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Priapism as A Warning Sign for Chronic Myeloid Leukemia

Boudreaux, J, BS, BA[1], Haller, CN, MD[2], Maslov, D, MD, MS[1,2], Bateman, L, MD[2]

[1]University of Queensland/Ochsner Clinical School, New Orleans, LA.
[2]Ochsner Clinic Foundation, Department of Internal Medicine, New Orleans, LA.

ABSTRACT

CML is a chronic myeloproliferative disorder associated with activating mutations in tyrosine kinases forming a BCR-ABL fusion gene, causing hyperproliferation of neoplastic myeloid progenitors [1,2]. The incidence of CML peaks in the fourth and fifth decades of life [3]. In the United States, nearly 4,500 cases are diagnosed annually [3]. The onset of the disease is insidious due to the nonspecific initial “B symptoms,” such as increased fatigability, weight loss and weakness. The natural history of CML is slow with a median survival of three years if untreated [3,4]. The current treatment is targeted therapies to the BCR-ABL tyrosine kinase using imatinib or nilotinib and hydroxyurea. Although B symptoms are common, priapism can be an initial presentation of CML in 1-5% of cases [5]. Priapism is a urological emergency defined by a sustained erection of four or more hours. Hyperviscous blood in leukemias, such as CML, prevents appropriate venous drainage of the corpora cavernosa, resulting in failure of detumescence [4]. This case reports highlights priapism as a presenting sign of CML and reviews the pathophysiology between the association.

Keywords: Chronic myeloid leukemia; Priapism

***Correspondence to Author:**
Boudreaux, J
University of Queensland/Ochsner Clinical School, New Orleans, LA.

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Case Report

A 52-year-old man presented to the Emergency Department due to an erection lasting for eight hours. The patient had another episode one week prior, which was treated at an outside hospital ED by penile draining. Relevant medical history includes a two-year history of thrombocytosis. The patient abuses cocaine, tobacco and alcohol and has an extensive psychiatric history, including diagnoses of anxiety, bipolar 1 disorder and schizoaffective disorder. The patient had previously been in a treatment facility for cocaine before presenting to hospital. The patient had been lost to follow up with Hematology for his thrombocytosis history. He was prescribed hydroxyurea but was noncompliant, as he believed the medications were causing his priapism. Positive genitourinary symptoms include penile pain, swelling and discharge. Patient denies dysuria, hematuria, scrotal swelling and testicular pain. Patient also reported easy bruising and bleeding. Other review of systems was negative.

Physical exam revealed an erect penis tender to palpation. Phenylephrine injection and penile aspiration were performed. The patient's priapism resolved and did not recur during his two-week admission.

During admission, laboratory findings included a white cell count of 20.81 K/μL with 8% basophils, red cell count of 4.08 M/μL, hemoglobin of 12.3 g/dL, platelets of 1,242K/μL. Bone marrow flow cytometric analysis showed polyclonal B and T lymphocytes of normal immunophenotype, granulocytes 88.3%, lymphocytes 2.1%, monocytes 0.5%, blast 5.9%, CD34 positive blasts and CD61 positive megakaryocytes. Blood test for *BCR/ABL* fusion p210 mutation was positive. An abdominal CT scan demonstrated splenomegaly, measuring 14.7 cm.

The patient received a therapeutic plateletpheresis, was prescribed imatinib and was discharged. The patient followed up in the Hematology clinic and started daily imatinib, which he tolerated well. After understanding the cause of his priapism, the patient remained compliant to both hydroxyurea and imatinib.

Discussion

Though rare, priapism can be an initial presentation of CML. There are only a handful of case reports supporting this.

First-line priapism treatment is intracavernosal injection of phenylephrine and aspiration of blood from the corpus cavernosum. Second-line therapies include penile decompression by creating a shunt between the corpus cavernosum and the glans penis or corpus spongiosum [4,5]. Additional supportive measures include oxygen supplementation, analgesia and intravenous fluids [4].

Possible etiologies of priapism include side-effects of medications, hematologic conditions, metabolic disorders, trauma, cancer, neurologic dysfunction and idiopathy. Some examples of hematologic disorders include thrombocytosis, sickle cell disease, leukemias, and myelomas. Examples of metabolic dysfunction leading to priapism include amyloidosis, diabetes and nephrotic syndrome. [4] Screening for antidepressants, illicit drug use and history of intracavernosal injections should be ordered [5]. A complete blood count with WBC differential can help in deducing etiologies due to blood dyscrasias or hemoglobinopathies [5].

Although CML may be attributed to this patient's priapism, it is also possible that cocaine abuse may have played a role. Further, in particular to this case, patient education played a large role in successful treatment. Once the patient better understood his diagnosis, he was compliant. This better communication between physician and patient may have saved his life.

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