



## Oncology

## Priapism – A Rare Presentation in Chronic Myeloid Leukemia: Case Report



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## ABSTRACT

Priapism is a complication rarely seen in leukemia. We report a 19-year-old man presented with persistent painful erection of penis for over 24 hours at home. The patient had underwent immediate irrigation and decompression of priapism by urologist at emergency services of the hospital. This approach resulted in a flaccid penis later. During hospitalization, peripheral blood smear and bone marrow aspiration confirmed the diagnosis of chronic myeloid leukemia.

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## Introduction

Priapism is defined as full or partial erection that continues more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation.<sup>1</sup> Ischemic priapism is marked by rigidity of the corpora cavernosa with little or no cavernous arterial inflow. In ischemic priapism time dependent changes occur in the corporal metabolic environment with progressive hypoxia, hypercarbia, and acidosis. The patient typically complains of penile pain after 6–8 hours, and the examination reveals a rigid erection.<sup>1</sup> Ischemic priapism accounts for the majority of cases.

Although SCD is a predominant etiology of veno-occlusive priapism cases in the literature, there are a wide variety of other causes ranging from urinary retention to insect bites.<sup>1</sup> Hematologic dyscrasias are also a major risk factor for ischemic priapism. Priapism may occur in patients with excessive white blood cell counts. The incidence of priapism in adult male patients with leukemia is 1%–5%.<sup>2</sup> When priapism presents in the oncology setting, evaluation and management of the predisposing condition must accompany

interventions directed at the penis. We report a case of a young adult who presented with priapism as the first manifestation of chronic myeloid leukemia.

## Case report

A previously healthy 19-year-old man presented to the Urological emergency services with history of an erect penis of over 24 hours duration (Fig. 1). His penis was erect, painful and swollen when he arrived at emergency department. There was no history of trauma, malaise, night sweats, joint pain, and cough. The patient gave history of two similar previous episodes lasting for few hours and subsiding on its own in the past 1 month.

The physical examination revealed that the liver was palpable 1–2 cm below the right costal margin, and spleen was 3 cm below the left costal margin. The conjunctiva appeared pale with no jaundice. The penis was erect, firm and tender with superficial venous engorgement. Laboratory data showed hemoglobin (Hb) 9.2 g/dl, hematocrit 25.7%, white blood count (WBC), 296,800/mm<sup>3</sup>, and platelet 936,000/mm<sup>3</sup>. Serum chemistries were unremarkable. Treatment of the priapism was initiated by corporal aspiration and phenylephrine irrigation at the emergency services under the impression of low flow type priapism because of the history and physical examination. The erection gradually reduced following aspirations and washes.

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**Figure 1.** Clinical photograph of an erect penis.

For hyperleukocytosis, the patient was admitted to the inpatient services and was diagnosed as a case of chronic myeloid leukemia on the basis of peripheral blood smear and bone marrow examination (Fig. 2). The Philadelphia chromosome was illustrated in the patient. He was started on hydroxyurea tablets at 1.5 g per day and Imatinib 400 mg per day. Allopurinol 300 mg daily with adequate hydration was also started for potential tumor lysis syndrome.

