








## CASE REPORT

# REVISED Case Report: Priapism as The Clinical Presenting Feature of Chronic Myeloid Leukemia: Case Report and 20-Year Literature Review [version 2; peer review: 2 approved]

Previously titled: 'Case Report: Priapism as The Clinical Presenting Feature of Chronic Myeloid Leukemia: Case Report and 20-year Literature Review'

Siprianus Ugroseno Yudho Bintoro <sup>1,2</sup>, Pradana Zaky Romadhon <sup>1,2</sup>, Satriyo Dwi Suryantoro <sup>1,3</sup>, Rusdi Zakki Aminy <sup>1</sup>, Choirina Windradi <sup>3</sup>, Krisnina Nurul Widiyastuti<sup>1</sup>

<sup>1</sup>Department of Internal Medicine, Airlangga University, Faculty of Medicine, Surabaya, East Java, 60131, Indonesia

<sup>2</sup>General Teaching Hospital Dr. Soetomo, Surabaya, East Java, 60286, Indonesia

<sup>3</sup>Universitas Airlangga Hospital, Surabaya, East Java, 60115, Indonesia

**V2** First published: 15 Jul 2021, 10:571  
<https://doi.org/10.12688/f1000research.53365.1>  
 Latest published: 13 Jan 2022, 10:571  
<https://doi.org/10.12688/f1000research.53365.2>

## Abstract





Priapism in chronic myeloid leukemia (CML) appears to be an infrequent manifestation as well as a crucial emergency. Here, we report an 18-year-old male presenting with a persistent erection of the penis for 20 days. We evaluated and compared the reported cases within 20 years discussing the management of priapism in CML. Cytoreductive therapy followed by leukapheresis, the administration of tyrosine kinase inhibitor, and intra-cavernosal blood aspiration may resolve the symptoms of priapism. Early intervention for cytoreduction and aspiration are the pivotal keys to successfully impeding the complications.


## Keywords

priapism, chronic myeloid leukemia, cytoreduction, penile-aspiration, cancer

## Open Peer Review

Reviewer Status  

	Invited Reviewers	
	1	2
<b>version 2</b> (revision) 13 Jan 2022		 report
<b>version 1</b> 15 Jul 2021	 report	  report

1. **Wulyo Rajabto**, University of Indonesia, Jakarta, Indonesia
2. **Ritu Gupta** , All India Institute of Medical Sciences (AIIMS), New Delhi, New Delhi, India

Any reports and responses or comments on the article can be found at the end of the article.

**Corresponding author:** Pradana Zaky Romadhon ([zaky.romadhon@fk.unair.ac.id](mailto:zaky.romadhon@fk.unair.ac.id))

**Author roles:** **Bintoro SUY:** Conceptualization, Formal Analysis, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; **Romadhon PZ:** Conceptualization, Data Curation, Formal Analysis, Investigation, Methodology, Resources, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; **Suryantoro SD:** Conceptualization, Formal Analysis, Methodology, Resources, Software, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; **Aminy RZ:** Conceptualization, Methodology, Project Administration, Writing – Original Draft Preparation, Writing – Review & Editing; **Windradi C:** Conceptualization, Formal Analysis, Investigation, Methodology, Project Administration, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; **Widiyastuti KN:** Conceptualization, Data Curation, Formal Analysis, Project Administration, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing

**Competing interests:** No competing interests were disclosed.

**Grant information:** The author(s) declared that no grants were involved in supporting this work.

**Copyright:** © 2022 Bintoro SUY *et al.* This is an open access article distributed under the terms of the [Creative Commons Attribution License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

**How to cite this article:** Bintoro SUY, Romadhon PZ, Suryantoro SD *et al.* **Case Report: Priapism as The Clinical Presenting Feature of Chronic Myeloid Leukemia: Case Report and 20-Year Literature Review [version 2; peer review: 2 approved]** F1000Research 2022, 10:571 <https://doi.org/10.12688/f1000research.53365.2>

**First published:** 15 Jul 2021, 10:571 <https://doi.org/10.12688/f1000research.53365.1>

**REVISED Amendments from Version 1**

As advised by the reviewer, we have simplified the title, paid attention to unclear sentences, removed and revised them. We also have added one recommended reference to our discussion.

