

## **Case study**

### **Priapism as the first manifestation of Chronic Myeloid Leukemia: a rare occurrence**

**Abstract:** Priapism is an involuntary, painful erection of the penis for more than 4 hours without any sexual stimulation or trauma. It is a urological emergency to avoid erectile dysfunction. We report a case of a young adult who presented to emergency with priapism and was eventually diagnosed to have chronic myeloid leukemia (CML). The priapism did not resolve with conservative measures. He, underwent a proximal Sacher's shunt surgery to reduce the priapism. Immediate initiation of hydration, oral administration of hydroxyurea, surgical correction and prompt addition of tyrosine kinase inhibitors (TKI) after an early diagnosis by FISH method is the key to managing such patients to reduce the consequences of erectile dysfunction.

**Key words:** priapism, chronic myeloid leukemia, erectile dysfunction

#### **Introduction:**

Priapism is an involuntary, painful penile erection that persists for > 4 hours and is unrelated to sexual interest or stimulation. It is classified into ischemic (low flow), arterial (high flow), or stuttering (recurrent or intermittent) (1, 2, 3). It is a urological emergency with an immediate goal of preventing erectile dysfunction(2). It can occur at any age, with an overall incidence of 1.5 in 1, 00,000 patients(1). Priapism can occur due to a wide variety of causes which commonly include infection, malignancy, CNS mediated, drug-induced or hemoglobinopathies (1). Almost 20% cases of priapism are due to an underlying hematological disorder of which chronic myeloid leukemia (CML) is one of the commonest (1, 4, 5). Priapism as a presenting feature of CML is

very rare and occurs in only in 1-2% cases (5). We report a case of a young adult who presented with priapism and eventually diagnosed as CML who was managed in a multidisciplinary manner.

**Case report:**

A 37-year-old gentleman presented to the emergency department with the painful erection of penis unrelated to sexual stimulation, drugs or trauma. The patient had experienced a similar episode six months ago, which had subsided spontaneously within two hours. There was a history of weight loss of around 10kgs in the last 3 years. There was no history of fever, loss of appetite or breathlessness or hyperviscosity symptoms. On examination, the penis was firm, erect and tender. The spleen was palpable 10 cm below left costal margin. There was no hepatomegaly or lymphadenopathy.

Initial Conservative measures such as corporal aspiration with cold saline irrigation and phenylephrine injection and distal caverno-glandular shunt (winter shunt) were ineffective to achieve detumescence.. Eventually, he underwent an emergency proximal Sacher's open shunt between the proximal corpora spongiosum and corpora cavernosum, which resolved the priapism (Figure 1). His complete blood count (CBC) showed hemoglobin of 13.2 g/dl, white blood cell (WBC) count of  $2,52,300 /\text{mm}^3$  and platelet count of  $3,54,000 /\text{mm}^3$ . Peripheral smear showed a neutrophilic shift to the left with 7% blasts and increased eosinophil and basophils (Figure 2a). Lactate dehydrogenase, renal and liver function tests, were within the normal range. . Fluorescent in situ hybridization (FISH) for BCR: ABL1 fusion on peripheral blood was positive in 97% cells confirming the diagnosis of CML (Figure 2b). Bone marrow examination was suggestive of chronic myeloid leukemia in chronic phase (Figure 2c, 2d).

Conventional karyotyping did not show any additional cytogenetic abnormality. RT-PCR for BCR: ABL showed p210 transcript.

The option of leukapheresis was not considered as the patient had to be taken up for surgery due to prolonged tumescence. e. He was started on Imatinib 400mg once a day, besides continuing hydroxyurea and allopurinol. With the normalization of WBC counts, the hydroxyurea was tapered and stopped in the following week, while continuing Imatinib at the same dose. On the last follow-up, his hemoglobin was 10.3 g/dl, and white blood count was  $5400/\text{mm}^3$  with an absolute neutrophil count of  $3300/\text{mm}^3$  and platelet count was  $2,10,000/\text{mm}^3$ . Quantitative molecular analysis showed optimal response with BCR: ABL (IS) ratio of  $< 0.45\%$  at 6 months from the time of diagnosis. The patient is on regular follow-up, has an active lifestyle, but able to achieve a partial erection. n.

### **Discussion:**

Priapism is an andrological emergency with an impotence risk of 50%. (2) The median age of presentation in a case series of 16 patients was 36 years (range: 21 to 56 years). The commonest cause was secondary to neuroleptics in 6 cases (37%), chronic myeloid leukaemia in 2 cases, sickle-cell anaemia in one case, radiotherapy in 1 case and the cause remained unknown in 6 cases (37%) (6).

