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CASE REPORT

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Priapism as an initial clinical manifestation in hematological disease: A case report and literature review

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Abstract

Priapism is defined as an erection lasting more than 4 hours and is considered a urological emergency in pediatrics. The goal of treatment is to prevent both short and long-term erectile dysfunction. The incidence in boys has not yet been estimated because priapism is considered a rare clinical manifestation. A 15-year-old male was transferred to our clinic with priapism of 48 hours' duration, following two unsuccessful attempts to drain the corpora cavernosa at another facility. Preoperative evaluation revealed hyperleukocytosis with leukemic blasts in peripheral blood, prompting a hematology consultation for a multidisciplinary approach. In the operating room, aspiration and lavage of the corpora cavernosa were performed. Due to unsatisfactory results, a percutaneous shunt procedure was attempted, followed by a corpora cavernosa–spongiosum shunt. Postoperatively, hematology continued to monitor the patient, eventually diagnosing him with type B lymphoblastic leukemia as the underlying cause of the priapism episode. Priapism is an uncommon clinical manifestation in the onset of systemic diseases, with hematological causes being the primary etiology. The diagnosis is clinical, based on a comprehensive medical history, physical examination, and complementary studies, including blood gas analysis of the corpora cavernosa and penile color Doppler ultrasound. Treatment is staged, beginning with patient stabilization and pain control, followed by more invasive surgical interventions if necessary. The understanding and management of priapism in children remain challenging for healthcare providers, and further studies are needed to ensure timely and effective management.

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Introduction

Priapism is a prolonged total or partial erection of the penis that lasts 4 hours, unrelated to sexual stimulation (1). It is considered a urological emergency, and its management aims to prevent erectile dysfunction, shortening of the penis and psychological consequences in the sexual sphere. It has been observed in the pediatric population that, once puberty begins, adolescents may fear painful erections during masturbation or normal sexual activity.

The incidence in men of any age is estimated at 0.3 to 1.5 per 100,000 urological emergency consultations per year, most frequently affecting men in their fifth decade of life (2). The reporting of cases of pediatric priapism is underestimated because it is considered rare, often due to the shyness of adolescents or their parents, who may be reluctant to seek medical attention immediately, especially in an environment like ours where health professionals continue to face taboos and prejudices about sexuality.

Our aim was to present a rare case of pediatric low-flow priapism as the initial manifestation of acute lymphoblastic leukemia, describe its diagnostic and therapeutic management, and review the literature to highlight pathophysiology, clinical approach, and prognosis in childhood hematologic-associated priapism.

Case description

A 15-year-old male, born via vaginal delivery at 39 weeks of gestation, had no significant pathological or surgical history, except for a maternal history of arterial hypertension. According to his mother, the condition began with two episodes of vomiting containing food, accompanied by dysuria, bladder tenesmus, and a sustained erection. She also reported that, one month earlier, he had experienced generalized pallor, night sweats, and polydipsia. Initially, they presented to a secondary care hospital, where an aspiration puncture of the corpora cavernosa was attempted without success.

Laboratory tests were performed, revealing hyperleukocytosis, severe anemia and blasts in peripheral blood. Initial support was provided, and the decision was made to transfer the patient to our health center. We received a patient with priapism 10/10 of apparent hematological origin, evolving for 48 hours. The laboratory test was repeated, showing the following results: hemoglobin 8.2 g/dl (11.8 - 16 g/dl), hematocrit 28.2% (35-48%), leukocytes 343,400/mm3 (4,700-12,200/mm3), platelets 98,000/mm^3 (150,000 – 450,000/mm^3), peripheral blood blasts 53%, CRP 16.20 mg/dl (0-0.50 mg/dl). A color Doppler ultrasound of the penis was performed, showing slight thickening of the soft tissues at the level of the glans, consistent with edema, irregular heterogeneous avascular cavernous bodies, with no evidence of flow on color Doppler (Figure 1). He was evaluated by the Hematology and Pediatric Urology services, who suggested an initial diagnosis of acute leukemia accompanied by low-flow priapism. Following informed consent, treatment was initiated with tumor lysis syndrome prophylaxis, including oxygen therapy, hyperhydration, hydroelectrolyte correction, and platelet concentrate transfusion. This was followed by opioid-based analgesia and local cooling while the operating room was being prepared for urological intervention.

