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Review Article

Priapism in Chronic Myeloid Leukemia: Imaging Findings in Cases of Chronic Myeloid Leukemia who presented with Priapism

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Abstract

Priapism is a urological emergency and must be diagnosed and treated appropriately. Specifically in known cases of leukemia painful turgid penis should raise suspicion of priapism in first hand and penile Doppler should be the first line of imaging modality. This information should be forwarded to patient and emergency department clinician.

Keywords

Priapism; CML; Imaging

Introduction

Priapism is rare and is characterized by prolonged, painful and irreducible erection which doesn't result in ejaculation. Sickle cell anemia, chronic myelogenous leukemia, chronic lymphocytic leukemia, and acute lymphoblastic leukemia are hematologic disorders that can be its cause [1]. In adult leukemic patients, the incidence of priapism is approximately 5% [2]. Priapism as a result of hematologic malignancy is most likely caused by venous obstruction from microemboli/thrombi as well as hyper viscosity caused by the increased number of circulating leukocytes in mature and immature forms [3]. In cases of hematologic malignancy, controversy has existed regarding the optimal treatment of leukemic priapism. Earlier series of case reports show successful detumescence with local radiation therapy, open surgical shunting, or combination of the two treatments [4]. More recent literature has focused on the use of cytoreductive modalities such as chemotherapy or combination chemotherapy and leukapheresis [5]. Because of the relatively rare occurrence of leukemic priapism and the small number of case series, there is no standard treatment protocol. Chemotherapy or radiotherapy may first be attempted. If detumescence is not achieved, then surgical shunting should be considered. Priapism is a full or partial erection that continues more than 4 hours beyond sexual stimulation and orgasm [6].

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Discussion

The term priapism has its origin in reference to the Greek god Priapus, who had a disproportionate permanent erection [7]. Priapism is caused by imbalance of penile blood inflow and outflow and it may be either low-flow (ischemic) or high-flow (non-ischemic/arterial) type. Differentiation from non-ischemic variety must be done and apart from history and clinical examination, cavernosal blood gas analysis and Color duplex ultrasound (US) are currently the most reliable diagnostic methods of distinguishing ischemic from non-ischemic priapism.

Low-flow or ischemic priapism results from venous occlusion and manifests as painful, rigid erection. Intracavernosal blood sampling in such cases show acidosis (pH<7.25), hypercarbia (PCO_2 >90) and decrease in oxygen tension (PO_2<30) [6]. In contrast to low-flow priapism, intracavernosal blood sampling from patients with high-flow priapism reveal bright red oxygenated blood (pH =7.4, PCO_2<40, PO_2>90) [8].

Color duplex ultrasound (US) of the penis and perineum is recommended in the evaluation of arterial priapism because it can identify approximately 70% of cases and can differentiate arterial from ischemic priapism as an alternative or adjunct to blood gas analysis [9,10]. Ultrasound should be performed in the lithotomy position and examination of the entire penile shaft and perineum is recommended. In high-flow (non-ischemic/arterial) priapism US will show turbulent flow at the fistula, which helps to localize the site of trauma since patients with arterial priapism have normal to high blood velocities in the cavernous arteries, while patients with low-flow (ischemic) priapism will have no blood flow in the cavernous arteries. The return of the cavernous artery waveform will accompany successful detumescence [9,11,12]. Color duplex US of the penis should be performed before aspiration in ischemic priapism. After aspiration, a reactive hyperaemia may develop with a high arterial flow that may mislead the diagnosis as arterial priapism.

The cytological changes in the corpora cavernosa of the penis of patients due to low flow (ischemic) priapism and high-flow (non-ischemic/arterial) priapism has been studied in detail and are as follows [13]:

Low flow priapism

The first reaction of the tissue to the hemodynamic derangement is trabecular interstitial edema.

Then at the cellular level, trabecular smooth muscle cells were found to be the first affected by the hemodynamic derangement.

