Chronic myeloid leukaemia presenting with priapism as the only symptom: A case report and review of literature.

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Case Report

Abstract

Objective: There have been few reports of priapism as complications of various haematological malignancies. Delays in establishing the cause and late institution of appropriate intervention in patients suffering from priapism could result in long-term complications, especially erectile dysfunction. We present here a case of chronic myeloid leukaemia (CML) in a 30 year old male who presented with priapism as the only symptom. Apart from presenting this as a rare case report, it also emphasizes the need for early full blood count in patients presenting with priapism.

Methods: The case note of the patient, as well as all available literature on the subject were reviewed.

Results: The 30 year old man who presented with a 9 (nine) day history of persistent painful non-sexually related penile erection was found to have chronic myeloid leukaemia (CML) following peripheral and bone marrow aspiration studies. He however had a delay in referral and diagnosis of his primary condition and eventually had erectile dysfunction. The priapism responded well to surgical intervention and use of cyto-reductive therapy. The erectile dysfunction could have been averted if complete blood count and, or peripheral blood film were carried out at the time of first presentation

Conclusion: Priapism due to hyperviscosity can be the first presentation in patients with Chronic Myeloid Leukaemia, and CML should therefore always be considered in a patient with priapism. High Index of Suspicion, as well as early full blood count and peripheral blood/Bone marrow examination among patients presenting with priapism will facilitate prompt and correct diagnosis, and reduce, if not prevent erectile dysfunction and other severe complications that may arise from priapism.

Keywords: Priapism, Chronic myeloid leukaemia, erectile dysfunction

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La leucémie myéloïde chronique présentant priapisme comme le seul symptôme: Un rapport de cas et revue de la littérature.

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Rapport de cas

Resume

Objectif: Il y a eu quelques rapports de priapisme comme complications de diverses hémopathies malignes. Les retards dans l'établissement de la cause et de l'institution tardive d'une intervention appropriée chez les patients souffrant de priapisme pourraient entraîner des complications à long terme, en particulier la dysfonction érectile. Nous présentons ici un cas de leucémie myéloïde chronique (LMC) chez un homme de 30 ans qui a présenté avec priapisme comme le seul symptôme. En dehors de la présentation de ce qu'un rapport de rares cas, il souligne également la nécessité d'une numération formule sanguine précoce chez les patients présentant un priapisme.

Méthodes: La note de cas du patient, ainsi que toute la littérature disponible sur le sujet ont été examinées.

Résultats: Le vieil homme de 30 ans qui a présenté avec une histoire 9 (neuf) de jour de la persistance douloureuse érection du pénis non liées sexuellement a été constaté que la leucémie myéloïde chronique (LMC) suite à des études d'aspiration de la moelle osseuse et de périphériques. Il a cependant eu un retard dans l'orientation et le diagnostic de son état primaire et a finalement dû dysfonction érectile. Le priapisme a bien réagi à l'intervention et l'utilisation de la thérapie cyto-réductrice chirurgicale. La dysfonction érectile aurait pu être évitée si la numération globulaire complète et, ou d'un film de sang périphérique ont été réalisées au moment de la première présentation

Conclusion: Priapisme en raison de l'hyperviscosité peut être la première présentation chez les patients atteints de leucémie myéloïde chronique, et la LMC doit donc toujours être considéré dans un patient avec priapisme. Haut Indice de suspicion, ainsi que tôt hémogramme et du sang périphérique / os examen de la moelle chez les patients présentant un priapisme facilitera un diagnostic rapide et correct, et de réduire, voire prévenir la dysfonction érectile et d'autres complications graves qui peuvent résulter de priapisme.

Mots-clés: priapisme, la leucémie myéloïde chronique, la dysfonction érectile

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INTRODUCTION

Priapism is a urological emergency that requires urgent intervention to prevent erectile dysfunction(1). It is defined as persistent, painful, penile erection not related to sexual stimulation. It is classified into high-flow (non-ischaemic) and low-flow (ischaemic) types(2). Low-flow priapism results from reduced penile venous drainage, while high-flow priapism results from increased carvenosal arterial inflow.

Priapism is a rare medical condition with a worldwide incidence of 1.5 per 100,000 persons per annum(3). Most cases (64%) of priapism are idiopathic while about 20% are caused by haematological disorders. Although haematological conditions are the major cause of ischaemic priapism, it is however rare in leukaemias. It is important therefore to evaluate all cases of priapism for leukaemias in order to facilitate early diagnosis, allow prompt institution of appropriate interventions, and avoid potential complications.

