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Acute Priapism as a First Manifestation of Chronic Phase CML; A Case Report

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Abstract

Priapism is a urological emergency that, if not treated promptly, can result in erectile dysfunction. 50% of cases of priapism in leukemia are caused by chronic myeloid leukemia, but priapism is only observed in 3% of chronic myeloid leukemia and very infrequently as the initial manifestation. This is a case of a 28-year-old male with no prior co-morbid presented with acute penile erection for eighteen hours. On examination, the patient was vitally stable and found to have a palpable spleen of 4cm below the left costal margin. Laboratory workup showed leukocytosis, low hemoglobin, normal platelets, and 2% myeloblasts on the peripheral film. Aspiration of the corpora cavernosa at the shaft of the penis was done and blood gas analysis showed acidosis, hypercarbia, and hypoxia consistent with ischemic priapism. Bone marrow aspirate showed chronic phase chronic myeloid leukemia. BCR-ABL by FISH showed p210 mRNA and the patient was started on Imatinib. Priapism in chronic myeloid leukemia is very rare as the first presentation and requires urgent initiation of cytoreduction therapy to prevent worse outcomes.

Keywords: Priapism, Imatinib Mesylate, Chronic Myelogenous Leukemia, BCR-ABL Positive, Cytoreduction Surgical procedures

Introduction

Priapism is a urological emergency that is defined as persistent penile erection for more than 4 hours irrespective of the stimulus.[1] This condition is considered an emergency due to the risk of fibrosis and erectile dysfunction. Priapism is divided into two types: ischemic and non-ischemic.[2] Hematological disorders are found in around 20% of cases of priapism. Chronic myeloid leukemia accounts for 50% of cases of priapism in leukemia. Chronic myeloid leukemia usually presents with lymphadenopathy and splenomegaly and/or hepatomegaly but priapism is seen in ≤ 3%.[3] It has been seen that chronic myeloid leukemia causes priapism by cellular hyper viscosity leading to venous occlusion.[4] We report a 28-year-old male who presented in the urology emergency department with priapism as the first presentation of CML.

Case Report

A 28-year-old male, with no known comorbid, presented in the emergency department with involuntary, prolonged, painful penile erection for 18 hours. There was a history of intermittent episodes of protracted penile erection without any stimulus for the last 10 months along with undocumented weight loss over the previous 8 months. There was no history of night sweats, bone pains, trauma, or any medications.

On examination, pulse was 94 beats per minute, blood pressure was 112/70 mmHg. Abdominal examination revealed the spleen 4cm below the left costal margin, while lymphadenopathy was not present. Local examination revealed an erect, swollen, engorged, and tender penis with engorged veins on its surface. At the presentation to the emergency department, his Hb was 8.5, WBC 439.5, and platelets 276.

The patient was treated with aspiration of the corpora cavernosa at the shaft of the penis which resulted in a drop of Hb from 8.5 to 6.6 as mentioned in Table 1. About 250 ml of dark blood was aspirated and the priapism was relieved. Blood gas analysis of the aspirated blood showed pH 6.6, pCO2 117.6 mmHg, and pO2.[9]

According to Table I, there was anemia (HB 6.6g/dL), leukocytosis (WBC 285,000 cells/mm3), with left shift (neutrophils 4.5% and lymphocytes 2.9%), presence of immature cells (myeloblasts, myelocytes, and metamyelocytes), increased ESR (35mm/hr) and increased LDH (752 U/L). The values of the complete laboratory workup are mentioned in Table I.

The peripheral film revealed normochromic normocytic, nucleated RBCs, polymorphic neutrophilic leukocytosis with left shift until the level of blasts (2%) noted, bimodal

peak at neutrophils and myelocyte, eosinophilia, platelet anisocytosis and thrombocytosis seen. The peripheral film was in line with chronic phase CML as shown in Figure 1. Final diagnosis was made on bone marrow aspiration and biopsy followed by detection of BCR-ABL gene by FISH.

Table I: Laboratory Test Results for Patient.

Test	Result	Normal Ranges
Haemoglobin (HB)	6.6 g/dL	(12.3-16.6)
Mean Corpuscular Volume (MCV)	92fL	(80-100)
White Blood Cells (WBC)	285,000 cell/mm3	(4.8-11.3)
Neutrophils	4.5%	(34.9-76.2)
Lymphocytes	2.9%	(17.5-45)
Eosinophils	4%	(0.3-7.4)
Basophils	3.4%	(0-1)
Monocytes	3%	(3.9-10)
Myeloblasts	2%	0
Myelocytes	30%	0
Metamyelocytes	21%	0
Platelets	216,000 cells/mm3	(154-433)
Erythrocyte Sedimentation Rate (ESR)	35mm/hr	(0-15)
Blood Urea Nitrogen (BUN)	23mg/dL	(7-20)
Serum Creatinine	0.81 mg/dL	(0.7-1.3)
Potassium	4.4 mEq/L	(3.5-5.2)
Uric Acid	4.05 mg/dL	(3.5-7.2)
Lactate Dehydrogenase (LDH)	752 units/L	(140-280)
Total Bilirubin	0.8mg/dL	(0-0.8)
ALT	28 mg/dL	(10-130)
AST	41 mg/L	(10-34)
ALP	109 mg/dL	(24-147)
PH	6.60	(7.35-7.45)
pCO2	117.6mmHg	(35-45)
pO2	9mmHg	(75-100)

Key: Blue = lower than normal value
Red= higher than normal value

The patient was started on intravenous 0.9% normal saline at 100 mL/Hour, oral hydroxyurea 2000 mg once daily for cytoreduction while awaiting BCR ABL results, along with paracetamol 500 mg thrice daily and oral allopurinol 300mg once daily for covering tumor lysis syndrome.

