Discontinuation of treatment in a chronic myeloid leukemia patient caused priapism: A case report

Case Report

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ABSTRACT

Priapism is an unusual andrological emergency that requires urgent intervention. It is characterized by prolonged penile erection for more than 4 h not related to sexual excitation. It is classified into three main types:veno-occlusive (ischemic), arterial (non-ischemic) and stuttering priapism. Many causes can predispose to ischemic priapism including thalassemias, sickle cell anemia, hypercoagulable status, neoplastic syndromes, pelviabdominal tumors and use of some drugs and medications. However, in the majority of cases of ischemic priapism no cause could be identified. Chronic myeloid leukemia (CML) is a chronic myeloprolifertative tumor that is characterized by a variety of vague clinical manifestations. Priapism as a complication of CML is a rare clinical presentation. It may cause priapism as a result of hypercoagulability due to hyperleukocytosis. Here, in this report, the case of a known CML male patient who presented with venoocclusive priapism for about 16 h after abrupt discontinuation of his cytoreductive therapy is described. Thus, all medical staff, either (General practitioners) GPs or specialized in different fields of medicine, should be aware of priapism as an emergency and that early diagnosis and multidisciplinary management minimize the inevitable damage of the erectile corpora cavernosa as well as improving the outcome.

Key Words: Andrological emergency, chronic myeloid leukemia, leukocytosis, priapism, veno-occlusive.

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INTRODUCTION

Priapism is a rare andrological emergency with an estimated incidence of 1.5/100000 male populations. It has been found that CML has a bimodal pattern, wherein it may affect first the children with sickle cell disease between the age of 5 and 10 years, and, later on, it may affect adult male individuals between the ages of 20 and 50 years. Priapism is characterized by prolonged penile erection lasting for more than 4 h that is not related to sexual excitation.

In general, it is classified into three main types: ischemic (low-flow or veno-occlusive), arterial (high flow or nonischemic) and stuttering (recurrent or intermittent) priapism[1].

Veno-occlusive priapism is considered as the most common, whereas in it accounts for more than 90–95% of cases. It is due to venous outflow obstruction leading to impaired perfusion of cavernous tissue with subsequent hypoxia, hypercapnia and acidosis. A single attack of ischemic priapism may end up in cavernosal fibrosis with subsequent erectile dysfunction, if it is not managed urgently or properly. Several etiologies have been mentioned in the literature causing ischemic priapism, which include hemoglobinopathies including thalassemias and sickle cell anaemia, hypercoagulable status, neoplastic syndromes, compressive pelviabdominal masses and use of some recreational drugs and medications (intracavernosal injections of papaverine, phentolamine and PGE1, anticoagulants, α-blockers, antidepressants and antipsychotics)[2]. However, in the majority of cases of ischemic priapism, no cause could be identified and, in such cases, are known to be idiopathic. Early detection and management of such cases by aspiration/irrigation along with the use of sympathomimetic agents gives a better outcome. However, if not managed early, the resolution of erection may take days, and erectile dysfunction occurs, and that may end up in penile prosthesis implantation[3].

Chronic myelodysplastic leukemia (CML) affects the bone marrow stem cell in which uncontrollable proliferation of mature granulocytes (neutrophils, eosinophils and basophils) and their precursors occurs. It was found that CML has an incidence of 1–2 cases per 100000 adults. Concerning the etiopathogenesis of CML, it has been

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described that a gene mutation resulting from a balanced translocation, t(9; 22) (q34; q11.2), including a fusion of the Abelson gene (ABL) from chromosome 9q34 with the breakpoint cluster region (BCR) gene on chromosome 22q11.2 is the cause. The resulting anomalous chromosome is known as 'Philadelphia chromosome' carrying the BCR-ABL oncogene that encodes for the expression of an oncoprotein known as BCR-ABL 1, which is responsible for the occurrence of CML[4].

In general, CML is divided into three distinctive phases on the basis of clinical characteristics and laboratory findings, which are chronic, accelerated and blast phases, wherein about 85% of patients present with the chronic phase at the time of diagnosis, and, years later, they may progress to an accelerated phase and blast crisis[5]. Unfortunately, in most of the cases, the diagnosis of CML is reached late, as it has a large variety of vague clinical manifestations that may include lymphadenopathy, fatigue, hepatosplenomegaly, weight loss, bleeding tendency, and thromboembolic phenomena due to hyperleukostasis[6]. Priapism as a complication of CML is rare, as only few cases have been reported in the literature ; hereby, our case report is about an adult who had encountered priapism due to CML after abrupt discontinuation of therapy.

CASE PRESENTATION

A 30-year-old male patient presented to the emergency department complaining of prolonged painful penile erection for about 16 h after discontinuation of cytoreductive therapy for the treatment of CML. He was diagnosed with CML since 6 months upon which he received treatment, but he was non compliant; he stopped it abruptly and did not receive the treatment for 2 weeks before priapism. There was no past history of prolonged penile erections, genital trauma or other drug intake apart from cytoreductive therapy for CML.

