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SHORT COMMUNICATION

PRIAPISM – AN UNUSUAL PRESENTATION IN CHRONIC MYELOID LEUKAEMIA: CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT
Chronic Myeloid Leukaemia (CML) counts for around 15% of haematological malignancies. We are reporting two cases in which initial presentation of the disease was with priapism. After initial investigative profile, the priapism was relieved by aspiration and decompression. Later investigations revealed hyperleukocytosis due to CML in both the cases. These cases demonstrate the importance of identifying the underlying cause of priapism, as it directly influences both initial and ongoing management.

Key Words: *Priapism, Chronic Myeloid Leukemia.*

INTRODUCTION

Priapism is persistent, prolonged and painful abnormal erection of the penis without accompanied sexual arousal (longer than 6 hours). The word 'Priapism' is related to 'Priapus' the Greek and Roman God of procreation whose symbol was an erect phallus.[1]

Traditionally priapism is either idiopathic or secondary. Idiopathic priapism is most common and may be due to thrombosis occurring in the venous plexus. Less commonly, priapism may be secondary to various disorders including sickle cell anaemia, trauma, leukaemia, cancerous invasion of the penis, drugs, alcoholic ingestion and various thromboembolic diseases.[2] About 20 percent cases of priapism are related to haematological disorders[3]. The incidence of priapism in adult leukaemic patients is about 1-5 percent and leukaemia is frequently associated with painful priapism.[3] The following cases illustrate priapism as an unusual presenting symptom of CML.

Priapism can also be categorised as low flow (ischaemic) or high flow (non ischemic). High flow priapism can be secondary to trauma to penile area. Priapism is an emergency situation. It is important to start appropriate therapy soon after the onset so as to have a reasonable chance of preserving potency.[4]

CASE REPORT 1

A previously healthy 21-year-old male was referred from local hospital for treatment of painful penile erection for the last 8 hours. There was no history of trauma, malaise, night sweats, joint pain, and cough. However, he gave history of weight loss of about 9 Kg and superficial bleeding tendencies (epistaxis and bruises) during the last 3 months. The vital signs revealed a body temperature of 38.2 C, pulse 109 beats / minute, blood pressure 137/64 mmHg and respiration rate of 21/minute. He was alert, anxious and oriented. The physical examination revealed that the liver was palpable 6 cm below the right costal margin, and spleen was 7 cm below the left costal margin. His conjunctiva was pale but did show no jaundice. The penis was erect, firm, and tender with superficial venous engorgement.

Laboratory data revealed haemoglobin (Hb) 8.3 g/dl, haematocrit 25.7%, white blood count (WBC) 316×10^9/L, with Platelet 670×10^9/L. Serum chemistry was unremarkable. Treatment of priapism was initiated and performed by cavernosa aspiration and epinephrine irrigation in the emergency minor operation theater. For hyperleukocytosis, he was admitted to the intensive care unit and was diagnosed as a case of CML on the basis of peripheral blood smear and bone marrow examination The Philadelphia chromosome (t(9:22) p210) was illustrated in the patient (Fig. 1). He was started on Hydroxyruea tablets at 4 grams per day. Allopurinol 300 mg daily with adequate hydration was also started for potential tumour lysis syndrome. Before discharge on 3rd day, his WBC dropped to $82x10^9$/ l and haemoglobin (Hb) was raised to 9.7 g/dl. No further episode of Priapism occurred during his admission period. The patient continues to report to us without any erectile disorder till date.

CASE REPORT 2

A 55-year-old man reported to our hospital with the complaints of persistent and painful erection for the last 12 hours. There was no history of recent inter-

course, trauma and use of drugs etc. The patient had no known medical problem previously.

On examination, the patient was moderately built and well nourished. He was anaemic but was not jaundiced and had no lymphadenopathy or sternal tenderness. Pulse was 98/min, blood pressure: 120/82 mm Hg. His spleen was palpable 7 cm below the left costal margin. Other systems revealed no abnormality on clinical examination. Local examination revealed an erect, swollen, engorged and tender penis with engorged veins on its surface.