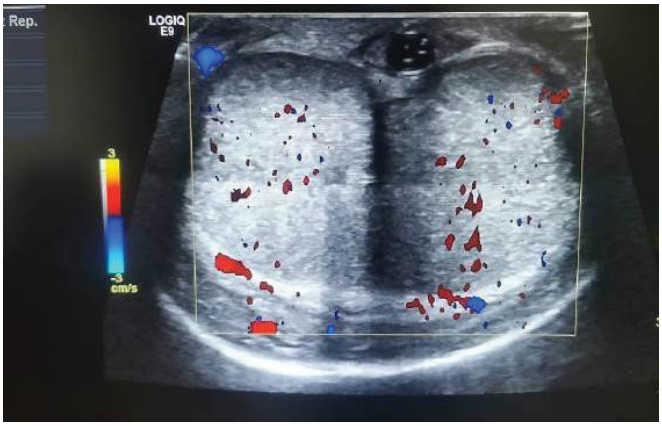

Figure 1: Color doppler ultrasound of the patient's penis

With the support of the anesthesiology team and under general anesthesia and ketamine supplementation, we performed an aspiration and lavage of the cavernous bodies without satisfactory reduction. Subsequently, an intracorporeal injection of diluted epinephrine (1ug / ml) 15 milliliters was administered twice, resulting in an improvement of 8/10. Next, a percutaneous distal fistula technique was performed with the Winter technique through the introduction of 18G catheters in the apex of the glans parallel to the urethra, showing little output of thick, dark blood. Finally, an open distal cavernous spongy fistula with the AL-Ghorab

technique resulting with an improvement of 6/10 (Figure 2).

After the procedure, the bone marrow aspiration and immunophenotyping were performed, resulting in a diagnosis of common B-type acute lymphoblastic leukemia with Philadelphia translocation t (9;22) with P210 BCR-ABL 1 fusion transcript. The patient remains under the care of the hematology team and, at the time of this presentation is receiving chemotherapy following to the BFM-2009 protocol in the induction phase, which is based on corticosteroid therapy and cyclophosphamide.

Literature review and discussion

Incidence

Case reports of childhood priapism are scarce and often represent an initial sign of hematological disorders. Approximately 65% pf cases are associated with sickle cell disease, followed by 10% linked to acute leukemia, trauma and idiopathic origin, and 5% attributed to drugs such as: trazodone, fluoxetine, methylphenidate and sildenafil (1).

Approximately two - thirds of patients experiencing an acute episode of priapism report previous intermittent episodes (3), which frequently go unnoticed due to self-perception of normality.

Pathophysiology and classification

The Consensus on the Evaluation and Treatment of Priapism, under the auspices of the American Foundation for Urological Disease and with the multidisciplinary participation of experts in pediatrics, hemato -oncology, psychiatry and urology, classified priapism into the following categories (4):

- Low flow, venous, ischemic: This is the most common form of priapism, characterized by a