The first description of priapism was noted in the year 1845 (7). The term “priapism” is from Priapus (ancient Greek god of fertility) (7). The pathophysiology depends on the aetiology of priapism which can be ischemic or non-ischemic type (8). Ischemic priapism behaves as compartment syndrome wherein there is stasis of blood within corpora leading to erect penis. Pain occurs due to tissue ischemia and muscle hypoxia. Ischemic priapism accounts for almost 95% of all cases (Table 1) (9). In ischemic priapism, it has been hypothesized that

tissue hypoxia leads to changes in nitric oxide (NO) synthase, which in turn causes reduced NO production and leads to pain (2, 9). Another hypothesis ischemic priapism is the infiltration of sacral nerves or CNS by leukemic cells (2)

Although patients with CML is one of the most frequent haematological causes of ischemic priapism, priapism as a presenting feature in CML is rare. A high WBC count with peripheral smear showing left shift, eosinophilia and basophilia along with splenomegaly should point towards a diagnosis of CML. CML may present in either chronic, accelerated or blast phase. Once suspected, the patient should be started immediately on hydration and allopurinol. Initial management is multidisciplinary with involvement of emergency medicine, haematologist, urologist, haemato-pathologist and transfusion medicine specialist. Due to high counts, it can present with symptoms of hyperviscosity (blurring of vision, respiratory discomfort or end-organ damage) (1, 2, 10).

Urgent leukapheresis is indicated in such patients whenever the facility is available and surgical intervention is not an emergency. (1, 10) Surgical approach is a therapeutic aspiration or corpora-glandular shunt surgery Proximal shunt surgery is offered for severe distal penile edema and tissue damage. Erectile dysfunction (ED) is a major complication seen in 90% cases where priapism persists for more than 24 hours (2, 5). In case, the surgical options fail, insertion of penile prosthesis may be offered before intra-corporal fibrosis ensues American Urological Association guidelines for the management of priapism reiterates the importance of urgent surgical intervention for ischemic priapism in addition to medical management (2).

Medical management of CML consists of hydroxyurea till WBC count normalizes along with tyrosine kinase inhibitors (TKI) like Imatinib, Dasatinib or Nilotinib (10)..

Response a monitoring is done by 3 monthly RQ-PCR for BCR: ABL as per the recommended guidelines with life long continuation of TKI (10).

**Conclusion:**

This case highlights the importance of evaluating the causes of priapism systematically. The patient had ignored his first episode 6 months ago, which had lasted for only 2 hours and did not seek medical attention. Initial management of priapism is a challenge to prevent long term erectile dysfunction s. High index of suspicion for CML and complete blood count along with peripheral smear evaluation at presentation will help rule out hematological cause for priapism. Managing the patient alongside the Urologist is pivotal in initial management of such cases.

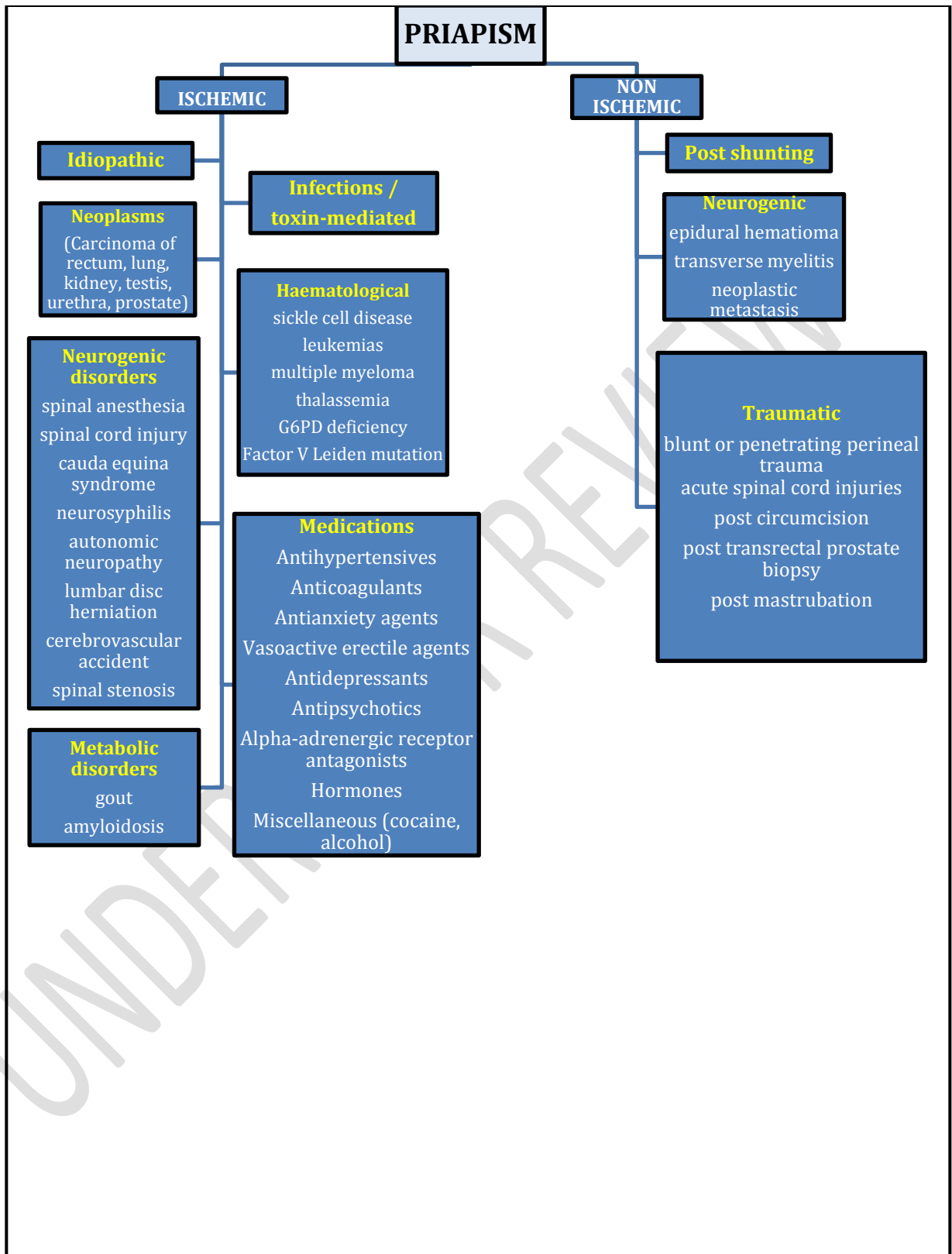
**Consent:** Signed consent was obtained from the patient and kept in his medical records.