General examination showed that the patient was pale and underweight. In addition, nontender mildly enlarged cervical, axillary and inguinal lymph nodes could be felt. Abdominal examination showed mild nontender hepatomegaly and huge splenomegaly. In contrast, genital examination showed that the penis was fully erect, tender and mildly edematous.

Initially, cavernosal blood aspiration was performed for blood gas analysis and showed ischemic pattern of priapism (i.e. hypoxia, hypercapnia and acidosis). A complementary penile duplex examination was performed and revealed absence of blood inflow in both cavernosal arteries, denoting veno-occlusive priapism. Afterwards, the patient was admitted for urgent evacuation for priapism as an initial time-dependent salvage procedure for the cavernous tissue. Using a butterfly cannula 19G, aspiration and irrigation with sympathomimetic (ephedrine) was performed several times under complete aseptic conditions in the OR and after performing penile block anesthesia using bupivacaine (without adrenaline) in a concentration of 0.25%. Fortunately, priapism was evacuated by this procedure.

Afterwards, the patient was admitted to the hematological ward for further investigations to rule out other causes of priapism, where complete blood count with differential, toxicology screening, hemoglobin electrophoresis, coagulation profile, pelviabdominal US, and semen and urine cultures were carried out. All of them were normal, but complete blood count with differential showed low hemoglobin level of 10.3g/dl, increased total leukocytic count (TLC) of 210000/mm^3, and low platelet count of 45000/ mm^3. In addition, the blood smear examination showed immature leukocytes in different stages of differentiation with 2% blasts ; basophilia was prominent without dysplasia. Thus, bone marrow aspirate was performed and showed marked hypercellularity due to marked granulocytic proliferation and increased dwarf megs; some Pseudo-Gaucher cells were seen, and myeloid blasts represent 4.4% myeloperoxidase immuno histochemistry, proving the myeloid origin of the blasts. Karyotype analysis showed the Ph1 chromosome BCR-ABL1 positive by FISH with dual-color; therefore, a final diagnosis of CML was reached. In addition, concomitant management of leukocytosis was performed, whereas hydroxyurea therapy (1000 mg, twice daily) associated with intravenous fluid hydration was started. Besides that, he was prescribed allopurinol (300mg/day) to avoid the occurrence of tumor lysis syndrome. Thereafter, with 2 weeks of chemotherapy by hydroxyurea, the patient's TLC and platelet counts were 65000 and 232000/mm^3, respectively. Later on, he will be a candidate to receive tyrosine kinase inhibitor when TLC becomes less than 50000/mm^3 to avoid tumor lysis syndrome. During the outpatient follow-up visits at 3 and 6 months from discharge, the patient mentioned that he experienced morning penile tumescence with no rigidity and some difficulty in coital erections; however, he was doing well on phosphodiesterase-5 inhibitors that were prescribed to him. Moreover, he did not experience any further attacks of priapism after being compliant on treatment and attending regularly for follow-up at the hematology unit.

DISCUSSION

Priapism is a rare andrological emergency. Early intervention is so crucial to preserve the erectile function of the penis. It has been mentioned that hematological disorders associated with hypercoagulability may precipitate in 20% of priapism cases. In contrast, sickle cell disease in children may account for 67% of cases, while leukemia accounts for 15% of cases of priapism. However, in adults, priapism associated with leukemia may account for 1–5% of cases. In addition, priapism in leukemic patients is much more common among CML than acute leukemia patients[7].

Several hypotheses have been suggested, trying to explain the pathogenesis of priapism in CML patients; however, all of them are still a matter of debate. They include the following :(i) hyperleukocytosis leads to leukostasis and hyperviscosity with subsequent microemboli and thrombi; (ii) venous outflow obstruction due to abdominopelvic vein compression by organomegaly; (iii) hypercellular infiltration of the sacral nerves by leukemic cells; and (iv) central nervous system involvement[8].

Because of the relatively small number of reported cases of priapism in CML patients, there is no standardized treatment protocol. However, the American Urological Association has published practice guidelines based on expert panel discussion and review of the limited data available that recommended the concurrent use of systemic cytoreductive treatment for CML and local cavernosal decompression to preserve the erectile function[9]. Urgent decompressive intervention through aspiration and irrigation with concomitant use of sympathomimetics (α-agonists) is the main standardized approach to preserve the erectile function. However, in refractory or neglected ischemic priapism (i.e. duration>48h), shunt operations or even urgent penile prosthesis implantation may be considered[3,10]. In contrast, systemic cytoreductive therapies used in CML patients include high-dose hydroxyurea, and tyrosine kinase inhibitors, such as imatinib, with or without concomitant leukapheresis, have been recommended, trying to reduce hypercoagulability[11].

CONCLUSION

Despite priapism being considered a rare clinical presentation of CML, it is recommended that, once it is clinically encountered, early diagnosis and the subsequent multidisciplinary therapeutic approach should be implemented, not only to control priapism and preserve erectile function, but also to prevent further inevitable complications.

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CONFLICT OF INTEREST

There are no conflicts of interest.

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