

ABSTRACT

Introduction :

Chronic Myeloid Leukemia (CML) is a chronic myeloproliferative disorder and a form of leukemia characterized by the increased and unregulated growth of myeloid cells in the bone marrow and the accumulation of these cells in the blood. Majority of undiagnosed patients may initially present to primary health cares and emergency departments with classical symptoms of anemia, unresolved infections and bleeding tendencies. Priapism is a rare symptom, about 5% of leukemic patients, and is most likely caused by venous obstruction from microemboli/thrombi and hyperviscosity caused by the increased number of circulating leukocytes.

Case Report :

A 26-year-old married male who presented with priapism to Emergency Department of Hospital Sultanah Nur Zahirah and was eventually diagnosed as CML.

Discussion :

Priapism is an andrological emergency, as the risk of impotence is 50% despite appropriate management. Priapism can either be idiopathic or secondary to several medical conditions including CML. Urgent Full Blood Count (FBC) and peripheral blood smear are the first diagnostic tools which can be obtained at emergency departments to help narrow down the cause of priapism which demand early intervention and treatment. Prompt urological referral and cavernosal aspiration is important for symptomatic relief and resolution of the obstructed blood flow.

Conclusion :

In general, as priapism is one of the rare manifestations of CML, high index of suspicion is vital once encountered and urgent FBC and peripheral blood smear are the first diagnostic tools in emergency care settings and should be performed immediately to prevent delayed diagnosis and management

INTRODUCTION

Priapism is both medical and surgical emergency that is characterized by prolonged and painful erection of the penis unassociated with any sexual arousal or desire(1). It is a debut appearance as a sign of hematological dyscrasia and a rare event. Approximately 50% of patients having priapism are at risk of impotence, despite appropriate therapy(1). If untreated, priapism may lead to penile necrosis and permanent erectile dysfunction. Early intervention is the key to reverse the anoxic metabolic derangements caused by the priapism. Priapism resolution following aspiration with or without irrigation is ~30%. Priapism can either be idiopathic or secondary to several medical conditions including Chronic Myeloid Leukemia (CML). The diagnosis of priapism as manifestation of CML will usually be suspected from the Full Blood Count (FBC) and peripheral blood smear. Hence, we would like to highlight a case of priapism seen at emergency department as first presentation of CML.

CASE REPORT

History Taking

- A 26-year-old married Malay man with no known previous illness presented to Emergency Department of Hospital Sultanah Nur Zahirah complaining of prolonged and painful penile erection.
- There is no history of trauma, mechanical stimulation, taking any erectile/traditional medications or undergo any radiation therapy prior to the symptom.
- He denied history of fever, night sweats, pus discharge or penile bleeding previously.

Physical Examination

General Examination	Mild pallor with good perfusion Vital signs unremarkable
Systemic Abdominal Examination	Enlarged spleen crossing the midline, no hepatomegaly or abdominal tenderness
Local Genitalia Examination	Erected and hardened corpus cavernosum, tense penile shaft, and superficial venous engorgement
Other Systemic Examinations	No significant abnormality or lymphadenopathy

Blood Investigations

Full Blood Count	
Total White Count (10 ⁹ /L)	270
Hemoglobin (g/L)	9
Platelet (10 ⁹ /L)	329

Table 1. Full Blood Count (FBC).

Full Blood Picture

Leucocyte	Hyperleukocytosis : granulocytic series seen at all stages of maturation, basophilia eosinophilia Differential counts : 5% blast cells, 3.5% promyelocytes, 14% myelocytes, 48% neutrophils, 2.5% metamyelocytes, 10% eosinophils, 16% basophils, 1% lymphocytes considering chronic myeloid leukemia in chronic phase
Erythrocyte	Normochromic normocytic anemia Nucleated red blood cells
Platelet	Large platelet with normal counts

Table 2. Full Blood Picture (FBP).

Molecular Study for BCR-ABL (PCR)

BCR-ABL fusion transcript detected in peripheral blood at 0.0693

Table 3. Molecular Study (PCR).