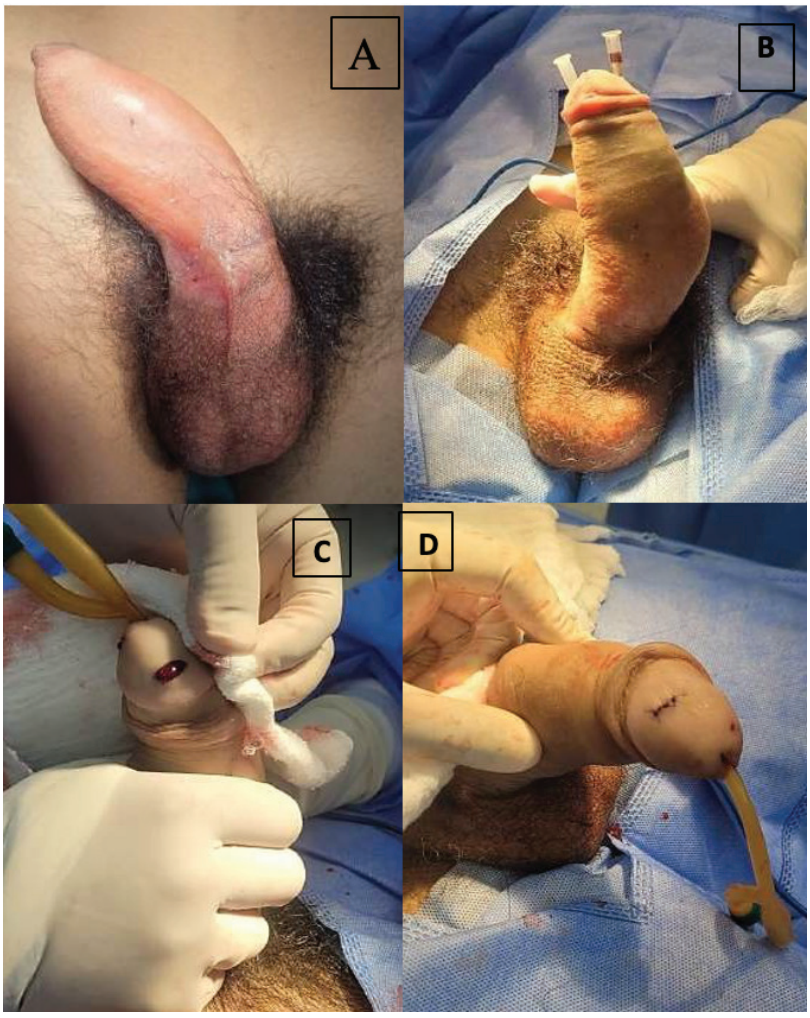

Figure 2: A. Priapism in a patient in a pre-surgical state, B. Percutaneous fistula with Winter technique, C and D. Open distal cavernospongiosus fistula with AL-Ghorab technique

painful erection with maximum rigidity. Clinically, it is defined by the absence of cavernous blood flow and behaving like a compartment syndrome.

- High-flow, arterial, non-ischemic: This form is almost always secondary to traumatic or iatrogenic causes due to the formation of arteriocavernous fistulas. It is not painful and does not present maximum rigidity. Although non-ischemic priapism requires timely evaluation and care, it doesn´t represent a medical emergency as isn't a compartment syndrome.

In pediatrics, neonatal priapism has been reported, most cases being idiopathic. It has been attributed to subclinical trauma of the perineal birth canal. There aren´t reports of Ischemic cases or cases caused by agitation the newborn (2). In all documented cases, complete functional recovery is reported and suggests a favorable natural history of benign pathophysiology.

Hematological causes of priapism include hemoglobinopathies as the primary etiology, less frequently hyperviscosity states such as in lymphoproliferative syndromes (5).

In leukemic conditions, there is a reduction in venous return and impairment of the normal detumescence mechanism during prolonged nocturnal erections. This is considered a rare syndrome characterized by multiple and recurrent episodes of ischemic priapism, also known as intermittent priapism.

Erections may be initiated by genital (reflexogenic) stimulation, central stimulation (audiovisual events, fantasy or memory) or central in origin (mediated by androgens during REM sleep in adolescents). The flow of the corpora cavernosa is facilitated by nitric oxide (NO) and prostaglandins (PGI2), which play a key role in regulating the interaction between stagnant blood and the trabecular wall.

