

# Priapism- A Rare Presentation of Chronic Myeloid Leukaemia

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Dear Editor,

Priapism is prolonged, painful and abnormal erection of penis unassociated with any sexual desire. If untreated, priapism may lead to penile necrosis and permanent erectile dysfunction. It is traditional to consider priapism as idiopathic and secondary. Idiopathic priapism accounts for 64% of all cases of priapism and may be due to thrombosis occurring in the venous plexus. About 20% of cases of priapism are caused by haematological disorders including sickle cell disease, leukaemia, various thromboembolic disease [1,2]. Sickle cell disease is the most common haematologic disorder causing priapism in children. The incidence of priapism in adult leukaemic patients is about 1-5 percent and leukaemia is frequently associated with painful priapism. Chronic Myeloid Leukaemia (CML) causes 50% of cases of priapism in patients with leukaemia [3,4]. In this report, we present a CML patient admitted to our hospital with priapism.

An 18-year-old male patient, with no previously known systemic disease, was admitted to emergency room with a first episode of painful, swollen and erected penis since about five days [Table/Fig-1].



**[Table/Fig-1]:** Presentation of the patient with priapism.

On physical examination, the patient had a large spleen (spleen was palpable 10 cm below the left costal margin) and no lymphadenomegaly. Total blood count showed a decreased haemoglobin 6.9 g/dL, increased leukocyte  $215 \times 10^9/L$ , (lymphocyte  $7.1 \times 10^9/L$ , neutrophil  $192 \times 10^9/L$ , monocyte  $3.8 \times 10^9/L$ , basophil  $16 \times 10^9/L$ , eosinophil  $9.1 \times 10^9/L$ ), and platelets  $470 \times 10^9/L$  count.

Peripheral blood smear revealed nearly 62% neutrophils, 8% metamyelocytes, 13% myelocytes, 5% promyelocytes, 6% basophils, 7% eosinophils and there were no blast cells. Coagulation profiles were normal and serum chemistries were unremarkable except that lactate dehydrogenase was 681 u/L. Sufficient hydration and allopurinol 300 mg/day was started for potential tumour lysis syndrome. A bone marrow aspiration and biopsy was performed from the iliac crest and the bone marrow aspiration smear demonstrated granulocytic hyperplasia with a maturation pattern. Acute leukaemia was excluded based on the bone marrow aspiration smear and flow cytometry findings. Diagnosis of CML was first suspected by identifying the typical findings in the blood and bone marrow.

Subsequently, cytoreductive therapy (hydroxyurea tablets, daily dose of 3 g) and leukopheresis were also started. At the end of five sessions of leukopheresis, leukocyte decreased to  $24 \times 10^9/L$  and the penile erection, swelling and ecchymoses were relieved. Cytogenetic analysis revealed the presence of Philadelphia chromosome and molecular studies confirmed the presence of BCR-ABL transcript, so the patient was commenced on imatinib 400 mg/day.

Hyperleukocytosis is caused by blasts of leukocytes and they are the most important cause in the pathogenesis of priapism [5]. This is usually seen in Acute Leukaemias, CML or Chronic Lymphocytic Leukaemia. In such emergencies where hyperleukocytosis is the cause, cytoreductive treatment should be started urgently with tumour lysis measures, if necessary; the patient should be treated with leukopheresis.

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