**Any further responses from the reviewers can be found at the end of the article**

**Introduction**

Priapism is a urological emergency due to persistence of an erection lasting more than 4 hours, whether or not it is related to sexual influence.<sup>1</sup> Priapism is a rare condition with an incidence of 1–5 cases per 100,000 people per year. Penile erection in priapism is regularly painless. There are two types of priapism, which are low-flow priapism and high-flow priapism. Low-flow priapism is provoked by a pathological condition of low venous blood flow causing stasis in the penile vessels. This condition is an emergency condition that can result in cell damage and fibrosis, thus it often requires immediate therapy. Meanwhile, high-flow priapism is caused by increased blood flow to the sinusoid arteries without offsetting the flow to the veins. One of the causes of high-flow abnormalities is penile injury, while low-flow priapism is commonly caused by blood disorders such as sickle cell anemia and chronic myeloid leukemia (CML).<sup>2–4</sup>

Priapism accounts for 20% of the hematological abnormalities while 1–5% of priapism are due to leukemia. The theory behind a priapism is the dysregulation of nitric oxide (NO) in penile vascularization. This occurs due to changes in NO synthase enzyme activity which decrease NO production by the corpora cavernosa. This ischemic condition induces platelet aggregation, thrombus, and tissue damage. Decreased NO interferes with smooth muscle tone and generates the priapism. Hyperviscosity conditions due to leukocytosis and adenosine-opiophins abnormalities is also involved in this condition.<sup>1</sup>

Currently, the approach to treat CML patients with priapism uses a combination of systemic therapy (chemotherapy with hydroxyurea or tyrosine kinase inhibitors and leukapheresis) and local intracavernosal therapy. Some cases with late manifestations cause erectile dysfunction, gangrene and penile abscess.<sup>5</sup> This case report and review aims to discuss the clinical characteristics and outcomes of CML patients who experience priapism.

**Case**

An 18-year-old unmarried male student, presented at the ER complaining of persistent erection of the penis. The patient complained of persistent erected penis for 20 days before admission. There was no phase without an erection in between. Previously, there was neither history of trauma to sexual stimulation, nor consumption of certain drugs. The patient also complained of mild genital pain along with the onset of erection. There were no complaints about discoloration of the penis; becoming reddish, bluish, or pale, also there was no numbness. The patient could urinate normally (see [Figure 1](#)).

The patient complained of tinnitus in his right and left ears for 15 days accompanied by blurred vision. The patient also felt that his left side of stomach was slowly enlarging for 5 months. There was no bleeding and fever. Before coming to the



**Figure 1.** Penis at day 2, day 6 (after intracavernosal blood aspiration), and day 9.

ER, the patient was hospitalized at the regional hospital and received a blood transfusion and was diagnosed with a blood disorder.

Physical examination revealed no anemia and icterus. The spleen was palpable showing Schuffner 4 and Hackett 3. There was no enlargement of the lymph nodes. His laboratory findings were hemoglobin 10.4 g/dL; leucocytes 421,000 cells/mm<sup>3</sup>; platelets 407,000 cells/mm<sup>3</sup>; white blood cells differential 4.3/6.8/81.3/4.9/2.7; blood urea nitrogen 9 mg/dL; serum potassium 0.5 mg/dL, uric acid 6.5 mg/dL. Peripheral blood smear showed normochromic anemia, normocytic anisopoikilocytosis, leukocytosis (3% myeloblasts, 6% promyelocytes, 4% myelocytes, 2% metamyelocytes, 5% stab neutrophils, 63% segment neutrophils, 4% eosinophils, 6% basophils, 5% lymphocytes, 2% monocytes, atypical lymphocytes (+)) concluded as CML. The patient received hydroxyurea 2000 mg once daily at night, paracetamol 500 mg TID, and an urgent leukapheresis.

The patient underwent leukapheresis once per day (three times since initial admission) with gradual improvement. Unfortunately, on the fourth day of treatment the patient felt a penis erection again with pain on a scale of 0–5. Local examination of the genitalia showed a maximal erected penis, with no discoloration indicative of hyperemia, cyanosis, or pallor. Blood gas analysis showed pH 6.95, pCO<sub>2</sub> 64 mmHg, HCO<sub>3</sub> 14 mEq/L, BE -18 unit. We concluded that the patient had ischemic priapism. Therefore, the patient underwent intracavernous aspiration producing 150 mL blood. Not long after that, the patient's penis returned to an erection with bleeding from the puncture wound. We then decided to give leukapheresis to the patient.

On the eighth day of treatment, the erection improved with pain scale of 1. Quantitative *BCR-ABL* examination showed a positive result of 65%, thus the administration of hydroxyurea was stopped and replaced by imatinib 400 mg once daily at night. On the twelfth day of treatment, the erection completely resolved and the patient was successfully discharged from the hospital.

## Discussion

This review presents data on patients who have priapism due to CML (see [Table 1](#)). Priapism occurred in the age ranging from 9–53. Patients usually had episodes of priapism for 18 h to 7 days. Not all patients with priapism showed a typical clinical examination of CML in the form of splenomegaly, but all of these patients had a hyperleukocytosis profile with a leukocyte count >200,000 cells/mm<sup>3</sup>. Some of them are equipped with data of peripheral blood smear with excessive blast and identification of *BCR-ABL* gene. A study by Minckler *et al.* was the only one reporting a resolved erection with a cold shower, whilst most other cases needed medical intervention.<sup>6</sup> Although the duration of symptoms varied, four cases reported complications following an episode of priapism. Patients with unfavorable outcomes once received hydroxyurea, imatinib but failed to undergo urological emergency therapy such as intra-cavernosa aspiration, surgical intervention, and embolization.