We report here a 30 year old, Nigerian male, who presented to the emergency department with nine days history of persistent, painful, penile erection. Investigations revealed that he had chronic myeloid leukaemia which was missed in two different health facilities 9 days prior to presentation to our facility.

Case presentation

A 30 year old Nigerian man of the Tiv tribe presented to our emergency department with 9 days history of persistent, painful, penile erection which he noticed on waking up from sleep. He had a stuttering episode a month prior to presentation. His last sexual activity was about 2 weeks prior to the onset of the symptom. He had no antecedent trauma to the penile area and no history of using sex enhancing drugs. He was not a known sickle cell disease patient. There were associated recurrent bone pains which started a year ago with occasional yellowness of the eyes. There was no history of gum bleeding, bleeding from any orifices, or into the skin but he complained of prolonged bleeding after minor cuts of one year duration. There was no fever, weakness, or cough.

He had been to two other hospitals prior to presentation. In the last hospital before ours, he had aspiration and irrigation without achieving sustained detumesence.

On examination, he was in painful distress, his pulse rate was 96 beats per minute, blood pressure was 130/80 mmHg. Chest was clinically clear. Abdominal examination revealed an enlarged, non-tender spleen of 10cm below the left costal margin. The liver and kidneys were not palpably enlarged. Penile examination showed an erect, edematous penis with hard shaft and soft glans and multiple blot clots at the puncture sites for aspiration. The superficial penile veins were engorged. (Fig.I)

Complete blood count showed moderate anaemia of 9.2g/dl, mild thrombocytopaenia of $86x10^9$/l, and marked neutrophil leukocytosis of $261x10^9$/l with neutrophil differential of 96%. On peripheral blood film examination, anisopoikilocytosis, polychromasia, and nucleated red cells were noted. The granulocytes were markedly elevated with complete spectrum of the maturing myeloid cells present. There were elevated numbers of basophils and eosinophils.

The Urea and uric acid were mildly elevated but creatinine was normal. Liver function test was normal. Bone marrow aspiration morphology showed myeloid hyperplasia consistent with chronic myeloid leukaemia.

Cytogenetic studies for BCR-ABL1 fusion gene and cavernosal blood gas analyses were not done due to lack of facilities.

Following the result of these investigations, the diagnosis of chronic phase CML with low flow priapism was made.

T he patient was commenced on hydration with 0.9% normal saline to alternate with 5% dextrose water. He had aspiration of the corpora cavernosa and irrigation with epinephrine and achieved detumescence which was not sustained. Allopurinol 300mg daily and hydroxyurea 2g daily were commenced immediately. He had bilateral T-shunts after the commencement of hydroxyurea and achieved detumescence which was sustained.(Fig.II)

On day 16, his neutrophil count dropped to $99.2x10^9$/l, his haemoglobin and platelets appreciated to 10.7g/dl and $317x10^9$/l respectively. He was discharged on 1g hydroxyurea daily. He was advised on the need to carry out cytogenetics analysis for Philadelphia chromosome. We plan to refer him to a centre where he can assess tyrosine kinase inhibitor if the result of this investigation is positive.

Follow up

The patient reported to us a month after discharge and complained of inability to achieve strong erection.

DISCUSSSION

Priapism is a condition that is characterized by unwanted and sustained erection that does not result from sexual desire and not relieved by sexual activity(4). It has a bimodal distribution of age of onset with peaks at 5-13 years of age and 21-29 years of age(5). In this case our patient presented at age of 30 years which is around the second peak age of onset. It is a surgical emergency because it is a form of compartment syndrome and therefore it is a serious medical condition that can affect sexuality and reproductive life in those affected(6).

Priapism results from either increased arterial inflow (high-flow) or, more commonly, from the failure of venous out flow (low-flow or ischaemic type), resulting in the trapping of blood within the erectile tissues of the penis(7, 8, 9). Our patient's priapism is most likely the low-flow or ischaemic type because of the finding of hyperleucocytosis, engorged penile superficial veins, and turgid penile shaft with soft glans which are the typical findings in low- flow, ischaemic priapism. Intracavernosal blood gas analysis would have been helpful in distinguishing these types but we do not have the facility to carry it out. In high-flow priapism, the intracavernosal blood gas analysis is normal but in low-flow priapism the intracavernosal blood pH is less than 7.00, the partial pressure of carbon dioxide (PCO2) more than 60 mmHg, and partial pressure of oxygen (PO2) is less than 30 mmHg(10, 11).

