CASE REPORT

A rare presentation of chronic myeloid leukaemia with priapism treated with corporoglandular shunting

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SUMMARY

Priapism is a rare clinical presentation of a patient with chronic myeloid leukaemia (CML). Herein, we present a young Nepalese man that presented to the emergency department with an acute and painful penile erection for two days. Clinically, he was pale and abdominal examination revealed hepatomegaly. Combined oncologic and initial urological intervention with carvernosal aspiration and intracavernosal phenylephrine failed to achieve detumescence. The patient underwent an emergency corporoglandular shunting eventually. In this case report, we discuss the management compared with previously reported cases.

INTRODUCTION

The clinical manifestation of chronic myelogenous leukaemia (CML) is insidious in nature. The disease can present with incidental elevated white blood cell (WBC), nonspecific symptoms of fever, fatigue or weight loss. Priapism is a rare presentation of this entity. The pathogenesis is related to hyperviscosity and leucostasis due to hyperleucocytosis. The treatment for such crises often involves oncologic and urology intervention. Due to the rarity of the manifestation of priapism in CML patients, the management remains controversial. Majority of the authors advocate CML-specific therapy (chemotherapy) alone, whereas some advocate additional urological intervention. In this report, we describe a man presented with priapism requiring surgical shunting. He had brief period of detumescence following carvenosal aspiration and Intracavernosal phenylephrine. The role of CML-specific therapy,particularly in leukapheresis is reemphasized.

CASE REPORT

A 28-year-old Nepalese man whom was previously healthy, was referred to our regional urology department for acute painful penile erection for two days from a local district hospital. There was no history of trauma, fever, night sweats or joint pain. Physical examination revealed pallor of the conjunctiva with normal sclera. He had hepatomegaly which was palpable 2cm below the right costal margin and splenomegaly. The penis was erected, firm and tender (Figure 1).

Laboratory investigation revealed haemoglobin (Hb) was 6.6 g/dl [13.0-18.0], haematocrit was 20.7% [40-54%], white blood count (WBC) was 294.1x10^9/L [4.0 -11.0] and platelet was 94x10^9/L [150-400]. Peripheral blood film revealed hyperleucocytosis with blast cell and abnormal WBC seen. (Blast is moderate to large in size, moderate to scanty cytoplasm, round nucleus and prominent nucleoli). The differential counts revealed 6% of blast cells, 26% of promyelocyte and 20% of myelocyte. Urgent referral was made to haematologist with subsequent diagnosis of CML. Tablet hydroxyurea, allopurinol and intravenous Cytarabine were initiated due to the diagnosis of chronic myeloid leukaemia (CML).

Emergency intracavernosal aspiration and phenylephrine irrigation was performed. After aspiration of 750ml of blood, there was a brief period of detumescent, but the erection re-occurred hours later. Penile arterial blood gas revealed a low flow type priapism with presence of acidosis [pH 7.13, pCO2 65mmHg, pO2 30mmHg, HCO3- 18.6mmol/L, Base deficit -7.6mmol/L]. An emergency corporoglandular shunting was performed under spinal anaesthesia.

Intraoperatively, a Foley's catheter was inserted draining clear urine. Stab incisions were made with a size 11 blade laterally over the glans penis at both sides of penile meatus (Figure 2a). The incisions were deepened into the corpora body allowing shunting of blood to flow into the glans. Approximately 200ml of dark colour blood was drained before it turned bright red. Following drainage, the penis turned flaccid. The stab incisions were approximated with Polyglactin 3/0 to prevent excessive bleeding (Figure 2b). In total, only four pints of pack cells were transfused peri-operatively to prevent hyper-viscosity and recurrence of priapism. Postoperatively, intravenous hydration and Cytarabine were continued. There was a reduction of white blood cell count to 14.1x10^9/L at post-operative day-6 of Cytarabine.

DISCUSSION

Priapism in CML patient is described as early in 1974. It occurs in 1–2% of CML male patients, with a bimodal age distribution of 5–10 and 20–50 years old. To our knowledge, less than 20 cases were published describing priapism as a complication of CML. Shaeer et al., reported that one third of these conditions require shunting procedure following failed initial cavernosal aspiration and phenylephrine injection.[1] Similarly, in our patient these initial measures did not resolve the erection which led to a surgical shunt in this patient.

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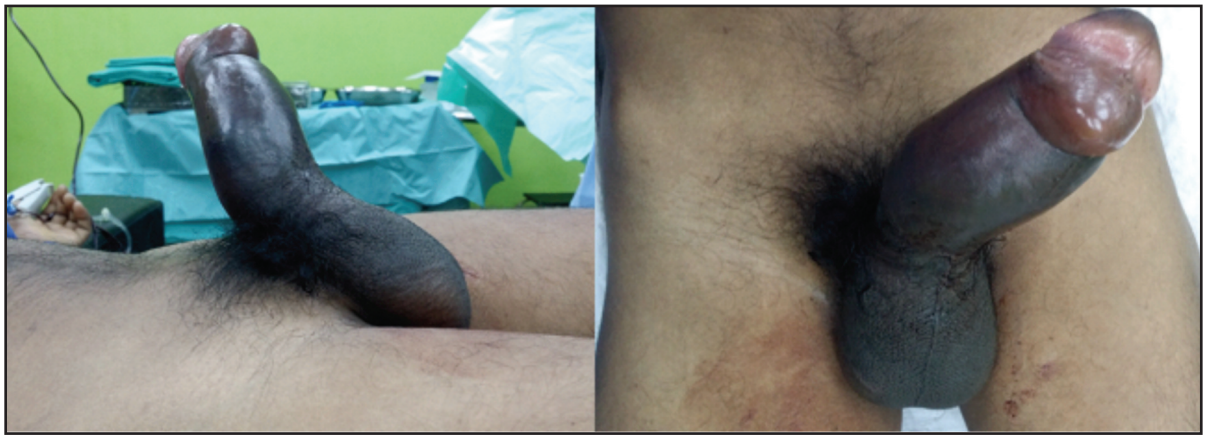

Fig. 1: Clinical picture of priapism before surgical shunting.