The patient in our study was 18 years old. However, based on the literature, patients in every age group are at risk of developing priapism. There are two peaks in the age distribution that tend to experience this condition. The peak in earlier age is between 5 and 10 years, especially in patients with sickle cell disease. Meanwhile, the second peak is at sexually active phase between 20 and 50 years. Apart from hypercoagulability, this condition may also be related to the abuse of erectile drugs.<sup>7</sup>

History and physical examination are important when encountering cases of priapism. Laboratory tests are required to check for impaired coagulation and serum electrolytes. Some patients who are at high risk for priapism include users of intracorporal injection therapy for erectile dysfunction, coagulation disorders such as sickle cell disease and CML.<sup>2,4</sup> In CML, hyperleukocytosis is thought to be the prime cause of priapism. The main mechanism is the aggregation of leukemic cells in the corpora cavernosa and dorsal veins of the penis. Other than that, mechanical pressure in the abdominal veins due to the enlargement of the spleen might also increase the risk.<sup>1</sup>

The data needed in the management of patients with this case are erection duration, pain scale, trauma, complete blood count, peripheral blood smear, penile blood gas analysis, bone marrow and polymerase chain reaction for *BCR-ABL* if necessary.<sup>1,2,4</sup> In CML, the most common type of priapism is the ischemic one (veno-occlusive). Patients usually complain of painful, rigid erection, with reduced to no cavernous blood flow at all. Priapism that lasts for more than 4 hours indicates a compartment syndrome and may require emergent medical intervention.<sup>8</sup>

The American Urological Association recommends that systemic treatment of an underlying disorder should not be the only one therapy for ischemic priapism. In this case, the patient had an erectile episode since 20 days before the admission. This phenomenon was likely due to the compartment syndrome, hence the intra-cavernous aspiration was required.<sup>1</sup>

**Table 1. Case report review from last than twenty years.**

No	Author	Country	Year	Age	Duration of priapism	Diagnosis of CML	Treatment of CML	Treatment of priapism	Outcome of the treatment
1	Gaye <i>et al.</i> <sup>4</sup>	Senegal	2020	46	48 hours	White Blood Cell: 52600/mm <sup>3</sup> , Platelets: 412000/mm <sup>3</sup> , Myelogram result: bone marrow hyperplasia. Karyotyping: Translocation between chromosomes 9 and 22	Imatinib (the dosage wasn't mentioned)	Aspiration of corpora cavernosa, injection of phenylephrine, hydroxycarbamide	Success
				9	36 hours	White Blood Cell: 82000/mm <sup>3</sup> , Platelets: 81000/mm <sup>3</sup> , BMA: acute myeloid leukemia	Vincristine and Prednisolone	penile skin refrigeration, rehydration, puncture of corpora cavernosa, injection of phenylephrine	Success
2	Rajabto <i>et al.</i> <sup>9</sup>	Indonesia	2020	44	4 days	physical exam: pale skin, conjunctival pallor, leukemic retinopathy in both eyes. Schuffer 2. Labs: anemia, hyperleukocytosis, microcytic hypochromic, anisopoikilocytosis, fragmentocytes, polychromic erythrocytes, a left shift, platelet count (355,000/ $\mu$ L), and hyperleukocytosis (399,560/ $\mu$ L).	IV fluid, Allopurinol 300 mg, Sodium bicarbonate 500 mg 3 times daily, hydroxyurea 1 gram three, Imatinib 400 mg times a day	aspiration of penile corpus, injection of epinephrine	suffered ED
							Positive BCR-ABL1 BMA: hypercellularity		
3	Dhar <i>et al.</i> <sup>11</sup>	India	2019	52	4 hours	Physical examination: massive splenomegaly of 8 cm below the left costal margin along with hepatomegaly of 3 cm below right costal margin. Blood count: left sided granulopoies, total leucocyte count of 239 $\times$ 109/L and platelet count of 625 $\times$ 109/L. BMA: findings of CML positive translocation of BCR-ABL	Hydroxyurea 500 mg TDS, Imatinib OD, Allopurinol 300 mg OD, adequate hydration	needle aspiration--> didn't work, went for Winters procedure	Success

