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Case Report

Priapism: a rare presentation in chronic myeloid leukemia

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ABSTRACT

Priapism is a rare presenting feature of Chronic Myeloid Leukemia (CML). It is an urological emergency which requires urgent treatment to prevent long term complications, in particular erectile dysfunction. Author report a case of 18 year old male presenting with persistent painful erection of penis for around 14 hours. The patient underwent immediate irrigation and decompression of priapism in emergency and was started on cytoreductive therapy. During hospitalization, peripheral blood smear and bone marrow aspiration confirmed the diagnosis of CML.

Keywords: Chronic myeloid leukemia, Complications, Emergency, Priapism

INTRODUCTION

Priapism is defined as prolonged, painful irreducible erection that continues for more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation and not resulting in ejaculation. Clinically and pathologically types of priapism are ischemic (low flow), non-ischemic (high flow) and stuttering priapism. Ischemic priapism is the most common subtype.[1] Hematological disorders leading to priapism are sickle cell anemia, essential thrombocythemia, CML, chronic lymphocytic leukemia, and acute lymphoblastic leukemia.[2] Authors report a case of patient presenting with priapism who was managed promptly in emergency and later diagnosed with CML.

CASE REPORT

An 18 year old male presented in emergency department with chief complaint of fever on and off since one month not associated with rigors and chills. He also complained of generalized weakness, fatigue, loss of appetite and weight loss. Patient passed urine following which he had persistent painful erection of penis for around 14 hours. His penis remained erect, painful and swollen. He denied recent intercourse, trauma, use of illicit drugs or any radiation therapy. The vital signs revealed a body temperature of 101 F. Blood pressure 122/84, Pulse rate 88/min, respiratory rate 14/min. Patient was alert and oriented to time, place and person.

Physical examination revealed that spleen was palpable 9 cm below the left costal margin while liver was palpable 6 cm below right costal margin. His conjunctiva was pale but no jaundice. Penis was erect, firm and tender with superficial venous engorgement. Urinalysis was normal. Laboratory data showed hemoglobin 9.7 gm/dl, hematocrit 27.1%, TLC- 3,62,650/mm^3, platelet count-5.27 lac/mm^3, DLC- N-43%, L 2%, E 3%, B 2%, myelocytes and metamyelocytes 39%, blast and promyelocytes 11%, reticulocyte count 1.3% and serum uric acid 6.8 mg/dl. His viral markers were non-reactive. Bone marrow aspiration revealed hypercellular marrow

with increased myeloid series showing immature forms. It demonstrated accelerated phase of CML. Quantitative real time PCR analysis of BCR-ABL gene rearrangement revealed percentage ratio of (p210 and ABL copy) 62.63%. The treatment of priapism was initially performed by cavernosa aspiration and irrigation with epinephrine in the emergency department. The erection was relieved later by this procedure. He was started on hydroxyurea therapy (50 mg/kg/day) and Imatinib 800 mg/day. Allopurinol 300 mg daily along with adequate hydration was also started. After 1 week the patients TLC and platelet count dropped to 1.68 lac/mm^3 and 3.36 lac/mm^3 respectively. Patient had no recurrent episode of priapism during the stay in hospital. On follow up at 1 month, TLC and platelet count was reduced to 3800/mm^3 and 1.56 lac/mm^3 following which dose of imatinib was reduced to 400 mg/day. The patient continues to report to us without any erectile dysfunction until the date of writing.

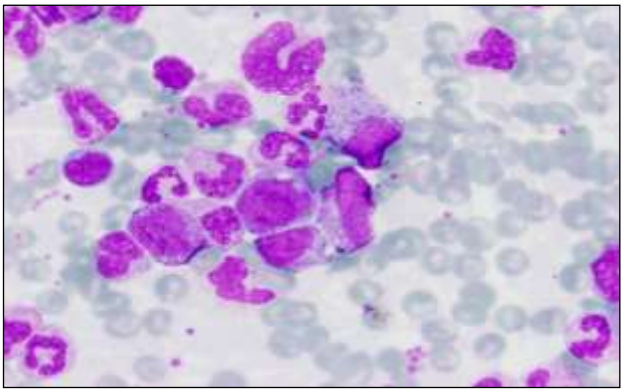

Figure 1: PBF film of patient in accelerated phase of cml showing "left-shift" of leucocytes with myelocytes, metamyelocytes and myeloblasts (1000x magnification).

DISCUSSION

Priapism is an andrological emergency with poor prognosis if not treated on time. If untreated within 24-48 hour, it can result in irreversible damage and fibrosis, leading to problems like erectile dysfunction or future episodes of persistent and prolonged priapism.[3] In adult leukemic patients, the incidence of priapism is approximately 1-5%.[4] Leucostasis as a result of hyperleukocytosis is present in around 12% of adult patients presenting with CML and around 60% of children diagnosed with CML.[5] Hematological conditions are the cause of 20% of cases of priapism in men. Priapism due to hematological disorder is most likely due to venous obstruction from micro emboli/thrombi as well as hyper viscosity due to increased number of circulating leukocytes in mature and immature forms.[6] It is also seen that increased production of cytokines and adhesion molecules by leukemia cells results in endothelial cell activation and leads to increased sequestration of cells in the microvasculature.[7]

Systemic therapies that are commonly used in CML patients include cytoreductive therapy, such as high-dose hydroxycarbamide and Tyrosine Kinase Inhibitors (TKIs), with or without the addition of leukapheresis to reduce hyperviscosity.[8] Therapeutic aspiration (with or without irrigation) or intra-cavernous injection of sympathomimetics should be initially tried to treat priapism. A penile block may be performed by injecting 10-20 ml of 1% lignocaine below the symphysis pubis to block the dorsal nerves to the penis. A tourniquet is applied to the base of the penis. A 16 or 18 gauge bio valve intra-venous catheter can be inserted into the corpus cavernosum laterally through the penile skin, avoiding the ventral urethra and dorsal neurovascular bundle. Around 20-30 ml of blood is aspirated and heparinized saline may be injected. Repeated aspirations over 1 hour may be needed and upto 50 ml of blood can be aspirated. On its own, aspiration has around 30% success rate.[9] In patients with leukemia induced priapism, the use of leukapheresis (mechanical white cell depletion) to rapidly reduce hyper viscosity in patients is also well-documented.[10]

CONCLUSION

This case is unique as priapism is a complication rarely seen in leukemia. The importance of prompt diagnosis and treatment of priapism cannot be overemphasized, as there is definite incidence of impotence following priapism. Besides the initial relief of priapism further workup and management of the underlying disease is more important. In this case, with use of a combined surgical and oncological treatment for priapism, the patient rapidly had relief of his clinical problem.

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