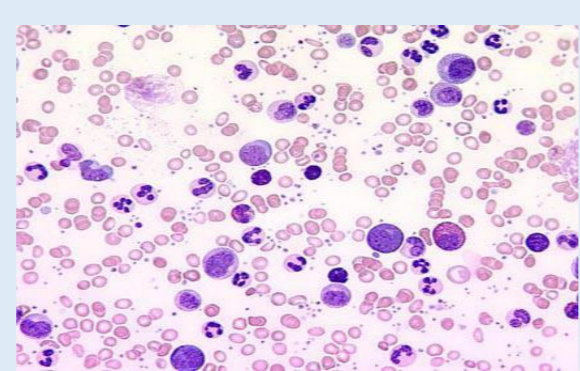


Figure 1. Blood film at 400X magnification demonstrates leukocytosis with the presence of precursor cells of the myeloid lineage. In addition, basophilia, eosinophilia, and thrombocytosis can be seen.

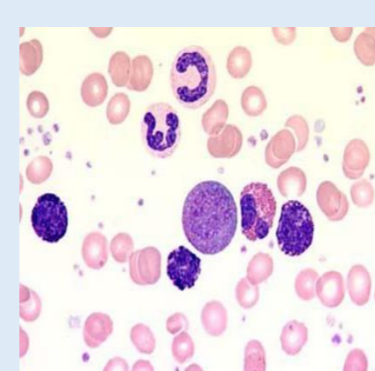


Figure 2. Blood film at 1000X magnification shows a promyelocyte, an eosinophil, and 3 basophils.

CASE REPORT (CONT)

Procedure

- Urgent referral to surgical department was done then patient underwent cavernosal aspiration, and approximately 60cc of blood evacuated.
- The pain was much improved following the analgesic and procedure, unfortunately, there was incomplete relief of the erection by the aspiration

Referral

- The patient was immediately referred to Urology Center in Hospital Raja Perempuan Zainab, Kota Bharu.
- Distal Corpus Spongiosum Shunt was done.
- Peripheral blood smear was sent for BCR-ABL translocation and turned out to be positive and these findings confirmed the underlying hematological malignancy to be Chronic Myeloid Leukemia (CML) and he was started on tyrosine – kinase inhibitor, Imatinib 400mg OD then total white count started falling to normal ranges.
- On subsequent follow up, his erectile function had improved and he was later referred to Urology Center in Hospital Kuala Lumpur and suggested for penile implant/prosthesis if developed irreversible penile damage.

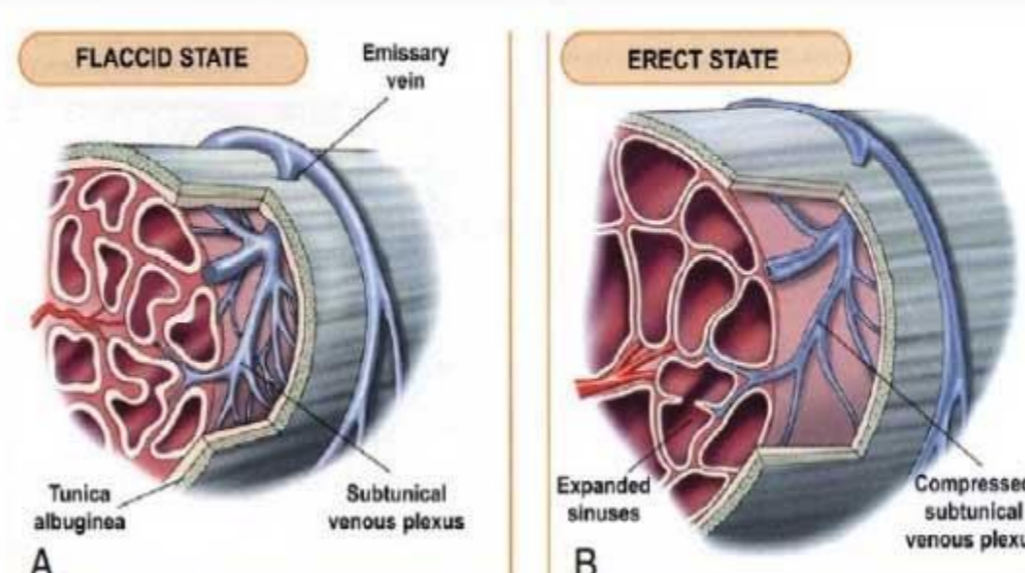


Figure 3. Corporeal relaxation causes external pressure on the emissary veins exiting the tunica albuginea, trapping blood in the penis and causing erection.