**Table 1.** Continued

No	Author	Country	Year	Age	Duration of priapism	Diagnosis of CML	Treatment of CML	Treatment of priapism	Outcome of the treatment
4	Becerra <i>et al.</i> <sup>12</sup>	Mexico	2018	52	6 day evolution	WBC: 282,000, platelets: $368 \times 10^3/\text{mm}^3$ BMA: acute phased CML translocation t (9:22)(q34; q11.2) with P210 BCR-ABL1 fusion transcriber	dastinib 100 mg/day +G15	corpora cavernosa irrigation and surgery penis shunts	Success
5	Khan <i>et al.</i> <sup>13</sup>	Pakistan	2018	16	264 hours	Leukocyte count: $614.8 \times 10^9$ , platelets $709 \times 10^{12}/\text{L}$ , peripheral smear: myeloid hyperplasia, neutrophilia. BMA: myeloid hyperplasia. Detection of BCR-ABL	Hydroxyurea, allopurinol	Glans-cavernosal shunt	Achieved detumescence, No info on ED
6	Qu <i>et al.</i> <sup>14</sup>	China	2018	18	72 hours	Hepatosplenomegaly 2-3 cm under arcus costae, blood count: white blood cell (WBC) $257 \times 10^9/\text{L}$ and platelets (PLT) $5450 \times 10^9/\text{L}$	Imatinib	Cavernosa-corporis spongiosum shunt	No ED at 3 months follow up
7	Clark <i>et al.</i> <sup>15</sup>	USA	2018	13	3 days	Blood count: WBC count of $350,000/\text{mL}$ ( $350 \times 10^9/\text{L}$ ) and platelet count of $450 \times 10^3/\text{mL}$ ( $450 \times 10^9/\text{L}$ ). Flow cytometry of blood: granulocytosis with no increase in blasts BMA: Philadelphia chromosome	leukapheresis, IV fluids, hydroxyurea, allopurinol, Imatinib	phenylephrine injection, three times corporeal irrigation	improved with phallus rigidity and tenderness
8	Kumar <i>et al.</i> <sup>16</sup>	India	2018	47	5 days	Hepatosplenomegaly, WBC: $279 \times 109$ , 91.2%BCR	Hydroxyurea, Imatinib	Aspiration and irrigation with phenylephrine, Winter's T Shunt	Successful treatment
				42	7 days	Splenomegaly 6 cm below costal margin, WBC: $390 \times 109/\text{L}$ , 70.7% BCR-ABL ratio	Hydroxyurea, Imatinib	Aspiration and irrigation	Successful treatment
				28	6 days	No hepatosplenomegaly, WBC: $206 \times 109/\text{L}$ , 75.3% BCR-ABL ratio	Hydroxyurea, Imatinib	Aspiration and irrigation with phenylephrine, Winter's T Shunt	Successful treatment

**Table 1.** Continued

No	Author	Country	Year	Age	Duration of priapism	Diagnosis of CML	Treatment of CML	Treatment of priapism	Outcome of the treatment
9	Sun <i>et al.</i> <sup>5</sup>	USA	2018	27	8 years, persistent erection 9 hours	Labs: anemia, WBC 450,010, Platelets 509,000/mm <sup>3</sup> BMA: 2% blasts, hypercellular bone marrow, granulocytic hyperplasia, small megakaryocytes. BCR-ABL did not reveal clonal evolution.	Leukapheresis, hydroxyurea 500 mg daily, allopurinol 300 mg daily, Imatinib 400 mg daily,	Corporal bpody aspiration, 1 dose of phenylephrine injection	Successful treatment
10	Huei <i>et al.</i> <sup>17</sup>	Malaysia	2018	28	48 hours	hepatomegaly 2cm below right costal margin, splenomegaly, anemia, WBC 294 × 10 <sup>9</sup> , platelets: 94 × 10 <sup>9</sup> /L Peripheral blood smear: hyperleucocytosis, blast cells	Hydroxyurea, allopurinol, intravenous Cytarabine	Intracavernosal aspiration, phenylephrine irrigation--> detumescent --> recurrent erection --> corpoglandular shunt	Successful treatment
11	Minckler <i>et al.</i> <sup>5</sup>	USA	2017	18	3 month intermittent	WBC: 588 × 10 <sup>3</sup> /uL, platelets: 109 × 10 <sup>3</sup> /uL peripheral blood: hyperleucocytosis with absolute neutrophilia and a peripheral blast count of 2%. bone marrow aspirate and biopsy: hypercellular marrow with 4% blasts FISH analysis: translocation t(9:22)	Hydroxyurea transition to imatinib 400 mg daily	Penile irrigation and aspiration	Success
12	Nerli RB <i>et al.</i> <sup>7</sup>	India	2016	19	duration: 24 hours	WBC 296800, platelet 936,000/mm <sup>3</sup> , BMA: hypercellular, increased megakaryocytes	Hydroxyurea 1.5 gram daily, Imatinib 40 mg daily Allupurinol 300mg daily per oral	Irrigation, decompression	Successful