## Discussion

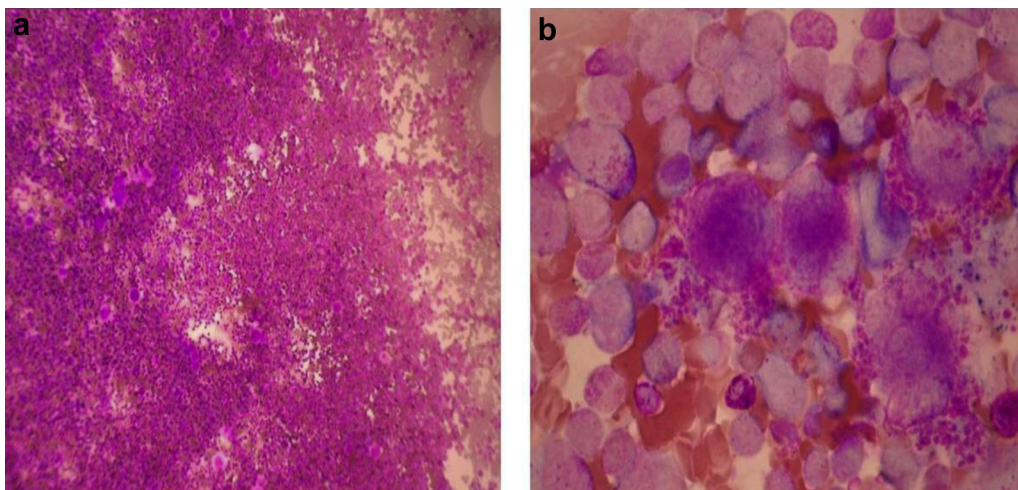
The condition priapism was named after the Greek god Priapus, thought to be the son of Zeus. It is believed that a jealous Hera or Aphrodite cast a spell over his mother while pregnant (either Aphrodite or Chloe) causing Priapus to be born with the affliction bearing his name and resulting in him being disowned by his mother.<sup>3</sup> Priapism is an urological emergency, which must be treated early to prevent erectile dysfunction. Priapism is a rare condition on its own with an incidence of 1.5 cases per 100,000 person-years. Hematological conditions are the cause of 20% of cases of priapism in men. Priapism secondary to penile metastasis is rare and one of the etiological factors.<sup>4</sup> Depending on the case series, priapism is seen in between 1 and 5% of male patients with all types of leukemia.<sup>2</sup> Of this small overall percentage, chronic myeloid leukemia (CML) accounts for 50% of all leukemic priapisms.

However, as a presenting feature of CML, priapism is rare in male patients, occurring in 1–2%. There is a bimodal age distribution in males of 5–10 and 20–50 years old, but it has been described in all age groups.<sup>1</sup>

There is little data available on the correct management of priapism in patients with CML; however the American Urological Association has published guidelines based on expert panel discussion and review of the limited data available.<sup>5</sup> American Urological Association strongly recommends that systemic treatment of an underlying disorder, such as CML, should not be undertaken as the only treatment for ischemic priapism.<sup>5</sup> Intra-cavernous treatment is required, and should be administered concurrently. As ischemic priapism is a compartment syndrome it requires treatment directed at the penis primarily. Systemic therapies that are commonly used in CML patients include cytoreductive therapy, such as high-dose hydroxycarbamide and tyrosine kinase inhibitors (TKIs), with or without the addition of leukapheresis to reduce hyperviscosity.<sup>3</sup>

Therapeutic aspiration (with or without irrigation) or intra-cavernous injection of sympathomimetics should be initially tried to treat priapism. A penile block may be performed but is not necessary. If performed, 10–20 ml of 1% lignocaine is injected below the symphysis pubis to block the dorsal nerves to the penis. A tourniquet is applied to the base of the penis. A 16 or 18 gauge biovalve intra-venous catheter can be inserted into the corpus cavernosum laterally through the penile skin, avoiding the ventral urethra and dorsal neurovascular bundle. Alternatively, the cannula may be inserted through the glans penis into the corpus cavernosum, which reduces skin bruising.<sup>3</sup> As the two corpora are interconnected, unilateral aspiration is sufficient. Twenty to thirty ml of blood is aspirated, and heparinized saline may be injected. This may be repeated a few times. Repeated aspirations over 1 hour may be needed and up to 50 ml of blood can be aspirated. On its own, aspiration has a success rate of approximately 30%.<sup>3</sup>

Our case is unique as priapism is a complication rarely seen in leukemia. The importance of prompt diagnosis and treatment of priapism cannot be overemphasized, as there is definite incidence of impotence following this condition. Besides the initial relief of priapism, the further workup and management of the underlying disease are more important. In our case, with use of a combined urological therapy and oncological treatment to priapism, the patient rapidly had relief of his clinical problem.



**Figure 2.** Bone marrow picture of chronic myeloid leukemia (H&E staining) – a) Under low power (10 $\times$ ), Hypercellular bone marrow. b) Under high power (100 $\times$ ), oil immersion – increased megakaryocytes with eosinophilic and basophilic myeloid precursors.

**Conflicting interest**

No.

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