**References:**

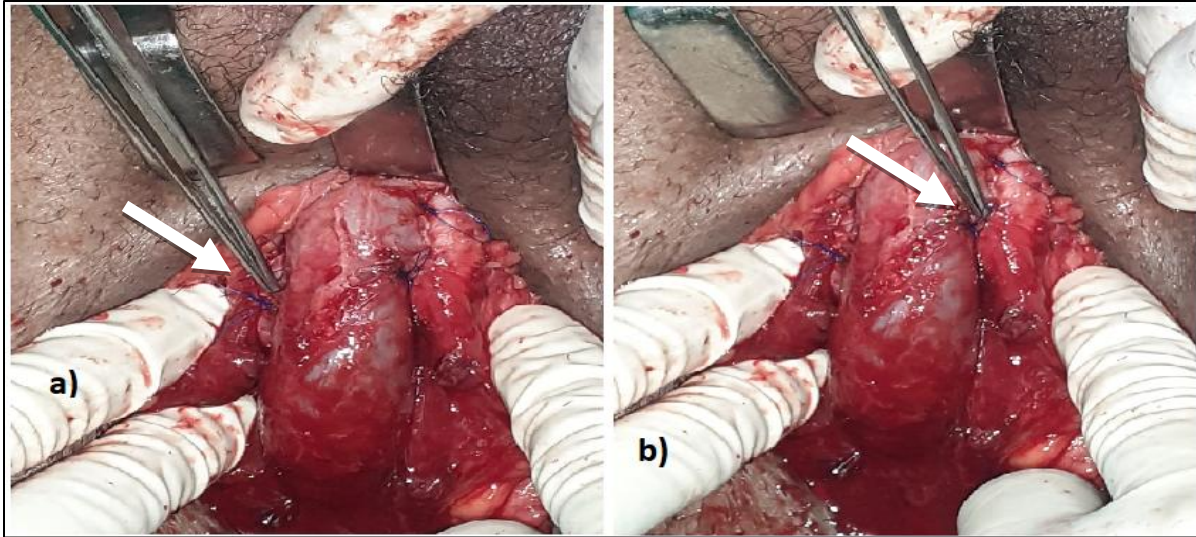
1. Rodgers R, Latif Z, Copland M. How I manage priapism in chronic myeloid leukaemia patients. *Br J Haematol.* 2012 Jul;158(2):155–64.
2. Montague DK, Jarow J, Broderick GA, Dmochowski RR, Heaton JPW, Lue TF, et al. American Urological Association guideline on the management of priapism. *J Urol.* 2003 Oct;170(4 Pt 1):1318–24.
3. Salonia A, Eardley I, Giuliano F, Hatzichristou D, Moncada I, Vardi Y, et al. European Association of Urology guidelines on priapism. *Eur Urol.* 2014 Feb;65(2):480–9.
4. Dhar J, Dhar J, Chhabra G, Khandelwal L, Khandelwal L, Batra A, et al. Priapism as a Debut Presentation of Chronic Myeloid Leukemia. *J Coll Physicians Surg Pak.* 2019 Jan; 29(1): 78–80.
5. Minckler MR, Conser E, Figueroa JJ, Scott AJ, Gaither J, Amini R. The Semantics of Priapism and the First Sign of Chronic Myeloid Leukemia. *Case Reports in Emergency*

