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Case Report
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Priapism: A Rare Presentation of CML

Rateesh Sareen[1], Menka Kapil[1] and Malpani BK[2]*

[1]*Department of Pathology & Transfusion Medicine, Santokba Durlabhji Memorial Hospital & Research Center, Jaipur.302015, India*

[2]*Department of Medicine, Santokba Durlabhji Memorial Hospital & Research Center, Jaipur.302015, India*

Abstract

We report a rare case of 17 years male presenting with priapism in medical emergency. The patient underwent immediate treatment and simultaneous investigations revealed a diagnosis of CML. Hyper leucocytosis causes low volume ischemic priapism requiring multidisciplinary approach-urologist, physician, oncologist and pathologist working at tandem to clinch the diagnosis. The unusual presentation of CML needs to be borne in mind while dealing with cases of priapism having splenomegaly.

Keywords: CML; Priapism; Hyperleukocytosis

Introduction

Priapism is persistent unprovoked abnormal full or partial erection of penis that continues for more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation [1]. Idiopathic Priapism is most common and may be due to thrombosis occurring in the venous plexus [2]. The association of Priapism is approximately 20% with hematological disorders [3,4]. Priapism as presenting complaint in chronic myelogenous leukemia (CML) is exceedingly rare finding. We report a case of 17 years boy presenting with Priapism in emergency and after preliminary treatment was diagnosed as CML.

Case Presentation

We report a case of 17 year male presenting in a tertiary care hospital with recent onset of priapism unrelated to sexual stimulation or drugs.

The general examination revealed hepatosplenomegaly.

The laboratory investigations revealed-BUN- 15mg/dl, Creatinine -0.8mg/dl, Sodium 139m mol/litre, potassium 4.0m mol/liter, chloride 98m mol/lit, glucose 100mg/dl, SGOT- 42U/L,SGPT 62U/L, total Bilirubin 0.6mg/dl, Direct Bilirubin-0.2mg/dl, Total protein-8.0gm/dl, Albumin 4.9gm/dl, globulin 3.1gm/dl , A/G ratio 1.58, Alkaline Phoshatase 102U/L, Gamma GT-102U/L, bleeding time was 2 minutes, clotting time—5 minutes and PT13.6. An automated complete blood count (CBC) demonstrated Hemoglobin-113g/L (reference range 130-170 g/L), white blood cell count 377.31 x 10^9/L (reference range 4-10 x 10^9/L) Platelet count 730 x 10^9/L (reference range 150-450 x 10^9/L) , Hematocrit 32.4% (reference range-36%-46%), differential count – Blasts+ Promyelocytes-12%, Myelocytes-15%, Neutrophils- 50%, Band forms-12%, Lymphocytes-03%, and Eosinophils-6% (Figure 1). A concurrent peripheral blood smear showed normocytic normochromic red blood cells. Test for HIV 1 & 2, Hepatitis B and C viral serology were nonreactive. Malarial smears and rapid malarial antigen test were negative. Routine urine examination did not detect any abnormality. Bone marrow examination and Bcr –ABL testing was advised. Penile Doppler ultrasonography showed low volume stasis as the cause of persistent priapism.

Aspiration was attempted following intra-cavernosal injection of phenylephrine. On repeat aspiration volume of 100ml of blood was aspirated and patient's priapism subsequently resolved. No surgery was performed. The patient after getting relieved from tumescence wanted to seek advice and treatment from cancer hospital and so was discharged on request. The latter follow up with patient confirmed bcr-abl positive CML as diagnosis.

Discussion

CML has hyperleucocytosis and it is not unusual for Priapism to occur but as the treatment requires a multidisciplinary approach of urologist, oncologist and pathologist with lack of awareness

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**Correspondence:*
Rateesh Sareen, Department of Pathology & Transfusion Medicine, Santokba Durlabhji Memorial Hospital & Research Center, Jaipur.302015, India.
E-mail: drrateeshsareen@yahoo.co.in

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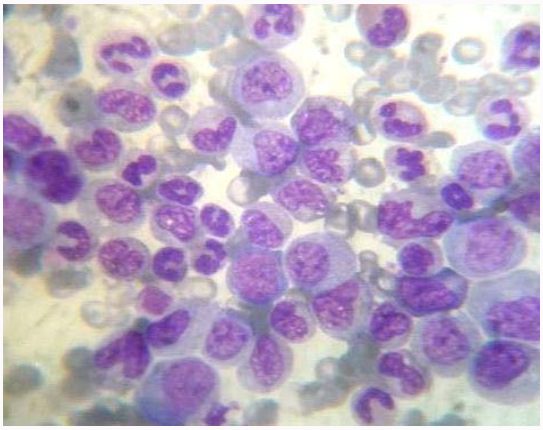

Figure 1: Peripheral Blood Smear (Leishman stain, 100x).

on the part of physicians in patients with priapism and splenomegaly mandates attention to this unusual presentation.

