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Priapism - A Rare Initial Presentation in Chronic Myeloid Leukemia: Case Report

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Abstract: ***Priapism is a complication rarely seen in leukemia. We report a 35-year-old man presented with persistent painful erection of penis for more than 13 days at home. The patient underwent immediate irrigation and decompression of priapism by urologist at emergency services of the hospital, which resulted in a flaccid penis later. During hospitalization, peripheral blood smear and bone marrow aspiration confirmed the diagnosis of chronic myeloid leukemia.***

Keywords: Priapism, Chronic myeloid leukemia, Emergency services, Low flow ischemia

1. Introduction

Priapism is defined as full or partial erection that continues more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation. [1] Priapism is a urological emergency, which must be treated early to prevent erectile dysfunction. Ischemic priapism accounts for the majority of cases described in the literature. Ischemic priapism is marked by rigidity of the corpora cavernosa with little or no cavernous arterial inflow. The patient typically complains of penile pain after 6–8 hours, and the examination reveals a rigid erection. [2]

Chronic myeloid leukemia (CML) is one of the myeloproliferative disorders and is a malignant clonal disease of the hematopoietic stem cells resulting in increased myeloid cells, platelets, and erythroid cells.[3] With the worldwide annual incidence of 1–1.5/100,000 population and M:F ratio of 1.8:1, CML accounts for 15–20% of all leukemias in adults. [2, 3]

Priapism is a rare initial presentation of CML occurring in 1–2% of cases and is of the low-flow (ischemic) veno-occlusive type associated with hyperleukocytosis and leukostasis or hyperviscosity syndrome. [2,3,4,5,6]

We report a case of a 35 year old male who presented with priapism as the first manifestation of chronic myeloid leukemia.

2. Case Report

The patient was a 35-year-old male married driver by occupation, who presented to the emergency department of RIMS Imphal Manipur with a 13-days history of spontaneous onset of prolonged painful penile erection. No associated history of trauma, sexual arousal/intercourse, use of aphrodisiacs or psychotropic drugs. He had no history of dysuria or urethral discharge.

He has had one similar episodes about 3 months prior to presentation, and it lasted overnight and resolved spontaneously in the morning. Patient had never consulted after the spontaneous detumescence. There was no history suggestive of sickle cell disease in the family.

On examination he was anxious looking and in painful distress, pale, anicteric and without significant peripheral lymphadenopathy or peripheral stigmata of SCD. He had hepatomegaly of 7cm below right subcostal margin and splenomegaly of 6cm below the left subcostal margin.

He had a non circumcised male phallus which was erect, turgid with differential warmth, mild blackish discoloration of penile skin and mild tenderness [Figure 1,2].

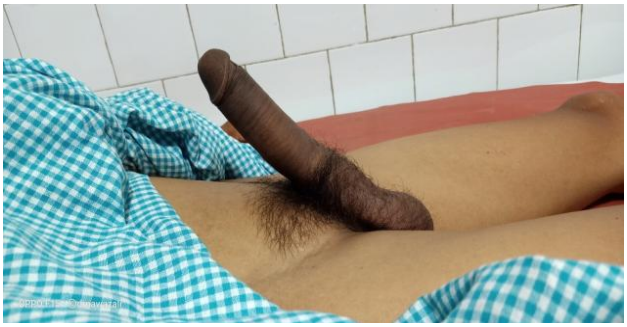

Figure 1

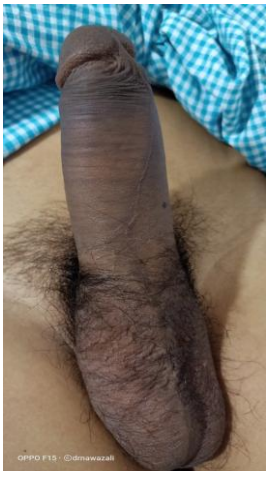

Figure 2

A working diagnosis of ischemic priapism was made by the attending urology team and this was further supported by the finding of absent arterial flow within the cavernosal arteries on penile Doppler ultrasonographic scan.

Treatment of the priapism was initiated by corporal aspiration and phenylephrine irrigation at the emergency services under the impression of low flow type priapism because of the history and physical examination. The erection gradually reduced following aspirations and washes.

His hemogram showed haemoglobin of 9.6gm/dl, a high white blood cell (WBC) count of 4.96lac/mm^3 and platelet count of 4.58lac/mm^3. The peripheral blood film shows predominantly normocytic normochromic red cells and a complete spectrum of the myeloid series with the following differential counts: myeloblasts 4%, promyelocytes 7%, myelocytes 24%, metamyelocytes 3%, eosinophils 6%, and lymphocytes 1%; with impression suggestive of CML in chronic phase.