Table 1. Continued

No	Author	Country	Year	Age	Duration of priapism	Diagnosis of CML	Treatment of CML	Treatment of priapism	Outcome of the treatment
13	Ergenc H <i>et al.</i> <sup>18</sup>	Turkey	2015	18	duration: 72 hours	Hepatosplenomegaly 2-3 cm under arcus costae, anemia, WBC 100.000, platelets 1.002.000/mm, peripheral blood smear: immature leukocytes. BMA: hypercellularity with myeloid hyperplasia, positive BCR-ABL translocation	Imatinib 400 mg once daily, allopurinol 300 mg once daily, leukapheresis	not mentioned	Success
14	Shaeer <i>et al.</i> <sup>2</sup>	Egypt	2015	21	6 days	palpable splenomegaly, WBC 410000, Philadelphia chromosome translocation	Leukapheresis, Imatinib 400 mg daily	failed several cavernosal aspiration and injection of epinephrine --> penile prosthesis	No complication throughout 6 months- follow up
15	Osorio <i>et al.</i> <sup>19</sup>	Spain	2014	24	14 hours, the second episode. The first episode was 4 months ago	WBC: 177.15×10 <sup>9</sup> , platelet was not mentioned, cytogenic diagnosis: showing CML	Imatinib	Corpora cavernosa aspiration, intracavernosa fenilefrin injection	not mentioned
16	Hazra <i>et al.</i> <sup>20</sup>	India	2013	14	6 hours, the second episode. The first episode was less than a month ago	WBC: 402.24×10 <sup>9</sup> , platelet was not mentioned positive BCR-ABL	hydroxyurea	Corpora cavernosa aspiration, intracavernosa fenilefrin injection	not mentioned
17	Vejjkovic <i>et al.</i> <sup>21</sup>	Serbia	2012	16	24 hours	Splenomegaly 6 cm below the left costal margin, anemia, WBC 226900, platelets 310,000/uL, Peripheral blood smear: immature leukocytes in various stages. BMA: CML.	Hydroxyurea 50 mg/kgBB/day, Allopurinol 300 mg/day	Cavernosal aspiration and phenylephrine irrigation	No recurrence at 2-months- follow-up
					24 hours	Splenomegaly 4 cm below costal margin, WBC 320×10 <sup>9</sup> /L, Platelet (Plt) 417×10 <sup>9</sup> /L BMA: extreme hypercellularity, BCR/ABL positive	leukapheresis, cytoreductive chemotherapy	leukapheresis	no follow up



Table 1. Continued

No	Author	Country	Year	Age	Duration of priapism	Diagnosis of CML	Treatment of CML	Treatment of priapism	Outcome of the treatment
18	Paladino <i>et al.</i> <sup>3</sup>	Spain	2011	16	48 hours	Splenomegaly, WBC 312.000, PLT: 60.000/mm <sup>3</sup> BMA: showing CML	no mention	Corpora cavernosa drainage	Erectile dysfunction
19	Gupta <i>et al.</i> <sup>22</sup>	India	2009	12	48 hours	Hepatosplenomegaly below the costal margins, anemia, WBC: 346 × 10 <sup>9</sup> /L, platelet count of 40,000/mm <sup>3</sup> , peripheral blood smear: immature myeloid leukocytosis. Cytogenesis: Philadelphia chromosome. BCR-ABL transcript was positive	hydroxyurea 4g/day IV fluid 3L/day, allopurinol, Imatinib 400mg/day, leukapheresis	Terbutaline 0.125 mg subcutaneously	Resolved by 24 h
20	Ilais Tazi <sup>23</sup>	Morocco	2009	33	duration: 22 hours	Palpable splenomegaly 4 cm below left costal margin, WBC: 40000/mm <sup>3</sup> , platelets 120000/mm <sup>3</sup> . Peripheral blood smear: immature leukocytes. Karyotype analysis: Ph1 chromosome, myeloid hyperplasia in the bone marrow.	Imatinib	Aspiration	Success
21	Castagnetti <i>et al.</i> <sup>24</sup>	Netherland	2008	9	several days	splenomegaly, anemia, WBC: 509 × 10 <sup>9</sup> /L, Philadelphia chromosome, BCR-ABL +	Hydroxyurea 1.5mg/m <sup>2</sup> /day, Cyclophosphamide 250 mg/m <sup>2</sup> /day for 2 days, leukapheresis	cytoreduction, antibiotics, anticoagulants	Fully resolved after 1 month
22	Yoshida <i>et al.</i> <sup>25</sup>	Japan	2007	29	48 hours	WBC 263000	Hydroxyurea 1g/m <sup>2</sup> /day	LMWH 90 units/kg SQ BID for 1 month, metamazole	fully resolved after 3 months
23	Lopez <i>et al.</i> <sup>26</sup>	Spain	2004	29	10 hours	WBC 414 × 10 <sup>9</sup> /L, BMA: hypercellularity, PLT: 1100 × 10 <sup>9</sup> /L	Cyclophosphamide 250 mg/m <sup>2</sup> /day for 2 days, leukapheresis	LMWH 90 units/kgBB SQ BID for 9 days, metamazole, morphine	fully resolved after 20 days
							Imatinib mesylate	Winter procedure	no evidence of recurrent
							corpora cavernosa aspiration, phenylephrine injection	corpus cavernosum aspiration, fenilefrin injection	Successful treatment