BCR ABL by FISH was positive so subsequent Quantitative BCR-ABL was sent showing p210 mRNA transcripts and estimated to represent 8.97% of total ABL1. The patient was started on Imatinib (Tyrosine kinase inhibitor) for treatment of the chronic phase of chronic myeloid leukemia.

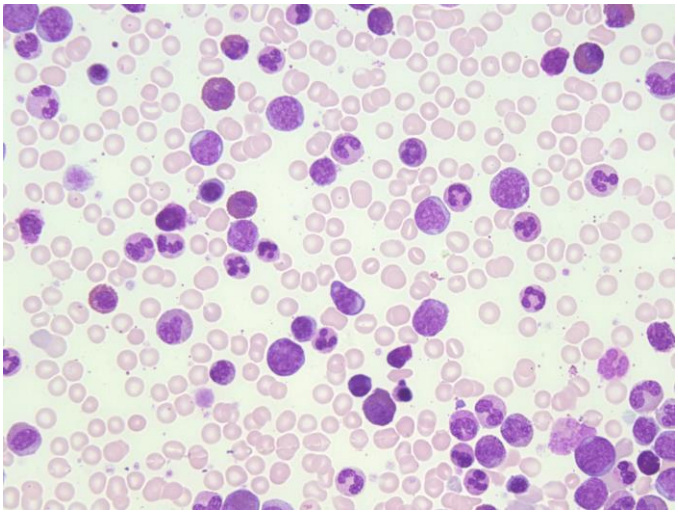

Figure 1. Chronic Phase of Chronic Myeloid Leukemia.

The peripheral film showing normochromic normocytic, nucleated RBCs, polymorphic neutrophilic leukocytosis with left shift until the level of blasts (2%) noted, bimodal peak at neutrophils and myelocyte, eosinophilia, platelet anisocytosis and thrombocytosis seen.

Discussion

Priapism is defined as sustained penile erection, irrespective of stimuli, for more than 4 hours. It is considered a urological emergency as it can ultimately lead to erectile dysfunction.[1]

Priapism is seen to have two peaks in age distribution. One in pediatric peak, mostly 5-10 years old, majority of cases due to sickle cell disease. The second peak is seen in the age group of 20-50 years old. Idiopathic priapism contributes around 64% to this group. While hematological malignancy contributes to 20% of cases. Fifty percent of priapism cases in leukemia are contributed by chronic myeloid leukemia.[3,4]

Priapism is divided into ischemic and non-ischemic types. Ischemic priapism is due to venous stasis and is called “low flow”, while non-ischemic priapism is a disorder of arterial flow and is called “high flow”. They can be usually differentiated based on history and examination but relevant lab workup is also necessary to confirm the diagnosis. In ischemic priapism blood gas analysis will show a pH of <7.25, pO2 of <30mmHg, and pCO2 of >60mmHg, while in non-ischemic priapism it will show a pH of around 7.40, pO2 of >90 mmHg and pCO2 of <40mmHg.[5] In our case, it was ischemic priapism as

confirmed by blood gas analysis of aspirated blood from the penile shaft.

The universally accepted mechanism of priapism in leukemia is sludging of blood in the corpora cavernosa which leads to painful and persistent erection.[6-8] The other described mechanisms are the congestion of corpora cavernosa secondary to raised intra-abdominal veins draining the spleen or by leukemic infiltration of the sacral plexus or central nervous system.[9]

CML is characterized by the presence of the (9;22) translocation also known as Philadelphia chromosome which can be detected by the presence of the BCR-ABL fusion gene. Priapism in CML is a rare entity and even more rarely, priapism is the first presentation of CML.[10]

The therapy concept includes emergency treatment of priapism in the form of aspiration and initiation of cytoreduction therapy for CML. If aspiration alone does not lead to detumescence then injection of a vasoactive agent (like phenylephrine) and heparin is indicated.[11]

In this case report, the patient made a complete recovery and had no sexual dysfunction owing to timely surgical intervention and initiation of cytoreduction therapy.

The importance of timely diagnosis of the cause of priapism is crucial for avoiding long-term complications and refractory priapism.

Conclusion

Priapism itself as the first presentation of CML is very rare and if not timely addressed can lead to debilitating consequences. Timely initiation of cytoreduction and targeted TKI therapy can improve outcomes. It is also necessary to exclude other causes of priapism.

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