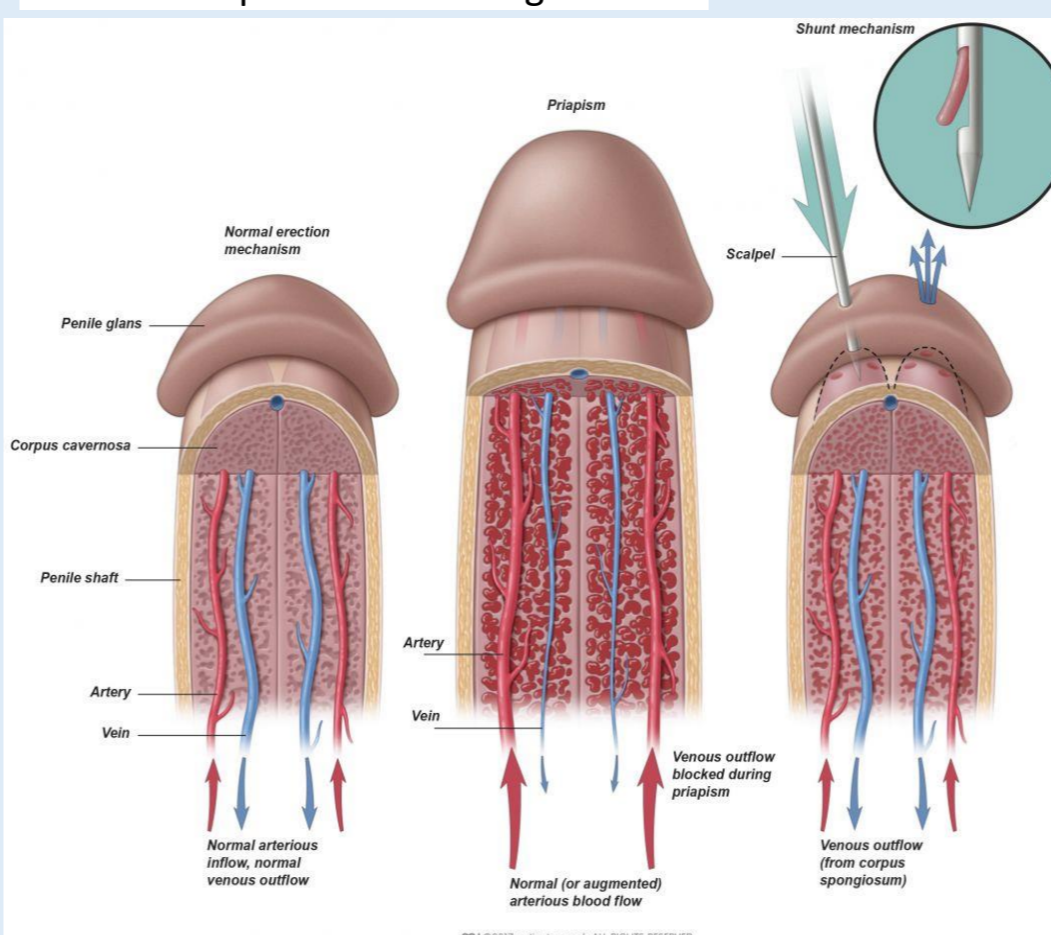


Figure 4. Penile shunt surgery is performed to restore an exit for blood and to re-establish blood circulation within the penis. A connection ("shunt") is created between the corpora cavernosa and the glans of the penis.

DISCUSSION

Chronic Myeloid Leukemia (CML) is a hematopoietic stem cells disease which is characterized by reciprocal translocation between chromosomes 9 and 22, resulting in the formation of the Philadelphia chromosome (Ph chromosome) with deregulated tyrosine kinase activity. These effects increase proliferation, affect differentiation, and block apoptosis.