- Medicine. 2017 May 30; 2017.
6. Benchekroun A, Lachkar A, Soumana A, Farih MH, Belahnech Z, Marzouk M, et al. [Priapism in adults. 16 cases]. *Ann Urol (Paris)*. 1998; 32(2): 103–6.
  7. Musa AU, Ndakotsu MA, Abubakar SB, Agwu PN. Chronic myeloid leukemia with an initial presentation as ischemic priapism: A case report and review of literature. *Archives of International Surgery*. 2017 Apr 1; 7(2): 68.
  8. Jameel TA, Mehmood KH. Priapism—an unusual presentation in chronic myeloid leukaemia: case report and review of the literature. *Biomedica*. 2009 Jul; 25: 197-9.
  9. Farhan S, Anjum F, Al-Qahtani FS, Al-Anazi KA. Chronic myeloid leukemia presenting with priapism. *Journal of Leukemia*. 2014 Dec 23.
  10. Hochhaus A, Saussele S, Rosti G, Mahon FX, Janssen JJ, Hjorth-Hansen H, Richter J, Buske C, ESMO Guidelines Committee. Chronic myeloid leukaemia: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of Oncology*. 2017 Jul 1; 28 (suppl\_4) : iv41-51.

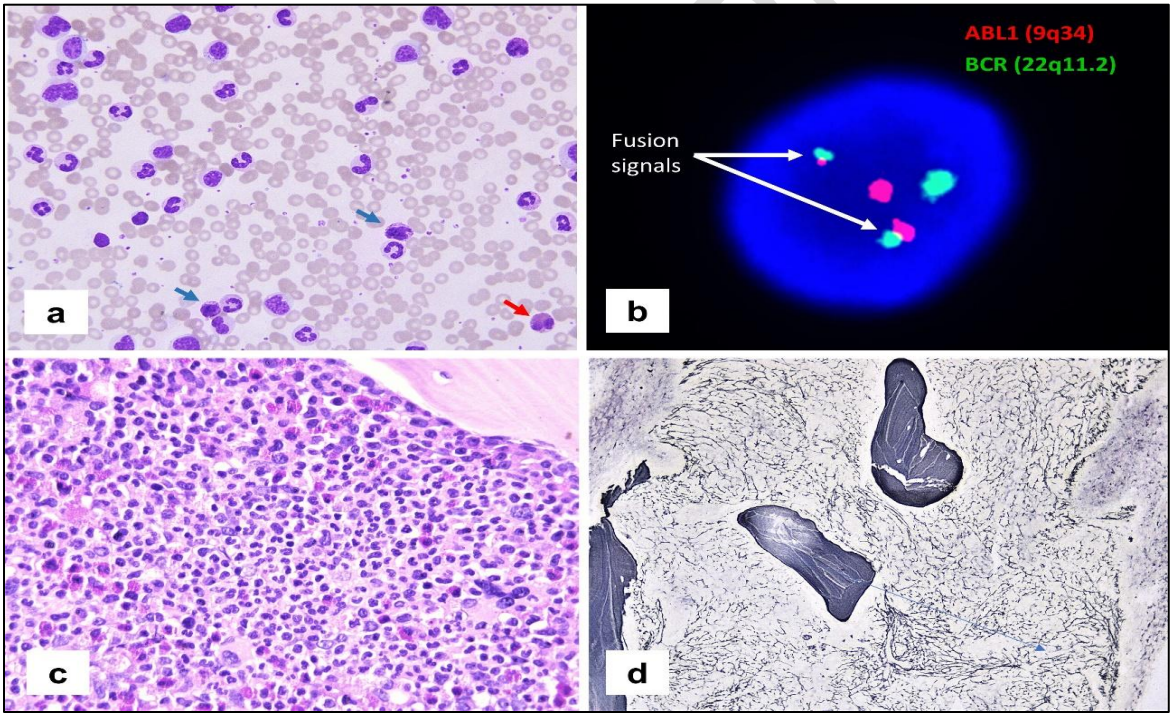
**Table 1: Causes of ischemic priapism:**



**Figures:**



**Figure 1:** Intra-operative images showing a) corpora-cavernosal anastomosis on left  
b) corpora-cavernosal anastomosis on right



**Figure 2:** a) Peripheral blood film showing left shift, eosinophils (red arrow) and basophils (blue arrows); Leishman stain x 400 b) FISH for BCR-ABL1 showing two fusion signals using dual colour dual fusion probes x 1000 c) Solidly cellular trephine biopsy showing myeloid predominance with increased eosinophil precursors; Hematoxylin & Eosin stain x 400 d) Reticulin stain on the trephine biopsy showing increased fibrosis (2+) x 100

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