Ischemia and consequently hypoxia alters the synthesis of NO and PGI2 of the capillary endothelium, leading homeostatic imbalance. This process promotes platelet aggregation, increasing white blood cell adhesion, and facilitates thrombus formation and tissue damage. Finally, hyperviscosity is caused by an excess of leukemic blasts accumulates in the corpora cavernosa and dorsal veins of the penis (2). Another factor that contributes to venous congestion of the corpora cavernosa is the mechanical pressure on the abdominal veins by splenomegaly, a common sign in childhood lymphoproliferative syndromes.

Diagnosis

A thorough clinical history and focused questioning about the priapism are essential to determine its etiology. Physical examination, along with the identification of extraurological signs and symptoms such as weight loss, night sweats, and splenomegaly may suggest malignancy (5).

A complete blood count and coagulation profile are crucial to identify potential hematologic diseases. Additionally, arterial blood gas analysis is recommended in cases of suspected hypoxemia or systemic acidosis. The distinction between high- and low-flow priapism can be made by aspirating blood from the corpora cavernosa. In low-flow priapism, the aspirated blood will appear dark and acidotic, whereas in high-flow cases, the blood will be bright red, well-oxygenated, and exhibit low CO2 pressure (3).

Doppler ultrasound is particularly useful in cases of high flow priapism, as it allows us to identify an arteriovenous fistula responsible for sustaining the erection (3).

Treatment

Treatment should be tailored to the patient's condition, progressing from conservative therapy to minimally invasive procedures, and, if necessary, invasive surgical interventions (6).

Conservative therapy: This includes hydration, oxygen therapy, and cold compresses (cytoprotection). These measures should be started simultaneously with complementary etiological studies. However, in cases where sickle cell disease is suspected, the application of local cold therapy may exacerbate priapism. Opioid-based analgesia may also help inhibit tumescence (2).

Aspiration and lavage of the corpora cavernosa: For this procedure, it is recommended to use 23-21 G butterfly needles in prepubertal children and 19 G in adolescents. The needle is inserted laterally at 3 and 9 o'clock positions to avoid damaging the corpus spongiosum and urethra, Blood is aspirated in aliquots of 3–5 milliliters and the corpora are rinsed with warm 0.9% saline solution (2).