Table 1. Continued

No	Author	Country	Year	Age	Duration of priapism	Diagnosis of CML	Treatment of CML	Treatment of priapism	Outcome of the treatment
24	Ponniah <i>et al.</i> <sup>27</sup>	United Kingdom	2004	19	18 hours	WBC 513×109/L	Leukapheresis	failed cavernosal aspiration + leukapheresis	No ED on follow up
25	Dogra <i>et al.</i> <sup>28</sup>	India	2003	18	10 days	hepatosplenomegaly, anaemic, WBC 320000, PLT was not mentioned	Intravenous hydration, furosemide, sodium bicarbonate, hydroxyurea, allopurinol, leukapheresis	Winters Procedure	impotent and enlarged penis at 3-months follow up
26	Meng-Wei Chang <i>et al.</i> <sup>8</sup>	Taipei	2003	21	19 hours	Hepatomegaly 6 cm below right arcus costae, Splenomegaly 7 cm below left arcus costae, anemia, WBC 216800, Platelet 1746,000/mm <sup>3</sup>	Interferon alfa-2a (6MIU/vial), allopurinol 300 mg daily	Aspiration, epinephrine irrigation	Success
27	Guerra <i>et al.</i> <sup>29</sup>	Spain	2002	53	12 hours	Wbc 968×109/L	Hydroxyurea	Corpora cavernosa aspiration	Successful treatment
28	Murayama <i>et al.</i> <sup>30</sup>	Japan	2001	14	4 days	WBC 510000, BMA: myeloid hyperplasia, karyotype analysis: chromosome Ph1	urokinase, hydroxyurea	embolization of bilateral pudendal artery	Reduced sexual potency
29	Rojas <i>et al.</i> <sup>31</sup>	Chilli	1998	22	duration: 36 hours	none	Leukapheresis	Surgical intervention	Unsuccessful (post treatment sexual dysfunction)

The intra-cavernous aspiration procedure can be accomplished by giving the anesthetic injection first under the symphysis pubis. The penis is tied with a tourniquet followed by insertion of a 16–18-Gauge bivalve intravenous catheter into the corpus cavernosum. When the two corpora are fused, aspiration of 20–30 mL of blood can be undertaken. This procedure has 30% chances of success.<sup>8,9</sup>

Systemic therapy is often used to reduce hyperviscosity is cytoreductive therapy such as high-dose hydroxycarbamide and tyrosine kinase inhibitors (TKI) with or without apheresis procedures. Hydroxycarbamide can be given 2–6 grams divided into four doses per day. This can reduce leukocytes by almost 60% in 24–48 h. In addition, TKI, such as imatinib, can be administered as soon as the diagnosis is confirmed. The recommended dose of imatinib is 400 mg once daily in the chronic phase, 600–800 mg once daily in the accelerated phase, and 800 mg once daily in a blast crisis.<sup>9</sup> Generally, IRIS study describes the effectiveness of imatinib therapy for complete hematological response (CHR), major cytogenetic response (McyR) and complete cytogenetic response (CcyR).<sup>4</sup>

Leukapheresis can promote a rapid decrease in intravascular leukemic cells, improve tissue perfusion and prevent leukostasis (generally show pulmonary and central nervous system manifestations). Once leukapheresis is given, it possibly can reduce the leukocyte count by 30–60%. However, compared to the chemotherapy, several previous studies have shown that this procedure had high all-cause mortality. According to 2016 apheresis guidelines, category 2 (second-line therapy) is recommended for grade 1B of acute myeloid leukemia (strong recommendation, moderate quality evidence), while category 3 (unclear role of apheresis) is recommended for acute lymphoblastic leukemia cases grade 2C (weak recommendation, low quality evidence). In this guideline, leukapheresis is not recommended for chronic myeloid leukemia.<sup>10</sup> Several cases of priapism in this case review reported a successful combination of leukapheresis with systemic oral CML therapy. A study by Rojas *et al.* was the only one reporting a failed leukapheresis.

This case report and review presents a comparative presentation of patient characteristics, clinical characteristics of CML, laboratory profile, and therapeutic intervention for CML with priapism. Clinical presentation and early intervention are pivotal keys to achieve favorable outcome and prevent complications. Systemic intervention combined with intraurethral therapy may add the success rate (see Figure 2).