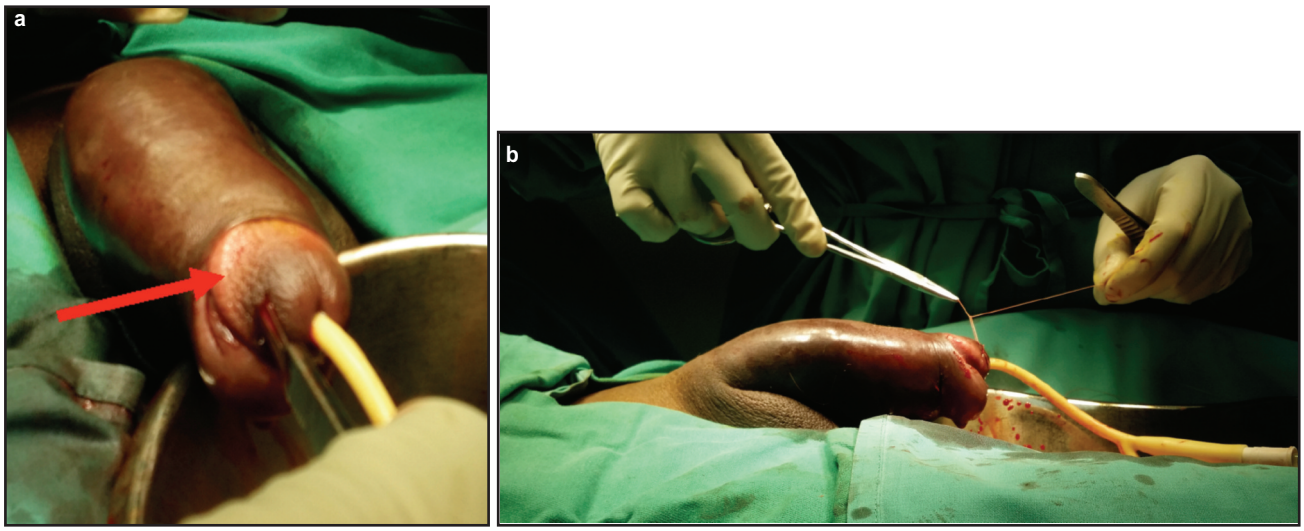

Fig. 2: Intraoperative picture showing corporoglandular shunting procedure.
(a) Stab incision made with size 11 scalpel at the glans (red arrow).
(b) Stab incision closed with absorbable stitches. Note the detumescence achieved following the shunting.

The priapism recurs despite a large amount of cavernosal blood aspiration primarily due to hyperleucocytosis. Despite best medical efforts by intravenous hydration with the administration of Cytarabine and Hydroxyurea, it took 4 days to reduce the WBC to less than 50 x10^9/L. Leukapheresis may be beneficial in such case. There were few cases reported successful detumescence following leukapheresis without needing a surgical shunt. Veljković D et al., reported the use of leukapheresis for one cycle on three consecutive days, Ponniah A et al., reported seven sessions within a day and Ergenc H et al reported single three hours session successfully achieve detumescence.[2-4] These patients' WBC were above 300 x10^9/L and leukapheresis were able to reduce the WBC to below 100x10^9/L (or 30-60% WBC reduction) after a single session. Due to unavailability of leukapheresis in our centre, the oncologic treatments in our patient were limited to chemotherapy and hydration.

Another practical aspect to be highlighted in the case is balanced transfusion of red blood cells. The target haemoglobin was kept at only 7g/dl. This is to reduce the risk of recurrent priapism with over transfusion.[1]

The clinical presentations of priapism with anaemia would suggest the differential diagnoses of sickle cell anaemia, glucose-6-phosphate dehydrogenase (G6PD) deficiency and thalassemia. However, 90% of patients with sickle cell anaemia usually present with priapism below the age of 20-year-old, and the disease is uncommon in Malaysia. Thalassemia usually presents with a history of multiple blood transfusion and clinical findings of hepatosplenomegaly which was not present in this patient that makes it an unlikely cause of priapism. A full blood picture with peripheral blood film excluded these differential diagnoses. The presence of hyperleucoystosis with the presence of blast cells led to the diagnosis of chronic myeloid leukaemia.

In conclusion, treatment of priapism in a patient with CML requires multi-disciplinary efforts from haematology-oncology and urology services. Reduction of WBC with chemotherapy, hydration and leukapheresis to decrease blood viscosity are the main oncologic treatment principle.

INFORMED CONSENT

Written informed consent was obtained from patient who participated in this study.

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