Asian Journal of Pharmaceutical and Health Sciences
www.ajphs.com

Priapism in Teenager Chronic Myelogenous Leukemia: A Rare Occurrence

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ARTICLE HISTORY
Received: 11-Jul-2011
Accepted: 20-Aug-2011
Available online: 10-Nov-2011

Keywords:
Priapism, CML, Treatment.

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ABSTRACT

Priapism in Chronic Myelogenous Leukemia(CML) is an unusual presentation but some cases are diagnosed due to complain of priapism. Priapism may be due to leukostasis associated with greatly exaggerated blood leukocyte count as low flow (ischemic) or high flow (non ischemic) type. Treatment option includes- aspiration, sympathomimetic agents, leukapheresis, cytotoxic therapy and shunt surgery. Study case was a 11-year-old boy presented with priapism and fullness in abdomen. Careful physical examination and blood investigations revealed CML. We treated conservatively and priapism resolved in 24 hours.

INTRODUCTION

Priapism is a prolonged, painful and persistent erection unassociated with sexual arousal. Although the injection of intracavernosal vasoactive substances is the most common cause of priapism but nearly 20% of all cases relate to a hematologic disorder. Such hematologic conditions include sickle cell anemia, chronic myelogenous leukemia (CML), chronic lymphocytic leukemia(CLL) and acute lymphoblastic leukemia(ALL) [1]. In children, the most common cause for priapism is a hematologic disorder such as sickle cell anemia (67%). Leukemia and idiopathic events are the next most common causes of priapism in children [2]. In adult leukemic patients, the incidence of priapism is estimated to be approximately 5% [3]. This article discusses the case of a 11-year-old patient who presents with priapism and is found to have chronic myelogenous leukemia.

CASE REPORT

A 11-years-old boy presented with complain of continuous painful erection of penis for last 12 hours and gave history of two similar episodes in last two months (Fig. 1). On further questioning patient complained of generalized weakness for 4 months and fullness of abdomen for one month. He denied history of any illicit drug and trauma.

On examination, Penile shaft was erect, stiff but glans was flaccid. The corpora cavernosa were engorged. Physical examination revealed normal cardiovascular and pulmonary systems. Spleen was palpable 4 cm below costal margin. Ultrasonography shows spleenomegaly and mild hepatomegaly. Because of the patient's young age and lack of predisposing causes for priapism, a complete blood count (CBC) with differential is ordered. The leukocyte count is 2,90,000/mm^3, platelet 5,50,000/mm^3, and hematocrit is 22%. Differential reveals 17% metamyelocytes, 5% myelocytes, and 7% promylocyte. Peripheral smear demonstrates immature leukocytes in various stages of differentiation (**Fig. 1**). Based on the CBC, the diagnosis of chronic myelogenous leukemia is made.

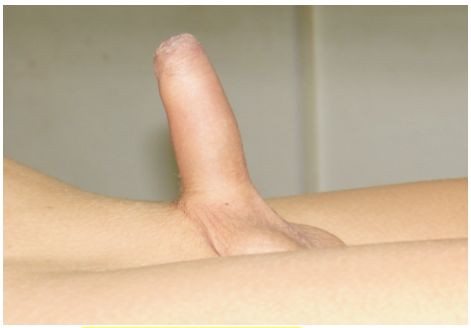

Fig. 1: Priapism in teenager

Because of the short duration of the patient's priapism (12 hours), mild penile pain at the time of examination, and the lack of ischemia in the penis, a urology service is consulted concerning treatment. The urologist elects conservative management and suggests aggressive treatment of the underlying condition instead of corporal aspiration/irrigation.

The patient was admitted in the hospital and received intravenous fluid hydration, hyper transfusion and allopurinol (400mg per day). Bone marrow aspiration was performed and confirms the diagnosis of chronic myelogenous leukemia and send for immunochemistry. Mean time we started hydroxyurea. The priapism resolved in 24 hours after starting treatment. After immunochemistry imatinib mesylate 400mg once a day started. Presently, the patient's leukemia is in remission. He is able to achieve an erection with manual stimulation and maintains the ability to ejaculate. The patient is currently sexually inactive.

DISCUSSION

Priapism is a prolonged, painful and persistent erection unassociated with sexual arousal. Priapism can be low flow (ischemic) or high flow (non ischemic) type.

Low-flow priapism results from decreased penile venous outflow causing stasis and presents as a painful, rigid erection. It is more common than high-flow priapism, and it is a medical emergency because irreversible cell damage and fibrosis can occur if treatment is not initiated within 24-48 hours. Low-flow priapism can be drug induced or caused by hematologic disorders (i.e. Sickle cell anaemia, thalassemia and Leukemias) and tumour infiltration[4]. Priapism as a result of hematologic malignancy is most likely caused by venous obstruction from microemboli/thrombi as well as hyperviscosity caused by the increased number of circulating leukocytes in mature and immature forms[5].

High-flow priapism results from increased arterial inflow into the cavernosal sinusoids, which overwhelms venous outflow. Clinical presentation is painless erection; irreversible cellular damage and fibrosis are rare. High-flow priapism often is the result of penile or perineum trauma and is not an emergency, treatment is elective[6].

First-line treatment of priapism is aspiration of blood from the base of the corpora cavernosa. The success rate with aspiration alone is approximately 30%. If the treatment is unsuccessful, instillation of the sympathomimetic agent phenylephrine hydrochloride[7].

In cases of hematologic malignancy, controversy has existed regarding the optimal treatment of leukemic priapism. Earlier series of case reports show successful detumescence with local radiation therapy, open surgical shunting, or combination of both treatments[2,8]. More recent literature has focused on the use of cytoreductive modalities such as chemotherapy or combination of chemotherapy and leukapheresis[9,10]. Because of the relatively rare occurrence of leukemic priapism and the small number of case series, there is no standard treatment protocol at this time. Chemotherapy or radiotherapy may first be attempted [11]. If detumescence is not achieved, then surgical shunting should be considered. This includes placing a shunt between the corpora cavernosa and glans, which allows for blood to flow in and out of the corpora cavernosa [12].

Other physicians have suggested a more specific time course. Suri *et al* [9] recommend that chemotherapy and leukapheresis should be initiated immediately. A similar conclusion was reached by Becker *et al* [10], who recommended chemotherapy and leukapheresis followed by surgical shunting if detumescence is not "immediately obvious." These articles seem to suggest an early, more aggressive use of surgery. Such urgent treatment is not always necessary in cases of leukemic priapism. However, in pediatric patients with priapism, 50% of cases are caused by chronic granulocytic leukemia, which carries a more favorable prognosis and greater life expectancy than other leukemias[3]. In such cases, an aggressive treatment protocol may provide a greater chance of maintaining normal erectile function. In this case study, surgical shunting was a less favorable option because of the patient's response to chemotherapy and hydration. Further review of such cases with long-term follow up is necessary to more precisely define an optimal treatment protocol.

CONCLUSIONS

A low flow priapism case should be thorouly examined and investigated. Priapism due to CML can be managed conservatively in safe mode.

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