Eventually, further discussion and study on other causes of priapism is essential as a meta-analysis stated that priapism might also be related to lymphoproliferative disorders.<sup>32</sup>

Treatment :	N	%
• Cytoreduction	19	54%
• Tyrosine Kinase Inhibitor	17	49%
• Leukapheresis	13	37%
• Penile aspiration	19	54%
○ Penile aspiration and sympatomimetic	10	29%
○ Penis-Shunt	6	17%
Outcome :		0%
• Success	22	63%
• Erectile dysfunction	3	9%
• Not mentioned	4	11%

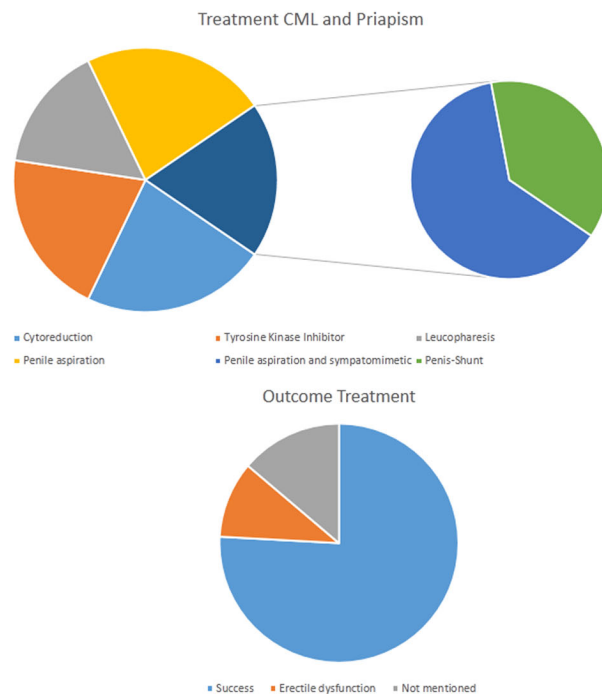


Figure 2. Treatment and outcome from priapism and CML.

## Consent

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

## Data availability

All data underlying the results are available as part of the article and no additional source data are required.

