CASE REPORT

Chronic myeloid leukemia presented with priapism: Effective management with prompt leukapheresis

H Ergenc, C Varım, C Karacaer, D Çekdemir[1]

Department of Internal Medicine, Faculty of Medicine, [1]Department of Hematology, Faculty of Medicine, University of Sakarya, Sakarya, Turkey

Abstract

Priapism is a painful medical condition in which the erect penis does not return to its flaccid state, despite the absence of both physical and psychological stimulation, within 4 h. Priapism is considered a medical emergency, which should receive proper treatment by a qualified medical practitioner. Treatment initially involves conservative measures, such as corporeal aspiration and irrigation with saline or dilute phenylephrine. If this fails, embolization or surgical shunting may be required. Priapism is more commonly associated with sickle cell hemoglobinopathy. However, hyperviscosity resulting from leukemia is a rare cause of priapism. We report a case of a 19-year-old man with an 18-h history of priapism secondary to undiagnosed chronic myeloid leukemia. We discuss the method of leukapheresis (mechanical white cell depletion) to reduce viscosity.

Key words: Chronic myeloid leukemia, leukapheresis, priapism

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Introduction

Priapism is an unprovoked painful prolonged erection of the penis. The unwanted, persistent erection is not caused by sexual stimulation or arousal, and priapism is usually painful. Priapism is an uncommon condition that needs immediate medical attention. Prompt treatment for priapism is usually needed to prevent tissue damage that could result in the inability to get or maintain an erection (erectile dysfunction). Other than the hemoglobinopathies such as sickle cell disorders and the thalassemias, hematological causes of priapism are rare indeed. Priapism has been seen in patients with either cellular (e.g., chronic myeloid leukemia [CML], essential thrombocythemia, polycythemia rubra vera) or plasma-related (e.g., multiple myeloma, Waldenstrom's macroglobulinemia) hyperviscosity. Hematological malignancy can cause low-flow veno-occlusive priapism secondary to hyperviscosity of circulating blood.[1] In patients with leukemia-induced priapism, the use of leukapheresis (mechanical white cell depletion) to rapidly reduce hyperviscosity in patients is well-documented.[2,3]

Address for correspondence:
Dr. C Varim,
Department of Internal Medicine, Faculty of Medicine, University of Sakarya, Sakarya, Turkey.
E-mail: ceyhunvarim@yahoo.com

This approach is usually associated with rapid resolution of symptoms. When such conservative approaches fail, aseptic aspiration of blood from the corpus cavernosa, followed by irrigation with saline and/or dilute phenylephrine, and surgical shunting, embolization or radiation therapy. The following case further illustrates the value of prompt leukapheresis in the treatment of priapism related to leukemia.

Case Report

We present here is an 18-year-old male, nonsmoker who presented to the medical emergency with sudden onset painful penile erection and scrotal swelling, which had persisted for the last 72 h [Figure 1]. There was no history of sexual stimulation, trauma, previous similar episodes, use of medications or any chronic illness. On physical examination, the patient had pallor. The spleen and liver

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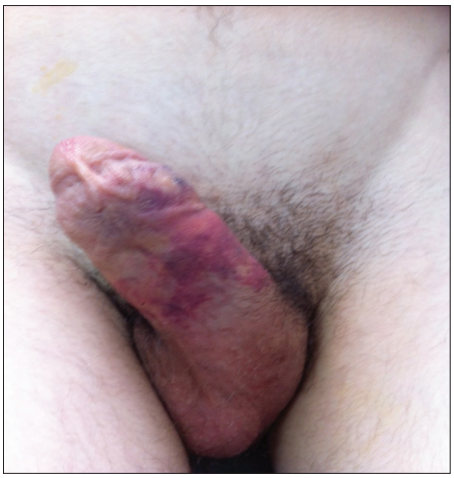

Figure 1: Priapism: Erect and painful penis with scrotal swelling

were 2–3 cm. palpable under arcus costa. Penis was erect, firm, and tender with prominent superficial veins. Urinalysis was normal. Other investigations, including liver functions, kidney functions, chest X-ray, and electrocardiography were unremarkable.

Hemoglobin concentration was 6 g/dL, white blood cell (WBC) count was 100,000/mm^3, platelet counts were 1,002,000/mm, lactate dehydrogenase 1009 IU/L (125–220 IU/L). A peripheral blood smear demonstrated immature leukocytes in various stages of differentiation (neutrophil 45%, metamyelocyte 40%, lymphocyte 6%, monocyte 4%, blast 3%, promyelocyte 1% and myelocyte 1%). Bone marrow analysis was suggestive of CML [Figure 2]. The bone marrow smear demonstrates hypercellularity with myeloid hyperplasia, no increase in blasts. Cytogenetic analysis showed found 45–46; XY, t (9;22) (q34;q11) in the genetic laboratory.

The BCR-ABL translocation was positive, and leukocyte alkaline phosphatase score (normal 30–140%) was decreased to 20%, which confirmed our suspicion of CML as the underlying etiology of priapism.