Priapism is uncommon in the general population but it has been recognized as a serious complication of sickle cell disease(12). It occurs in about 40% of men with sickle cell anaemia(13). Less common causes of priapism are trauma to the penis, leukemia, cancerous invasion of the penis, drugs, alcoholic ingestion, various thromboembolic disease, and intravenous fat for parenteral nutrition(14).

Chronic myeloid leukaemia (CML) is a clonal disease that results from an acquired genetic change in a pluripotential haemopoietic stem cell. This change leads to the proliferation of the cell and the accumulation of a population of cells that gradually displaces normal haemopoiesis. The end result is a greatly expanded total myeloid mass(15).

Cytogenetically, CML results from a reciprocal translocation of chromosomal materials between the long arms of chromosome 9 and chromosome 22, [t(9;22)(q14;q11)]. This translocation leads to the generation of the fusion gene BCR-ABL1 and subsequently formation of a fusion protein with increased tyrosine kinase activity.

CML presents majorly with features of hyper metabolism, splenomegaly, and features of bone marrow failure. Savage et al (16) reported that Fatigue or lethargy was the most common symptom of CML followed by bleeding, weight loss, and complaints attributable to splenic enlargement. Our patient did not present with these common symptoms but admitted to a year history of prolonged bleeding following injuries which he did not consider serious enough to require medical attention. If he had presented a year earlier, when he first noticed the abnormal bleeding, his diagnosis would have been established and treatment commenced. Symptoms of leucostasis including priapism have been noted to be rare and unusual in CML(14, 16). It has been suggested that the low incidence of leucostasis in CML was probably related to the inverse correlation between the WBC and haemoglobin concentration, which tends to prevent a marked rise in whole blood viscosity(17-20)

Many mechanisms for the development of priapism in CML patients have been postulated but cardinal among them is hyperleucocytosis, which is described as white cell counts in the excess of 100 cells $x10^9/l$. Our patient had a total WBC count of $261x10^9/l$ at presentation. The different pathophysiologic mechanisms for priapism in CML are sludging of leukemic cells in the corpora cavernosa and the dorsal veins of penis, venous congestion of the corpora cavernosa from mechanical pressure on the abdominal veins by the splenomegaly and infiltration of the central nerve.

Management of priapism depends on whether it is high or low-flow. Leukaemic priapism is usually ischaemic (low-flow) and therefore a urological emergency requiring urgent intervention. Our patient had hyperleucocytosis and features consistent with low-flow. Because leukaemic priapism is a relatively rare occurrence, there is no standard treatment recommended for it. However, it is strongly recommended by the American Urological Association that systemic treatment of an underlying disorder, such as CML, should not be undertaken as the only treatment for ischaemic priapism(21). Intracavernous treatment is required, and should be administered concurrently. The index patient had aspiration of

the corpora cavernosa and irrigation with epinephrine and achieved detumescence which was not sustained. Allopurinol 300mg daily and hydroxyurea 2g daily were commenced immediately to ensure cytoreduction. Cytoreduction could also be achieved by carrying out leucopharesis, but we are yet to commence this procedure due to lack of facility.

He had bilateral T-shunts and achieved detumescence which was sustained. This strongly supported the recommendation made above concerning combining the treatment for the primary cause of priapism with intracavernosal intervention. T shunt is a form cavernoglanular shunt, other cavernoglanular shunts include the Ebbehoj, Winter and Al-Gorab shunts. These are distal shunts. Other forms of treatment for priapism include proximal shunts such as the Quackles and caverno-venous shunts. On follow up patient complained of inability to achieve erection for sexual activity. This is usually a complication of delay in achieving detumensence within 24-48 hours of development of priapism. Our patient had priapism for 9 days.

We are reporting this case to underscore, and emphasize the value of simple and inexpensive laboratory investigations (complete blood count and peripheral blood film) in the diagnosis and management of patients with priapism.

Conflict of interest: No conflict of interest was declared.

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Figure I: Penis prior to bilateral T-shunts and commencement of cytoreductive therapy

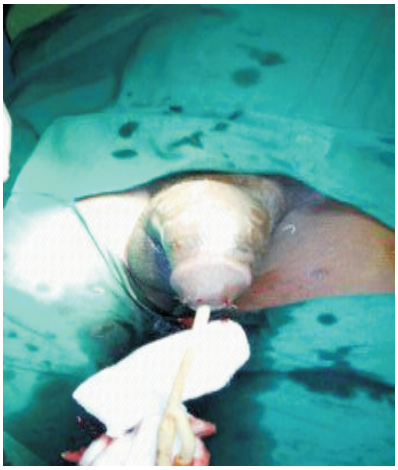

Figure II: Penis immediately after bilateral T-shunts