Priapism can occur at any age and two peaks of age are noticed [5], the first one in the age group of 5-10 years in children with Sickle cell disease (SCD) and the second one in patients of 20-50 years of age with active sexual activity [6]. In children SCD accounts for 67% of cases of priapism while in adults leukemia accounts for 15% of the total cases [7]. Again in adults with leukemia, 50% of cases are due to CML [8,9]. It is rare to encounter priapism in CML and that too as an initial presentation is exceedingly rare phenomena [10]. Priapism can be of low flow type and high flow priapism. Low flow or ischemic priapism results from decreased penile venous outflow that causes stasis resulting in tissue ischemia and attenuated functional erection. It manifests as painful, rigid erection and presents as an acute emergency because irreversible cellular damage and fibrosis occur if treatment is not administered within 24-48 hours [11]. The most common causes of low flow type are idiopathic, hematological disorders, tumor infiltration or drug induced [12-14]. On the other hand high flow or arterial priapism results from increased arterial inflow into cavernosal sinusoids which is in excess of venous outflow and present in a painless manner, associated with trauma to penis or perineum resulting in injury to the internal pudendal artery [15,16]. It is not an actual emergency in patients with high flow priapism and treatment can be on elective basis.

One of the concerns with respect to priapism is that most physicians are aware of long term sequel so that it can be avoided with prompt diagnosis and treatment. The prolonged repetitive episodes of priapism result in ischemia and fibrosis of corpus cavernosa of penis, potentially leading to impaired sexual function and impotence. Hyperleukocytosis is thought to be the cause of priapism in patients with leukemia. Different mechanisms [15,16] proposed are as follows:

1. Venous congestion of corpora cavernosa due to mechanical pressure on the abdominal veins by splenomegaly.

2. Slugging of leukemic cells in corpora cavernosa and dorsal vein of penis.

3. Infiltration of sacral nerves with leukemic cells.

4. Infiltration of central nervous system with leukemic cells.

5. Hyper leukocytosis causing elevation of whole blood viscosity causing vascular occlusion.

In our case significant leucocytosis with hepatosplenomegaly attributed to priapism.

The differentiation of low and high flow priapism can be done with detailed history, examination and blood gas analysis of blood within corpora cavernosa as well as penile Doppler study [17]. The clinical history encompasses family history of SCD, current medications, trauma, malignant disease and the use of intracavernosal agents. In low flow priapism the penile shaft is rigid & painful with soft glans whereas in high flow priapism the entire penis is rigid & painless. On intracavernosal blood gas analysis, low flow priapism has ph<7.00, pCo_2>60mm of Hg and pO_2<30mm of Hg [18]. The high flow priapism have normal intracavernosal blood ABG values. The outer preliminary investigations include complete blood count, hemoglobin electrophoresis, coagulopathy state, serum chemistry and drugs. Priapism is an exceptional presentation in male patients with CML [19]. There is studies in literature where diagnosis of CML was made retrospectively after obtaining results of laboratory investigations [20]. As in our case, hyperleukocytosis caused stasis and possibly microthrombi in cavernosal circulation which precipitated priapism[21].Hyperleukocytosis is the presence of blast cell count or WBC exceeding 50 to 100 x 10^9/L. It leads to leukostasis that is the cause of hearing loss, papilledema, cerebellar dysfunction memory function loss, acute renal failure, respiratory depression and intracranial hemorrhage [21,22].

Chisick et al [23] have mentioned about initial management of priapism in CML patients which is quite useful in resource limited developing countries like India. The treatment algorithm stress upon the utility of peripheral blood film examination, for patients having priapism & splenomegaly [24]. Penile blood testing and penile Doppler Ultrasonography are not considered essential in CML causing priapism as it is already known to be ischemic type. These interventions would unnecessary delay the emergency treatment[24]. Therapeutic aspiration with heparinized saline irrigation was performed within four hours. The procedure was repeated twice to bring immediate relief and rapid detumescence. In case of persistent erection for 24-48 hours surgical shunt should be performed. The involvement of urologist in early intervention is beneficial. The other therapies mentioned in literature include-cyto reduction with chemotherapeutic agents, leukapharesis, allopurinol hydration and tyrosinekinase inhibitor therapy [5,15].

Our case is rare in the sense that the presentation itself is of priapism. The importance of prompt diagnosis and treatment of priapism cannot be over emphasized as there is definite incidence of impotence after this condition. The patient was promptly relieved of his clinical problem; diagnosis of CML was made but as the patient opted for taking treatment at a dedicated cancer hospital he was advised to go. On a month follow up the patient reported regression of blood counts to normal and no incidence of priapism.

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