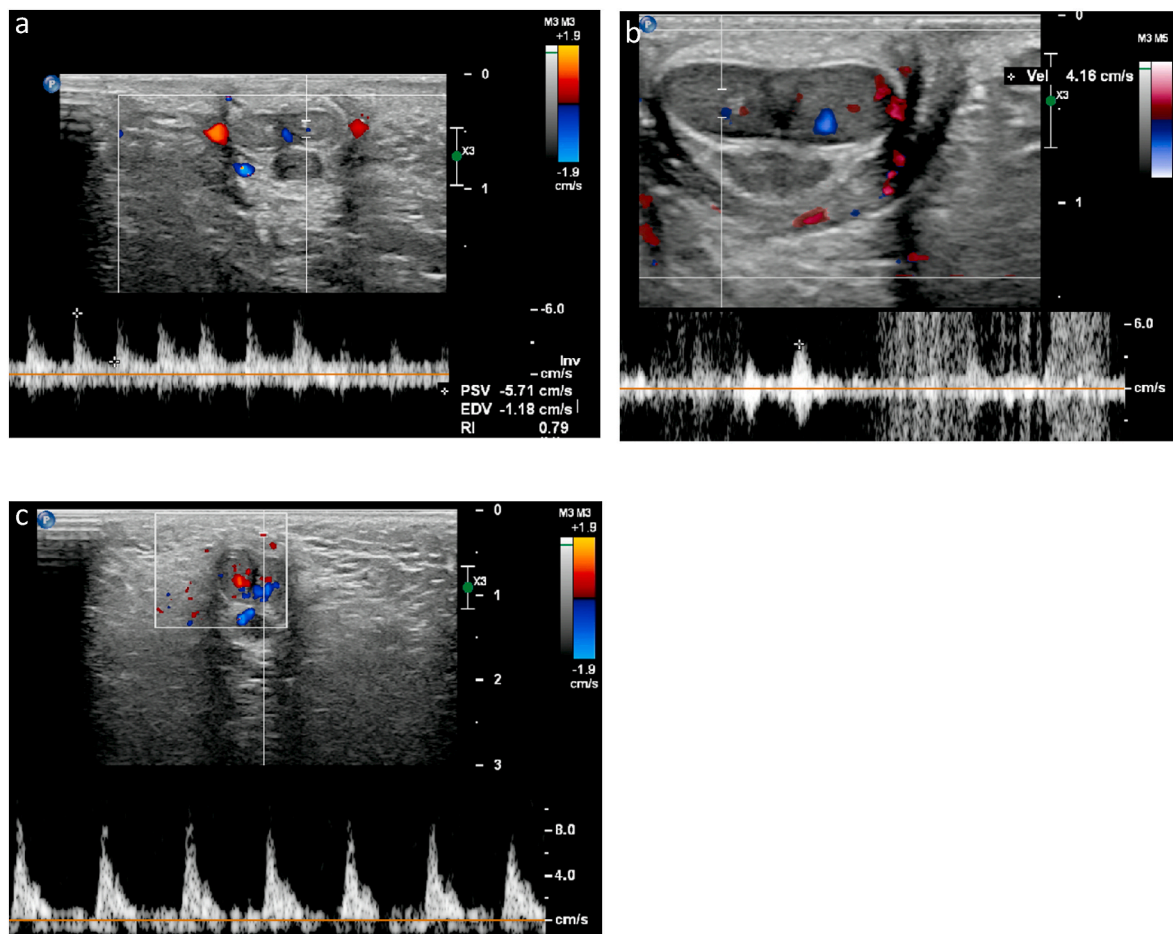


Fig. 1. Penile doppler ultrasonography showing arterial flow to the left corpus cavernosum (a), right corpus cavernosum (b), and dorsal artery of the penis (c).

Declaration of interest

None.

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CRediT authorship contribution statement

Luke L. Wang: Conceptualization, Investigation, Visualization, Roles/, Writing - original draft, Writing - review & editing. **Claudia Berrondo:** Conceptualization, Project administration, Supervision, Writing - review & editing.

Declaration of competing interest

The authors declare no conflicts of interest.

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