İmatinib 400 mg once daily along with allopurinol 300 mg once daily and intravenous fluids to maintain hydration were started. Although we started the medication immediately, WBCs count was still 406,000/mm^3 and painful penile erection continued 3 days. Leukapheresis was done immediately because of the high risk of erectile disfunction began. Leukapheresis took 3 h. Patient lied on a bed or reclining chair, with a tube into a vein in each arm. One tube removed blood and passed it into a machine that removed WBCs, including the leukemia cells. The rest of his blood cells and normal blood fluid (plasma) went back into his body through the tube in his other arm. After leukapheresis the WBC count was 40.000/mm^3 The patient responded to the treatment and were able to achieve an erection with

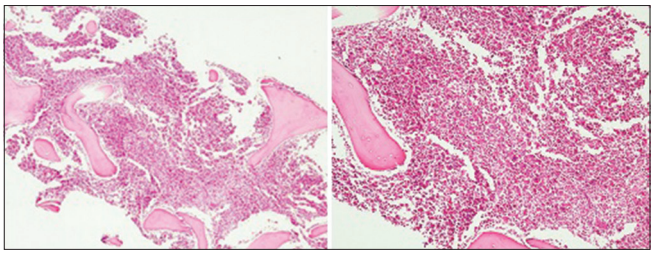

Figure 2: The bone marrow smear demonstrates hypercellularity with increase megakaryocytes and neutrophil (H and E, ×40, ×100)

manual stimulation and maintains the ability to ejaculate. At present, the leukemia is in remission for 6 months.

Discussion

Priapism is a sustained penile erection in the absence of sexual activity. Priapism is generally defined as an unwanted erection lasting more than 4 h.[1]

Priapism can occur at any age, and two peaks in age distribution are described. A pediatric peak, 5–10 years old, is noted owing to sickle cell disease (SCD) in black patients. One of the common manifestation that can significantly affect quality of life of men with SCD, which is the development of prolonged, painful erections known as low-flow or ischemic priapism that result in tissue ischemia and attenuated or absent return of functional erections. Reports of the lifetime prevalence of ischemic priapism in SCD range from 2% to 35%, respectively. High flow priapism, which results from increased arterial flow generally caused by trauma, does not result in tissue ischemia and has not been associated with increased risk in SCD patients.

The secondary peak occurs in patients with active sexual activity age of 20–50 years old. Idiopathic priapism is the most common (64%) while approximately 20% are related to hematologic disorders. In CML, priapism is an unusual presentation and seldom to encounter. An important aspect of priapism is that most physicians will never encounter. The poor experience will result in a delay of treatment and irreversible squeal. Hence, all physicians should understand that long-term sequela can be avoided with prompt diagnosis and treatment. Priapism can occur at any age, and two peaks in age distribution are described. A pediatric peak, 5–10 years old, is noted owing to SCD in black patients. The secondary peak occurs in patients with active sexual activity age of 20–50 years old. Idiopathic priapism is the most common (64%) while approximately 20% are related to hematologic disorders.

Prompt recognition and appropriate treatment of an episode of priapism in patients with CML are critical. This is the result prolonged, or repeated episodes of priapism can result ischemia and fibrosis of corpus cavernosa of the

penis, potentially leading to impaired sexual function and impotence. The goal of management for stuttering priapism is prevention of future episodes.[1] A practical approach to the diagnosis and management of priapism in patients with CML will be presented here. Priapism is an unusual and rarely presentation for CML. Hyperleukocytosis is thought to be the cause of priapism in patients with leukemia. Three different mechanism have been described: These are venous congestion of the corpora cavernosa resulting from mechanical pressure on the abdominal veins by the splenomegaly; Sludging of leukemic cells in the corpora cavernosa and the dorsal veins of penis and infiltration of the sacral nerves and central nervous system with leukemic cells.[4,5] In our case, significant leukocytosis with hepatosplenomegaly supports the first mechanism in the pathogenesis. Leukostasis is a medical emergency most commonly seen in patients with CML in blast crisis.[6] It is characterized by an extremely elevated blast cell count and symptoms of decreased tissue perfusion. An important aspect of priapism is that most physicians will never encounter it. This limited or no experience can contribute to a delay in diagnosis and treatment, and irreversible squeal. So it is crucial for physicians to understand that long-term sequela can be avoided with prompt diagnosis and treatment. Priapism should be treated promptly to avoid long-term complications. Although hematological causes are rare, they can easily be excluded with a full blood count and peripheral blood smear. In addition, the rapid detumescence can be achieved with leukapheresis and should be a priority in the management of leukemia induced priapism where the resources and expertise for leukapheresis are available. If leukapheresis fails, transglandular cavernosumspongiosum surgical shunting or radiation therapy should be considered.

References

1. Keoghane SR, Sullivan ME, Miller MA. The aetiology, pathogenesis and management of priapism. BJU Int 2002;90:149-54.
2. Agrawal DK, Jha S, Verma A, Tripathi AK, Singh BN. Priapism, complicating chronic myeloid leukemia – A case report. Indian J Cancer 1991;28:51-2.
3. Becker HC, Pralle H, Weidner W. Therapy of priapism in high counting myeloid leukemia – A combined oncological-urological approach. Two case reports. Urol Int 1985;40:284-6.
4. Savona M, Talpaz M. Chronic myeloid leukemia: Changing the treatment paradigms. Oncology (Williston Park) 2006;20:707-11.
5. Ponniah A, Brown CT, Taylor P. Priapism secondary to leukemia: Effective management with prompt leukapheresis. Int J Urol 2004;11:809-10.
6. Cuttner J, Conjalka MS, Reilly M, Goldberg J, Reisman A, Meyer RJ, *et al.* Association of monocytic leukemia in patients with extreme leukocytosis. Am J Med 1980;69:555-8.

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