This reaction leads to structural & functional transformation of trabecular smooth muscle cells to fibroblast-like cells.

In low flow priapism, Intra cavernous clot formation &endothelial destruction occurs if more than 48 hours.

By this time trabecular inflammation becomes visible and most of the smooth muscle cells were either transformed to fibroblast-like cells or had undergone necrosis.

This type is more common and is an emergency because irreversible cellular damage and fibrosis can occur if treatment is not administered within 24 to 48 hours [14].

It results in long term squeal of erectile dysfunction or predisposition to frequent, prolonged episodes of priapism [15].

The causes of low-flow priapism are idiopathic, hematologic disorders, tumor infiltrate, or drug induced [6,8].

High-flow or arterial priapism

High flow priapism differs in that it results from increased arterial inflow into the cavernosal sinusoids, which overwhelms venous outflow and clinical presentation is usually painless and less rigid [6].

Irreversible cellular damage and fibrosis are rare in this variety [14].

This type of priapism is usually occurs following trauma that results in injury to the cavernosal or helician artery which establishes a fistula between the cavernosal artery and the corpus cavernosum and an unregulated inflow occurs [16], a fact supporting the view that high flow priapism is a more benign and prognostically more favorable form of priapism

Leukemic priapism is mostly low flow ischemic type and caused by hyper-leukocytosis. This may result from aggregation of leukemic cells in the corpora cavernosa and the dorsal veins of penis or from venous congestion of the corpora cavernosa due to mechanical pressure on the abdominal veins by the splenomegaly. Alternately infiltration of the sacral nerves with leukemic cells or infiltration of the central nerve system may be the cause [7,17].

Treatment

It is noted that there is no standard treatment proposed for leukemic priapism since the data is limited. The American Urological Association suggests that systemic treatment of the underlying disorder should not be the only treatment for ischemic priapism [18]. Priapism in CML is ischemic which a compartment syndrome is and the treatment must be focused at the penis. Various systemic therapies used in CML patients are cytoreductive therapy like Tyrosine kinase inhibitors (TKIs) and high-dose hydroxycarbamide, sometimes with the addition of leukapheresis to reduce hyperviscocity. Leukapheresis is a potent relatively safe procedure which is used for symptomatic alleviation in an acute setting. It is also noted that it is rarely used prophylactically or special situations like priapism, CNS involvement etc.

Initial penile intervention may utilize therapeutic aspiration with or without irrigation. If priapism persists even after aspiration/irrigation, intracavernous injection of sympathomimetic drugs should be performed. Therapeutic aspiration or intracavernous injection of sympathomimetics may be the primary intervention. In a trial two patients with ischemic priapism for ≥36 hours were successfully treated with high-dose intracavernosal phenylephrine (mean dose 45,000 μg) without any adverse event. Both patients did not develop any impotence [19]. It has been noted that the success rate with symphathomimetics agents is 43-81%. Various side effects noted with this treatment is acute hypertension, reflex bradycardia, tachycardia, headaches, palpitations and cardiac arrhythmia. Various surgical procedures have been tried only when the sympathomimetic agents have been failed [20]. The first-line choice of surgery procedure is the cavernoglanular shunt which can be done with a large biopsy needle (winter) or a scalpel (Ebbehoj) inserted percutaneously through the glans. The important side effect to worry about it is erectile dysfunction which is more common in the proximal shunts. Surgical shunting should be the last resort if detumescence is not achieved in 24-48 hours. The purpose of shunt is to establish a new venous outflow and restore normal arterial flow to the corpora cavernosa [6]. To attain resolution step by step management approach should be undertaken.

Conclusion

Priapism is a urological emergency and must be diagnosed and treated appropriately. Specifically in known cases of leukemia painful turgid penis should raise suspicion of priapism in first hand and penile Doppler should be the first line of imaging modality. This information should be forwarded to patient and emergency department clinician.

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