Liver and renal function tests, uric acid, PT/INR, Ca^{2+}, Mg^{2+} and PO_4^{2-}were within normal limits, while urinalysis revealed traces of protein and red blood cells.

Haematology consultation was sought out and advised for maintaining hydration with intake of 3 liters/day, tab febuxostat 40mg. Cytoreduction done with only oral hydroxyurea at a dose of 1000 mg in three divided doses over 24h was commenced as per haematologist protocol, as there were no facilities for leukapheresis available at our center.

Kariotype and cytogenetic analysis was not done as the patient was poor and cannot afford said investigations from outside as facility in our institute was not available.

3. Discussion

The condition priapism was named after the Greek God Priapus, son of Zeus. It is believed that a jealous Hera cast a spell over his mother while pregnant causing Priapus to be born with the condition bearing his name.

Priapism is a persistent penile erection that lasts beyond 4 h, and/or is unrelated to sexual stimulation [4]. It is a rare in occurrence with an incidence of 1.5 cases per 100,000 person-years[4].

Hematological disorders are the leading cause of priapism accounting for 20% of the cases and include SCD, hyperviscosity syndromes as seen with the myeloproliferative diseases, hypercoagulable states such as deficiencies of proteins C and S, antiphospholipid syndromes, and amyloidosis[4]. Priapism is seen in about 1–5% of male patients with all types of leukemia[4].Though CML accounts for half of the cases with leukemic priapism, it is rare for priapism to be the initial presentation of CML as this occurs in only 1–2% of cases[4].

In CML, hyperleukocytosis causes ischemic priapism via aggregation of the leukemic cells in the corpora cavernosa and dorsal veins of the penis as well as infiltration of the sacral veins or the central nervous system by the leukemic cells.

The algorithm for the initial management of priapism in CML provided by Chisick *et al*[7] seems to be apt for our resource-constrained setting where CML cases usually present late coupled with dearth of diagnostic and therapeutic facilities for its management. According to this algorithm, splenomegaly in a priapic patient should raise the possibility of CML or a related hematological disorder, and in line with this, urgent full blood count with peripheral blood film should be requested [4,7].The outcome of these two initial tests will give a direction to additional testing such as hemoglobin electrophoresis, coagulation screening, or bone marrow studies. Penile blood testing and penile Doppler USS are not considered essential in CML as priapism in CML is already known to be of ischemic type, thus intervention should not be delayed on account of waiting to conduct these investigations[7].

Upon suspicion of CML, parenteral hydration and allopurinol should be commenced to minimize the risk of tumor lysis syndrome, while urgent consultation with the hematology, urology, apheresis services, and hematopathology, units should be sought.

The American Urologic Association recommends that for ischemic priapism, intracavernous treatment should be administered concurrently with systemic treatment of an underlying disorder, such as CML[4,8,9], thus initiation of cytoreduction with hydroxyurea is advocated, while more specific chemotherapy with a tyrosine kinase inhibitor (TKI) should be introduced as soon as possible. If available, combination of chemotherapy with leukocytopheresis is preferred as a single session of leukocytopheresis can reduce WBC count by about 30–60% in cases of hyperleukocytosis [7, 9].

A systematic approach is employed with the intracavernous treatment[4,7]. An initial therapeutic aspiration (with or without heparinized-saline irrigation) is conducted and if priapism persists despite several attempts, intracavernous injection of sympathomimetics is performed. It is advocated that surgical shunts should only be considered when sympathomimetics agents have failed [4].The corporoglanular is considered the primary choice of shunt procedure due to its ease and fewer complications. Presence of severe distal penile edema and tissue damage may necessitate proximal shunt procedures which are more difficult and more likely to develop complications such as urethral fistula, purulent cavernositis, and erectile dysfunction [4]. Failure of nonoperative or the less invasive shunt procedures should prompt insertion of penile prosthesis before significant intracorporal fibrosis sets in, which may make future surgical interventions unfruitful[4].

Our reported case depicts a late diagnosis of CML with an initial presentation as ischemic priapism. In the case at hand, a complete blood picture was helpful in early diagnosis of CML and early initiation of targeted chemotherapy. It is therefore recommended to have a CBC examined at presentation of any case of ischemic priapism of unknown

etiology. Besides the initial relief of priapism, the further workup and management of the underlying disease are more important. In our case, with use of a combined urological therapy and oncological treatment to priapism, the patient rapidly had relief of his clinical problem.

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