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CLINICAL AND MEDICAL CASE REPORTS
Case Report

Chronic Myeloid Leukaemia with Priapism….A Rare Finding

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Abstract

Chronic myeloid leukaemia is a myeloproliferative neoplasm of adult. As leucostasis as a result of hyperleukocytosis is commoner among children than adults. Priapism is a rare presenting feature of Chronic myeloid leukaemia. Hyperleukocytosis is considered to be the cause of priapism. Our case highlights this rare presentation of Chronic myeloid leukaemia and its management in suboptimal condition.

Keywords: *Chronic Myeloid Leukaemia; Priapism; Hyperleukocytosis*

Introduction

Chronic myeloid leukaemia is a myeloproliferative neoplasm which develops when a single, multipotent haemopoietic stem cell acquires the Philadelphia chromosome. Chronic myeloid leukaemia is mainly a disease of adult with an annual incidence of 1 case per million children in Western countries [1]. Leucostasis as a result of hyperleukocytosis is commoner among children than adults [2]. Priapism is a rare presenting feature of [3] chronic myeloid leukaemia. In chronic myeloid leukaemia, hyperleukocytosis is considered to be the cause of priapism. Priapism is clinically defined as a persistent penile erection that continues hours (4h) beyond, or is unrelated to, sexual stimulation [4]. Here we present an interesting case of Chronic myeloid leukaemia in a 15 year adolescent boy with hyperleukocytosis presenting with priapism.

Case Report

A 15 year old male presented to pediatrics department with complaints of painful erection of penis for last 48 hours. His penis was erect, painful and swollen. He did not give any history of trauma, fever, weight loss, malaise, abdominal distension, sexual intercourse before occurrence of priapism. No such episodes was noted in past. Patient did not complaint of visual blurring or headache. Physical examination revealed pallor and splenomegaly which was 15 cm below left costal margin. No evidence of jaundice or peripheral lymphadenopathy. The penis was erect and tender with superficial venous engorgement (Figure 1). Laboratory investigations revealed Hb 8.1 g/dl, TLC 462.5 X 10^9/l, platelet 361 X 10^9/l. Peripheral smear revealed normocytic normochromic anemia, nucleated RBC, increased WBC, differential cell count revealed neutrophils-47%, metamyelocytes-9%, myelocytes-36%, eosinophils-07%, basophils-01%, no blasts and promyelocytes. Serum lactic dehydrogenase was 690 u/ml. The ultrasound of penis and scrotum was suggestive of ischemic priapism and bilateral varicocele. Patient was diagnosed as a case of myeloproliferative neoplasm and suggested for BCR-ABL fusion oncogene studies for confirmation and started on treatment for hyperleukocytosis and priapism. Corporal aspiration with irrigation was done to decrease the swelling. There was incomplete relief after one procedure and he denied for further aspiration. Patient was advised for leukopheresis also but he refused for same. BCR-ABL by RTPCR was positive for major transcript. In bone marrow study, the aspirate was dry tap and

biopsy was hypercellular with marked myeloid hyperplasia with myelo poly peak, no dysplasia seen, megakaryocyte showed dwarf in morphology, no blasts seen (Figure 2). He was discharged with persistent priapism due to financial reasons. Adequate treatment in the form of Hydroxyurea, analgesics, allopurinol and dasatinib (50 mg once daily) was started. On follow up after 1 week, priapism subsided and TLC became 109 X 10^9/l. After 1 month, his CBC was with Hb 8.6 g/dl, TLC 13.9 X 10^9/l, platelet 396 X 10^9/l TLC.

