

Priapism as Rare Presenting Clinical Feature of Chronic Myeloid Leukemia.

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ABSTRACT

Priapism in chronic myeloid leukemia (CML) appears to be an infrequent manifestation as well as a crucial emergency. Here, we report a 45-year-old male presenting with involuntary and persistent penile erection since last 7 days. He was treated with needle aspiration of corpora cavernosa. In post operative labs he was found to have anemia and hyperleukocytosis. Patient was subsequently diagnosed with chronic myeloid leukemia.

Keywords: Priapism, chronic myeloid leukemia, penile aspiration, urology emergency. BCR-ABL.

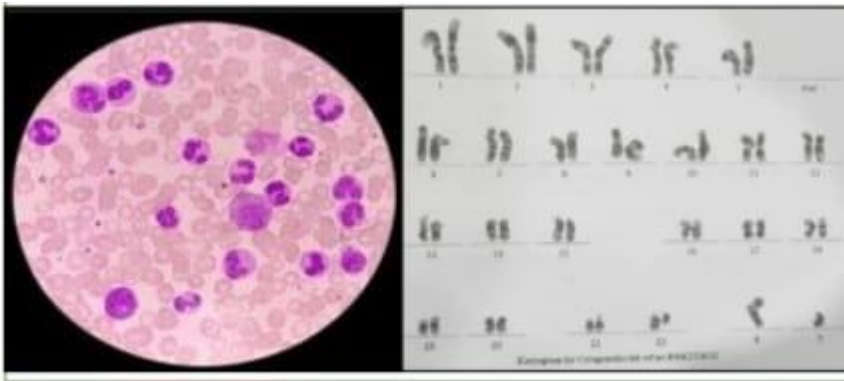
Introduction:

Priapism is an involuntary, prolonged and persistent painful penile erection without any sexual stimulation. This is an emergency condition that requires immediate management. The etiology of priapism can broadly be categorized as low flow (ischemic) and high flow (nonischemic). Hematological disorders constitute the etiology of almost 20% of cases of priapism. It is most commonly seen in sickle cell disease (40%–50% of cases) followed by leukemia (5% of cases). The most common leukemia presenting with priapism is chronic myeloid leukemia (CML). Ischemic priapism in CML patients occurs due to hyperleukocytosis resulting in hyperviscosity and venous obstruction by thrombus and microthrombus. CML is driven by the BCR/ABL1 gene that codes for a constitutively active tyrosine kinase, resulting from a balanced reciprocal translocation between chromosome 9 and 22, known as Philadelphia chromosome.

CASE REPORT:

A 45-year man presented to urology OPD with chief complaints of involuntary, prolonged, painful penile erection lasting for 4-6 hours since last 7 days. He had no previous history of trauma, intake of new medications, fever, and weight loss. He gave no history of headache, giddiness, visual disturbances and breathlessness. On examination, his vitals were stable with presence of pallor. On per-abdomen examination, he had a massive splenomegaly of 8 cm below the left costal margin. On local examination, the penis was erect, along with tenderness and superficial venous engorgement. Rest of the general physical and systemic examinations yielded no significant findings. Immediately on presentation, needle aspiration was performed, but there was no improvement. Then, shunting between the engorged corpora cavernosa and the corpus spongiosum of the glans penis was done. Routine lab parameters were sent, the

complete blood count of the patient showed a hemoglobin of 7.9gm/dl with a total leucocyte count of 3,75,900/cmm and platelet count of 125 x 10⁹/L. His peripheral smear was suggestive of left sided granulopoiesis with granulocytic precursors myelocytes 25%, metamyelocytes 20%, promyelocyte 15% and 3% blasts. Bone marrow biopsy and aspiration done which confirmed the findings of CML in the chronic phase. Bone marrow aspirate sample was sent for cytogenetics study and FISH analysis which confirmed t (9;22) (q34;q11.2) and BCR-ABL1 fusion positivity in 92% cells out of 200 cells. These findings confirmed the underlying hematological malignancy to be CML and the patient was started on hydroxyurea 500 mg TDS, Imatinib 400 mg OD along with Allopurinol 300 mg OD, and adequate hydration. Patient's WBC count started falling and by the third day, the patient became symptomatically better, and his total leucocyte count fell to 2,01,200/cmm after five days of treatment. Follow-up after one month revealed total leucocyte count of 2623/cmm, penile softening and complete improvement of priapism with no complaints of sexual dysfunction.



DISCUSSION:

Priapism is classified as low flow (ischemic) and high flow (nonischemic). Low-flow priapism involves poor muscle contraction resulting in venous congestion and is commonly caused by intracavernosal injections for impotence or erectile dysfunction. Other causes include sickle cell disease, hematological malignancy, malignant infiltration of the penis, drugs, and alcohol.^[1]

All cases of priapism. In CML, priapism is seen as the initial presentation, and usually patients have a history of recurrent episodes of self-remitting priapism (stuttering priapism) before reporting to the physician. The mechanism of priapism in CML is thought to be due to leukostasis and hyperviscosity. Therefore, such patients may also have other features of hyperviscosity in the form of hearing difficulty, breathlessness, visual blurring, or giddiness.^[2]

Unlike the other common complications of CML, priapism is more commonly seen in chronic phase than in blast crisis. Therefore, the response to first-generation TKI is optimal.^[3] The treatment modalities recommended include immediate medical management (hydroxyurea and analgesia), hydration, and TKI (imatinib). Apart from systemic treatment, it has been described that local intracavernous therapy in the form of penile aspiration, instillation of sympathomimetic drugs such as phenylephrine, and penile shunt procedures are also beneficial in bringing down the penile swelling.^[4,5]

CONCLUSION:

We conclude that although detection of priapism as the presenting manifestation of CML is rare, it is a medical emergency. Early management with local and systemic treatment prevents long term sequelae.

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