Approximately 20% of CML cases were diagnosed incidentally. The most common clinical manifestations of CML are nonspecific symptoms which includes anorexia, malaise, weight loss, sweating, bleeding episodes due to platelet dysfunction and abdominal discomfort due to enlargement of the spleen(1). Rarely, patients may present with leukostatic complications of hyper leukocytosis such as thromboembolic phenomena, hearing loss or priapism

Priapism is a pathological condition of persistent penile erection in the absence of sexual excitation. The incidence of priapism is 1.5 per 100,000 people(1). It can either be idiopathic or secondary to several medical conditions. About 20% of cases of priapism are caused by hematological disorders. In patients with leukemia, 50% of cases of priapism are due to CML and priapism is less common in patients with acute leukemia than in patients with chronic leukemia. The incidence of priapism in adult leukemia ranges between 1 and 5%.

DISCUSSION (CONT)

The main pathophysiology behind chronic myeloid leukemia is hyperleukocytosis which results in hyperviscosity that leads to priapism. This condition caused leukostasis and possibly microthrombi in the cavernosal circulation which precipitated priapism.

The following five pathophysiological mechanisms of priapism in CML have been postulated (1):

- venous congestion of the corpora cavernosa resulting from mechanical pressure exerted by the enlarged spleen on the abdominal veins
- sludging of leukemia cells in the corpora cavernosa and the dorsal veins of the penis
- local infiltration of sacral nerves by leukemia cells
- central nervous system infiltration by leukemia
- hyperleukocytosis causing elevation of whole-blood viscosity and this ultimately causing complication due to vascular obstruction.

In a patient presenting with priapism, the following are essential to make diagnosis of the cause of presentation: detailed history, thorough physical examination, appropriate laboratory investigations including Full Blood Count (FBC), differential cell count and peripheral blood smear. In emergency department, FBC, differential cell count and peripheral blood smear can help as the first diagnostic tools in narrowing down the diagnosis of suspected hematological malignancy particularly CML. Characteristic FBC features for diagnosing CML are as follows: absolute leukocytosis (median of 100,000/ μ L) with a left shift and classic "myelocyte bulge" (more myelocytes than the more mature metamyelocytes seen on the blood smear); blasts usually number <2%; absolute basophilia is nearly universal, with absolute eosinophilia in 90% of cases; monocytosis is often seen, but generally not an increased monocyte percentage; absolute monocytosis is more prominent. Platelet count is usually normal or elevated. At times, the diagnosis of CML can only be made retrospectively after obtaining results of laboratory investigations.

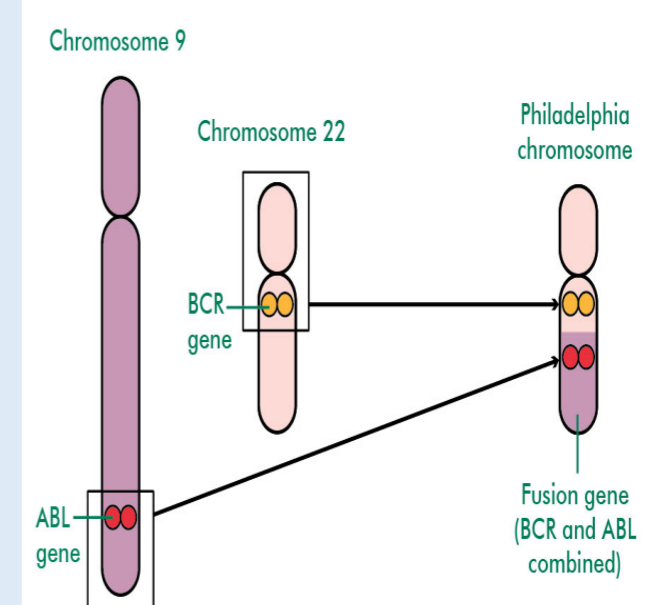


Figure 5. This diagram shows how the Philadelphia chromosome is made when a section of chromosome 9 joins on to a section of chromosome 22 which form the Philadelphia Chromosome (fusion gene of BCR-ABL)

CONCLUSIONS

Priapism is an andrological emergency and is one of the rare manifestations of CML as a result of hyper viscosity caused by hyperleukocytosis. Thus, it is essential for physicians to have a high index of suspicion regarding the possible causes of priapism once encountered since it demands early intervention and treatment. Urgent FBC and peripheral blood smear are the first diagnostic tools in emergency care settings and should be performed immediately to prevent delayed diagnosis and management.

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