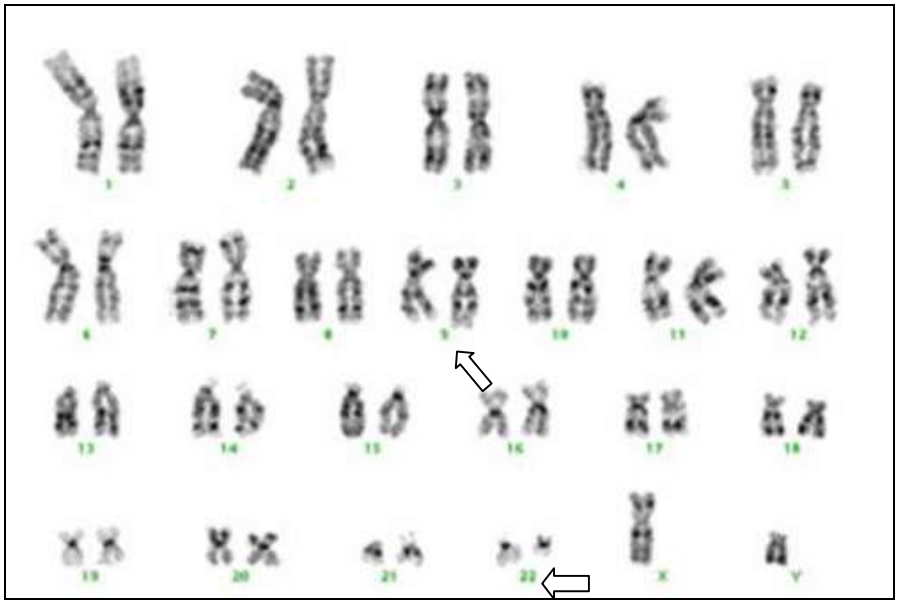

Fig 1: *Karoytypic arrangement showing translocation between chromosome 9 and 22.*

Immediate laboratory investigations revealed Hb 9 g/dl, haematocrit 27.8%, with the total leukocyte count (TL-C) 282×10^9/L. Platelet 260 × 10^9/L. All other initial investigations were within normal range.

In view of urgency of the situation, the patient was treated with simple wide bore needle aspiration of the corpora cavernosa at the shaft of the penis. About 70 ml of dark blood was aspirated and the priapism was relieved.

Keeping in view the clinical presentation, he was admitted and following battery of tests was performed, complete blood count (CBC), complete biochemical profile, neutrophil alkaline phosphatase staining of the peripheral blood. Bone marrow aspiration and trephine biopsy, the Philadelphia chromosome (t(9:22)) was observed on cytogenetic studies and confirmed by the FISH studies on the bone marrow aspirate samples.

Patient was diagnosed as a case of CML based on basis of investigations and was started on Hydroxyruea tablets at 3.0 gram/day and allopurinol 300 mg daily while adequate hydration was also started for potential tumour lysis syndrome. Dose of Hydroxyruea was gradually increased to 4.0 gram per day. Patient responded well and with in three days, his peripheral counts dropped to 28.6 × 10^9/I. No episode of priapism occurred during his stay in the hospital. Chemotherapy was readjusted accordingly and patient was allowed to leave the hospital with advice of regular checkup in out doors.

DISCUSSION

CML accounts for 15–20% of adult leukaemias.[5] Most common presenting feature of CML is raised white cell count i.e. hyperleuckocytosis. This disorder refers to a WBC count 100 × 10^9/l or more.[6]

Hyperleukocytosis is convincingly considered to be the cause of priapism in patients with leukemia. Different mechanisms are thought to be operative: Most commonly agreed process is the aggregation of leukaemic cells in the corpora cavernosa and the dorsal veins of penis or venous congestion of the corpora cavernosa resulting from mechanical pressure on the abdominal veins by the splenomegaly.[6] Alternately hypothesis is of infiltration of the sacral nerves with leukaemic cells or infiltration of the central nerve system.[6,7]

Priapism is a pathological condition characterized by penile erection that persists for longer than six hours and is unrelated to sexual stimulation. This condition is exclusive to men and typically involves the paired corpora cavernosa.[9]

Approximately 20% of priapism cases are related to haematological disorders, and the incidence of priapism in adult patients with leukemia is about 1%–5%.[9] Priapism is traditionally defined as either low-flow (ischaemic) or high-flow (non-ischaemic). Low-flow or ischaemic priapism results from pathologically decreased penile venous out-flow that results in intracavernosal stasis. It manifests mostly in a painful, rigid erection. This type is more common

and represents an actual emergency because of irreversible cellular damage and fibrosis that occur if treatment is not administered within 24 to 48 hours.[10]

The causes of low-flow priapism include idiopathic, haematological disorders, tumour infiltrate or drug induced.[11] High-flow or arterial priapism differs in that it results from increased arterial inflow into the cavernosal sinusoids, which overwhelms venous outflow and clinical presentation is painless. In contrast to low-flow priapism, intracavernosal blood sampling from patients with high-flow priapism reveals bright red oxygenated blood and thus irreversible cellular damage and fibrosis are rare.[12] This type of priapism is usually due to penis or perineum trauma that results in injury to the internal pudendal artery. This establishes a fistula between the cavernosal artery and the corpus cavernosum so that unregulated inflow occurs. It is not an actual emergency in patients with high-flow priapism, and treatment can be on an elective basis.[13]

About the management of priapism, there have been many methods described in the literature. Spinal anaesthesia, ice water enema, ice packs, radiotherapy,[14] fibrinolytic agents and anticoagulants have been tried but no significant success rate after releaved obtained.[15] The painful erection of the patient, immediate aspiration and irrigation of the corpora cavernosa as well as injection of α-adrenergic agents is recommended.[16]

It is well know that a higher number of leucocytes in hyperleukocytic syndrome leads to the formation of leukocyte aggregates and thrombi which further result in occlusion of small vessels[17]. Many studies have shown that a leukocyte count greater than 100.0×10^9/L or more is a major contributor of an elevation of the whole-blood viscosity.[18] In our cases, the leukocyte count on admission was 316.0×10^9/L and 288×10^9/L. Thus, it is assumed that leukocyte aggregates and/or thrombi formed may have resulted in leukocyte aggregates which initiated the sequences leading to priapism.

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