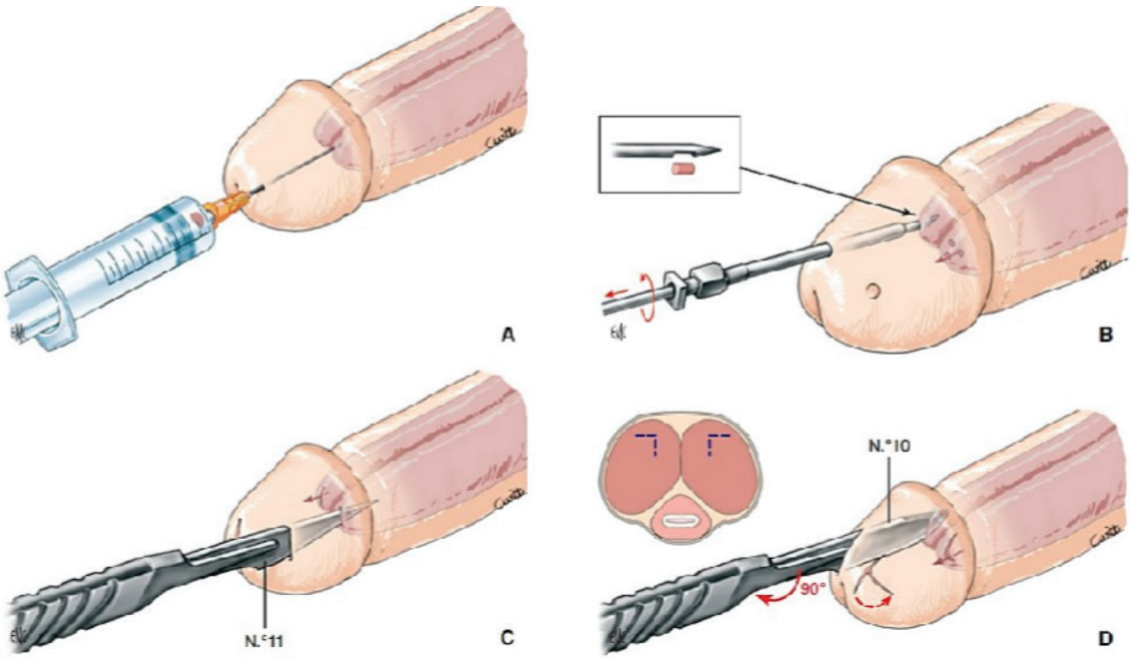

Figure 3: Percutaneous distal cavernospongiosus fistula, A. Needle puncture of the glans, B. Winter's technique, C. Ebbehoj's technique, D. Brant's technique (7)

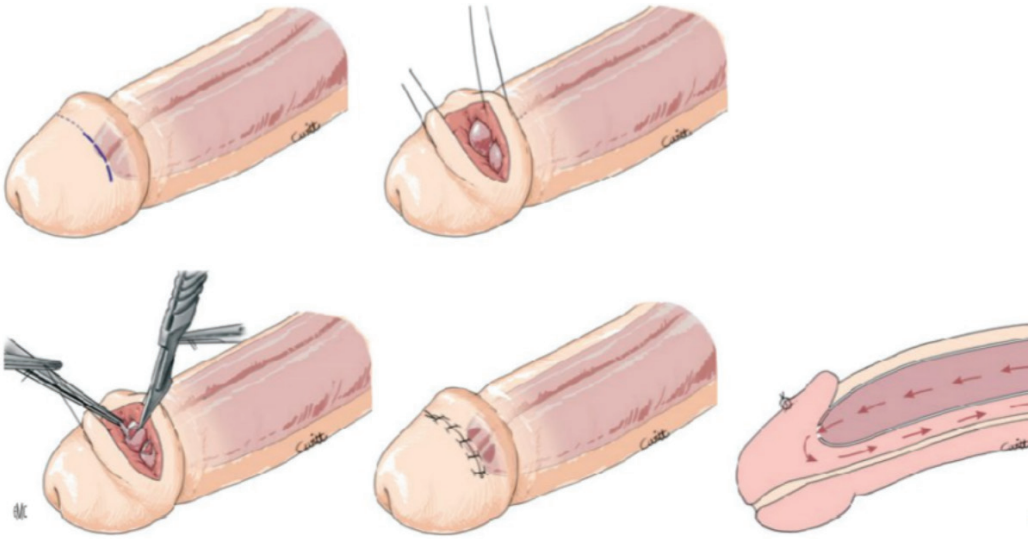

Figure 4: Open distal cavernospongiosus fistula according to the Al-Ghorab technique with cavernous dilation (7).

Intracorporeal injection: A diluted adrenaline infusion (1 µg/mL) may be administered. This sympathomimetic agent induces alpha-adrenoceptor-mediated contraction of cavernous smooth muscle and vasoconstriction of cavernous arteries (2). Although side effects such as dizziness, hypertension, reflex bradycardia, tachycardia, and arrhythmias are rare, patients should be carefully monitored during the procedure. This technique can be successfully performed under local anesthesia in children aged 4 to 18 years (6).

Ketamine anesthesia and caudal epidural catheter placement: Performed by anesthesiology in an operating room setting, this involves a caudal block, typically with bupivacaine. Ketamine has also been reported as an effective anesthetic agent for this procedure (6).

Penile derivations (invasive surgical techniques) (7):

Distal cavernospongiosus fistulas (first-line intervention): These are distal cavernospongiosus

anastomoses designed to drain thick, viscous, incoagulable and blackish blood through a fistula using a shunt. Its effectiveness is often temporary and its "blind" performance constitutes a risk, although insignificant, of injury to the urethra. These techniques can be classified as follows:

- Percutaneous: Winter technique, Ebbehoj and T-shunt, Brant (Figure 3).