Figure 1: *Shows priapism due to hyperleukocytosis.*

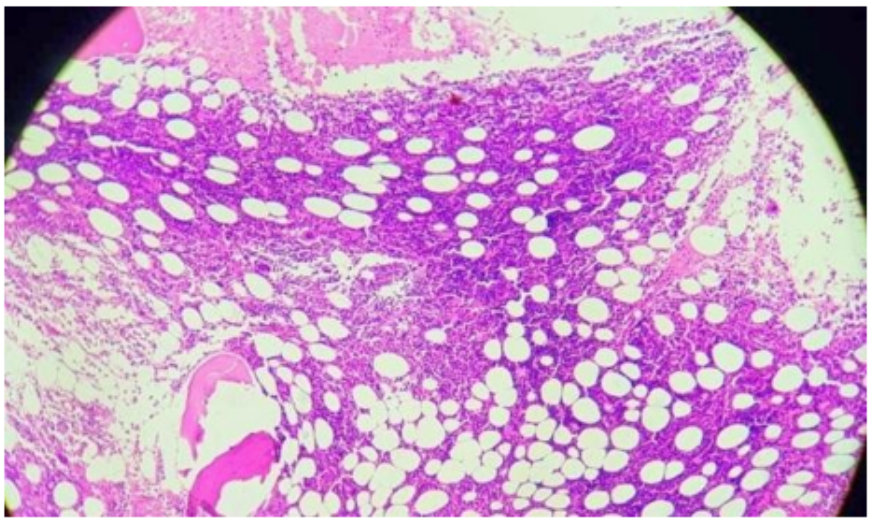

Figure 2: *Shows hypercellular marrow with increased and dwarf megakaryocytes.*

Discussion

Chronic myeloid leukaemia is the commonest adult leukemia in India with incidence of 0.6 - 2.2/100,000 population. However, it represents barely 3 percent of all childhood leukemia [5]. Literature regarding biological, clinical and molecular characteristics of Chronic myeloid leukaemia in childhood and young adults is sparse. Chronic myeloid leukaemia is often diagnosed accidentally when the patient present with an increased WBC and/or enlarged spleen. In one study of Chronic myeloid leukaemia in adolescents from AIIMS, India reported weakness, fever, stress due to splenomegaly, bleeding as most common presenting symptom [6].

Once in a while, patient may present with leukostasis complication due to hyperleukocytosis such as bleeding, hearing loss, priapism and visual blurring [7].

In the era of chronic myeloid leukaemia treatment, imatinib and other tyrosine kinase inhibitor (TKI) discovery was a breakthrough invention. Imatinib, nilotinib and dasatinib are TKI which are approved in first line therapy for chronic myeloid leukaemia.

The subject of adult and young oncology 15 - 29 years has been focus of attention in the last decade. Diverse issues revolve around AYA oncology. One of them deals with distinct pharmacokinetics and drug metabolism in this population [8,9]. Lack of adherence can occur in 27 - 60% of patients and is another obstacle for satisfactory results [10]. Financial burden is a very important facet of disease. Even a routine clinic visit may have monetary or logistical implication. Our patient refused leukapheresis on the basis of financial and psychosocial ground. TKI was procured via government sponsored health insurance scheme and thus continuous supply was assured.

Hyperleukocytosis is defined as > 50 - 100 X 10^9/l. Acute and chronic leukemia both can manifest as hyperleukocytosis [8]. It leads to leukostasis which in turn can cause hearing loss, visual blurring, headache, papilledema, intracranial hemorrhage, respiratory distress, acute renal failure and priapism. Treatment of hyperleukocytosis mandates management of primary disease, hydroxyurea and leukapheresis [11,12].

The condition priapism was named after the Greek god Priapus. It can be idiopathic or secondary to many medical conditions as shown in table 1. Priapism is an unusual presentation in male patients with Chronic myeloid leukaemia, particularly at this young age [8,11-14]. It is an emergency and needs multidisciplinary approach. The pathophysiology of priapism in Chronic myeloid leukaemia states following mechanisms. 1) Venous congestion of the corpora cavernosa due to mechanical pressure from splenomegaly, 2) sludging of leukemia cells in the corpora cavernosa and dorsal vein of penis, 3) local infiltration of sacral nerves, 4) CNS infiltration, 5) Hyper viscosity leading to vascular obstruction [10-14].

Primary (idiopathic)
Secondary
Hematological Hemoglobinopathies-sickle cell disease that is common in children
Hyperleukocytosis related - Acute leukemia, chronic leukemia
Hyperviscosity - polycythemia vera, multiple myeloma, Waldenstorm Macroglobulinemia
Neurologic-spinal shock
Malignancy-infiltration by cancer
Trauma-perineal, penile, pelvic
Drugs-anticoagulants, PDE5 inhibitors, cocaine, alpha blockers
Infection-Mumps orchitis, malaria
Metabolic disorders-Gout, Total parenteral nutrition, hemodialysis, amyloidosis

Table 1

Presently, no standard guidelines exist for managing these types of patients. The previous literature mentions use of cytoreductive therapy, leukapheresis, local aspiration and irrigation, intracavernous injection of sympathomimetics [15], surgical shunts and oral terbutaline [16].

Aforementioned patient was treated with cytoreduction, aspiration and TKI. This led to gradual reversal of priapism and hopefully no erectile dysfunction. Our case highlights the rare presentation of Chronic myeloid leukaemia and its management in suboptimal condition.

Conclusion

Any patient presenting with priapism must undergo detailed history, comprehensive physical examination, and suitable laboratory test. Physician at primary/secondary level should be apprised of this condition to be related to hematological disorders with prompt institution of supportive medical therapy and ensure timely referral. Almost half of them run the risk of erectile dysfunction [3], which hopefully can be averted by appropriate therapy. Our case highlights the rare presentation of Chronic myeloid leukaemia and its management in suboptimal condition and we performed clinicopathological correlation to reach the right diagnosis of patient.

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