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Case Report

Priapism and its Sequelae in a Young African Man with Myeloid Neoplasm

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Abstract

I present a rare case of priapism in a young African man with myeloid neoplasm, who developed erectile dysfunction due to delay in making correct diagnosis and instituting appropriate treatment. Leukaemic priapism being a rare presentation needs more awareness and formation of standard treatment protocol to avert the undesirable irreversible damage that may be associated with this condition.

Keywords: Priapism; Erectile dysfunction, Chronic myeloid leukaemia; Awareness.

Introduction

Priapism is a urological emergency that can be defined as persistent, painful, penile erection that is unrelated to sexual stimulation and lasting more than four hours [1]. The condition is found in about 5% of male patients with Leukaemia [2,3].

Priapism can be classified pathologically into two: Ischaemic and non ischaemic depending on the degree of affectation of the vascular mechanisms that control penile erection [4].

Ischaemic priapism has been associated with hyperviscosity syndromes as seen in some haematological conditions like Chronic myeloid Leukaemia, Polycythaemia rubra vera and sickle cell disease [5,6].

The hyperviscosity syndrome leads to sludging of blood within the corpora cavernosa and persistent engorgement thereby causing tissue hypoxia and ischaemia of the smooth muscle [7].

Pryor et al reported that Ischaemic priapism that last more than 24hours usually leads to erectile dysfunction in about 90% of men [8].

Case Report

A 20-year-old male who was referred from a primary health facility to emergency unit of our hospital on account of persistent painful penile erection that is unrelated to sexual stimulation of three days' duration. There was no previous history of such in the past. Not a known sickle cell disease patient. There was positive history of significant weight loss, headache and tinnitus. The patient had repeated aspiration from the penile shaft at referral center to relieve the penile erection to no avail.

He was reviewed by the Urologist and a tentative diagnosis of priapism? cause was made. He then had complete blood count done which showed marked leukocytosis on account of which Hematologist were invited to review. Packed cell volume 22%, white cell count: 626x 109 /L and Platelet count 89,000 x 109/L. Differentials: Myeloblast 25%, Promyelocyte 15%, Myelocyte 20%, Metamyelocyte 15%, Band form 15%, Basophil 5%, Eosinophil 5%.

The peripheral blood film showed marked leukocytosis. Bone marrow aspiration cytology was hypercellular with myeloid metaplasia. The myeloblast plus promyelocyte was 40%.

On examination, he was in painful distress, febrile, pale, anicteric, a cyanosed and no significant peripheral lymphadenopathy.

Findings in the urogenital system showed massive swelling of the penis and scrotum.

Abdomen is grossly distended; Liver was enlarged and 10cm below the right coastal margin.

The spleen was also palpably enlarged and measured 20cm below left coastal margin.

In view of the peripheral blood film and bone marrow aspiration cytology which were consistent with chronic myeloid leukaemia in blast phase. A diagnosis of priapism secondary to chronic myeloid leukaemia was made.

He was resuscitated with 2 units of blood transfusion and Hydroxyurea 1gm tid was commenced immediately. He was also hydrated with Intra venous fluids, had parenteral antibiotics, allopurinol and analgesics.

The patient made some clinical improvement with gradual reduction in the total white cell count from 625 x 109/L to 468 x 109/L within 72hours of commencement of Hydroxyurea.

The scrotal and penile oedema also regressed significantly but the patient failed to achieve penile erection. He subsequently developed depression and was referred to be seen by clinical psychologist.

Discussion

Our patient presented with ischaemic priapism of three days' duration following chronic myeloid leukaemia in blast phase.

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Chronic myeloid leukaemia is a myeloproliferative disease characterized by the presence of philadephia chromosome BCR/ ABL fusion with translocation chromosome 9:22.

Chronic myeloid leukaemia is usually associated with marked leukocytosis in the peripheral blood and myeloid hyperplasia in the marrow as in the case of this index patient. Total white cell count being 625x 109/L at presentation.

Priapism seen in chronic myeloid leukaemia as in the case of our patient is a feature of leucostasis or hyper viscosity [9,10].

The patient was referred from the primary health care center after three days of failed repeated penile cavernosa muscle aspiration to relieve the priapism. This was rather late for a good outcome.

Ischaemic priapism is a urological emergency which must be treated early often within 48hours of presentation to prevent erectile dysfunction. Becker et al, 1985 and Morano et al 2000 documented that ischaemic priapism not given appropriate treatment within 24-48hours of onset can lead to irreversible damage and fibrosis and if lasted for 5-10 days may lead to erectile dysfunction and impotence in 35-90% of men [3,5].

Our patient already had the priapism for 72hours before diagnosis could be established and appropriate treatment of cytoreduction of the marked elevated leukocytosis could be initiated with hydroxyurea.

The patient made significant clinical improvement within four days of commencement of hydroxyurea evidenced by reduction in the total white cell count from 625 x109/L to 465 x109/L. However, despite significant gradual regression in scrotal and penile shaft oedema our patient was unable to achieve penile erection and subsequently developed depression.

The primary concern in any patient presenting with priapism should be to establish the cause which will then inform the line of management. The time interval between the onset of the priapism and the time appropriate intervention is instituted is also key to the outcome.

All these were not in favour of our patient.

The use of cytoreductive agents, chemotherapy and leukopharesis have been a better approach to the treatment of ischaemic priapism in the recent times.

However, Leukaemic priapism being a rare presentation needs more awareness and formation of standard treatment protocol to avert the undesirable irreversible damage that may be associated with this condition as in this pathetic case of our patient.

Conclusion

I presented a rare case of erectile dysfunction in a young African man in blast phase of chronic myeloid leukaemia who had priapism for three days before receiving appropriate necessary intervention.

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Competing interest: No known competing interest.

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