- Open: Al-Ghorab technique (Figure 4).

Proximal fistulas (second-line intervention): If bilateral distal fistulas are properly performed but fail, proximal cavernospongiosus anastomoses are considered. These include: Quackels technique (proximal cavernospongiosus anastomosis via the perineal route), Grayhack technique (sapheno- cavernous anastomosis), or Barry technique (dorsal vein-corpus cavernosum anastomosis with or without a short saphenous graft) are still sometimes used, even though they are more invasive and more complex.

Forecast

The most critical prognostic factor for the preservation of erectile function is the duration of the priapism episode. In the adult population, priapism lasting less than 24 hours has been reported to result in a 92% likelihood of returning to baseline erectile function, compared to only 22% if the episode persists for more than 7 days (4). However, the prognosis may be more favorable in prepubertal and children. This relative protection could potentially be explained by the absence of androgenic activity, although this hypothesis remains under investigation (3). Currently, no prospective multicenter studies have evaluated the quality of sexual life following an episode of childhood priapism.

Conclusions

Priapism is an uncommon clinical manifestation that may present as an initial sign of systemic diseases, with hematological conditions being the leading cause.

Hyperviscosity states in which ischemia, hypoxia and disruptions in the homeostatic regulation of erection mediated by nitric oxide and prostaglandins occur, represent the pathophysiological mechanism of priapism in lymphoproliferative diseases.

The diagnosis of priapism is clinical, based on a through clinical history, physical examination and complementary studies such as cavernous body blood gases and color Doppler ultrasound which help delineate local ischemia. Treatment is then phased, starting with patient stabilization and pain management, followed by more invasive surgical interventions, such as spongiosacavernous or venocavernosal fistulas. Throughout this process, multidisciplinary management is essential, with emphasis on identifying and addressing the underlying disease.

Understanding and managing priapism in childhood remains a challenge due to the limited availability of case studies and clinical guidelines for appropriate management Additionally, there is a lack of empowerment regarding children's sexuality and the level of care available within the public health system.

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Contributions

Research concept and design: JPOC, DFGC, FAAV

Data analysis and interpretation: JPOC, DFGC, FAAV

Collection and/or assembly of data: JPOC, DFGC, FAAV

Writing the article: JPOC, DFGC, FAAV

Critical revision of the article: JPOC, DFGC, FAAV

Final approval of the article: JPOC, DFGC, FAAV

All authors read and approved the final version of the manuscript.

References

1. Patel SR, Reddy A, Dai M, Passoni N, Khera M, Koh CJ. Is urgent surgical management necessary for priapism in pediatric patients with hematologic conditions? J Pediatr Urol. 2022;18(4):528.e1-528.e6.
2. Donaldson JF, Rees RW, Steinbrecher HA. Priapism in children: a comprehensive review and clinical guideline. J Pediatr Urol. 2014;10(1):11-24.
3. Jesus LE, Dekermacher S. Priapism in children: review of pathophysiology and treatment. J Pediatr (Rio J). 2009;85(3):194-200.
4. Rodríguez Villalba R, Garcia S, Puigvert Martínez A, Pomerol I Montseny JM, Munárriz R. Priapismo [Priapism]. Actas Urol Esp. 2005;29(10):961-8.
5. Becerra-Pedraza LC, Jiménez-Martínez LE, Peña-Morfin I, Nava-Esquivel R, Villegas-Martínez JA. Priapism as the initial sign in hematologic disease: Case report and literature review. Int J Surg Case Rep. 2018;43:13-7.
6. Karagözlü Akgül A, Uçar M, Özçakır E, Balkan E, Kılıç N. Rare emergency in children: Priapism and stepwise treatment approach. Ulus Travma Acil Cerrahi Derg. 2022;28(4):464-70.
7. Bondil P, Carnicelli D. Tratamiento quirúrgico del priapismo. Urología. 2017;49(4):1-11.