## References

- Rodgers R, Latif Z, Copland M: **How I manage priapism in chronic myeloid leukaemia patients.** *Br J Haematol.* 2012; **158**(2): 155–64. [PubMed Abstract](#) | [Publisher Full Text](#)
- Shaer OK, Shaer KZ, AbdelRahman IF, et al.: **Priapism as a result of chronic myeloid leukemia: case report, pathology, and review of the literature.** *J Sex Med.* 2015; **12**(3): 827–34. [PubMed Abstract](#) | [Publisher Full Text](#)
- Paladino N, Roldán D, Caram MS: **[Priapism in pediatrics: initial presentation of chronic myeloid leukemia].** *Arch Argent Pediatr.* 2011; **109**(5): e104–8. [PubMed Abstract](#) | [Publisher Full Text](#)
- Gaye O, Thiam NM, Cassell A, et al.: **Unusual Presentation of Priapism Associated with Acute and Chronic Myeloid Leukemia in Two Patients: Emergency Management.** *Case Rep Urol.* 2020; **2020**: 4982432. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Sun HH, Zhang JH, DeWitt-Foy M, et al.: **Urologic Management of Priapism Secondary to Chronic Myeloid Leukemia.** *Urology.* 2019; **125**: 24–8. [PubMed Abstract](#) | [Publisher Full Text](#)
- Minckler MR, Conser E, Figueroa JJ, et al.: **The Semantics of Priapism and the First Sign of Chronic Myeloid Leukemia.** *Case Rep Emerg Med.* 2017; **2017**: 2656203. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Nerli RB, Magdum PV, Hiremath SC, et al.: **Priapism – A Rare Presentation in Chronic Myeloid Leukemia: Case Report.** *Urol Case Rep.* 2016; **4**: 8–10. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Chang MW, Tang CC, Chang SS: **Priapism—a rare presentation in chronic myeloid leukemia: case report and review of the literature.** *Chang Gung Med J.* 2003; **26**(4): 288–92. [PubMed Abstract](#)
- Rajabto W, Djanzonie JAC, Prastithita LB, et al.: **Priapismus as Leukostasis Manifestation in Chronic Myeloid Leukemia.** *Acta Med Indones.* 2020; **52**(4): 420–2. [PubMed Abstract](#)
- Schwartz J, Padmanabhan A, Aqvi N, et al.: **Guidelines on the Use of Therapeutic Apheresis in Clinical Practice—Evidence-Based Approach from the Writing Committee of the American Society for Apheresis: The Seventh Special Issue.** *J Clin Apher.* 2016; **31**(3): 149–62. [PubMed Abstract](#) | [Publisher Full Text](#)
- Dhar J, Dhar J, Chhabra G, et al.: **Priapism as a Debut Presentation of Chronic Myeloid Leukemia.** *J Coll Physicians Surg Pak.* 2019; **29**(1): 78–80. [PubMed Abstract](#) | [Publisher Full Text](#)
- Becerra-Pedraza LC, Jiménez-Martínez LE, Peña-Morfin I, et al.: **Priapism as the initial sign in hematologic disease: Case report and literature review.** *Int J Surg Case Rep.* 2018; **43**: 13–7. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Khan A, Shafiq I, Shah MH, et al.: **Chronic myeloid leukaemia presenting as priapism: A case report from Khyber Pakhtunkhwa.** *J Pak Med Assoc.* 2018; **68**(6): 942–4. [PubMed Abstract](#)
- Qu M, Lu X, Wang L, et al.: **Priapism secondary to chronic myeloid leukemia treated by a surgical cavernosa-corpora spongiosum shunt: Case report.** *Asian J Urol.* 2019; **6**(4): 373–6. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Clark AJ, Hsu P, Darves-Bornoz A, et al.: **Case 3: Priapism in a 13-year-old Boy.** *Pediatr Rev.* 2018; **39**(12): 617–9. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Kumar P, Rahman K, Kumari S, et al.: **Priapism as a rare presentation of chronic myeloid leukemia.** *J Cancer Res Ther.* 2018; **14**(6): 1442–3. [PubMed Abstract](#) | [Publisher Full Text](#)
- Huei TJ, Lip HT, Shamsuddin O: **A rare presentation of chronic myeloid leukaemia with priapism treated with corporoglandular shunting.** *Med J Malaysia.* 2018; **73**(6): 420–2. [PubMed Abstract](#)
- Ergenc H, Varim C, Karacaer C, et al.: **Chronic myeloid leukemia presented with priapism: Effective management with prompt leukapheresis.** *Niger J Clin Pract.* 2015; **18**(6): 828–30. [PubMed Abstract](#) | [Publisher Full Text](#)
- Villegas Osorio JF, Corchuelo Maíllo C, Cuevas Palomino A, et al.: **Ischaemic priapism as a presentation of chronic myeloid leukaemia.** *Arch Esp Urol.* 2014; **67**(8): 708–11. [PubMed Abstract](#)
- Hazra SP, Priyadarshi V, Gogoi D, et al.: **Pediatric priapism: a rare first manifestation of leukemia.** *APSP J Case Rep.* 2013; **4**(3): 39. [PubMed Abstract](#) | [Free Full Text](#)
- Veljković D, Kuzmanović M, Mičić D, et al.: **Leukapheresis in management hyperleucocytosis induced complications in two pediatric patients with chronic myelogenous leukemia.** *Transfus Apher Sci.* 2012; **46**(3): 263–7. [PubMed Abstract](#) | [Publisher Full Text](#)
- Gupta A, Seth T, Gupta A: **Successful use of terbutaline in persistent priapism in a 12-year-old boy with chronic myeloid leukemia.** *Pediatr Hematol Oncol.* 2009; **26**(1): 70–3. [PubMed Abstract](#) | [Publisher Full Text](#)
- Tazi I: **Priapism as the first manifestation of chronic myeloid leukemia.** *Ann Saudi Med.* 2009; **29**(5): 412. [PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
- Castagnetti M, Sainati L, Giona F, et al.: **Conservative management of priapism secondary to leukemia.** *Pediatr Blood Cancer.* 2008; **51**(3): 420–3. [PubMed Abstract](#) | [Publisher Full Text](#)
- Yoshida K, Kinoshita H, Taniguti H, et al.: **Priapism complicated by chronic myelogenous leukemia (CML); a case report.** *Hinyokika Kyo.* 2007; **53**(5): 323–5. [PubMed Abstract](#)
- Allué López M, García de Jalón Martínez A, Pascual Regueiro D, et al.: **Priapism as an initial presentation of chronic myeloid leukaemia.** *Actas Urol Esp.* 2004; **28**(5): 387–9. [PubMed Abstract](#)
- Ponniah A, Brown CT, Taylor P: **Priapism secondary to leukemia: effective management with prompt leukapheresis.** *Int J Urol.* 2004; **11**(9): 809–10. [PubMed Abstract](#) | [Publisher Full Text](#)
- Dogra PN, Kumar P, Goel R, et al.: **Long duration priapism in blast crisis of chronic myeloid leukemia.** *J Assoc Physicians India.* 2004; **52**: 170. [PubMed Abstract](#)
- Cruz Guerra NA, Ramos LC, Linares Quevedo A, et al.: **Priapism secondary to chronic myeloid leukemia: value of initial treatment with cavernous lavage plus adjuvant methoxamine.** *Arch Esp Urol.* 2002; **55**(3): 320–1. [PubMed Abstract](#)
- Murayama K, Shibuya A, Ishii S, et al.: **[Embolization of the bilateral internal pudendal arteries for intractable priapism in a child with chronic myelogenous leukemia].** *Rinsho Ketsueki.* 2001; **42**(11): 1117–1. [PubMed Abstract](#)
- Rojas B, Cabrera ME, Kliwadenko W, et al.: **[Priapism in a patient with chronic myeloid leukemia].** *Revista medica de Chile.* 1998; **126**(8): 978–80.
- Ali EA, Sardar S, Yassin MA: **Priapism in lymphoproliferative disorders: A systematic review.** *Hematol Oncol Stem Cell Ther [Internet].* 2021. [Publisher Full Text](#)

# Open Peer Review

Current Peer Review Status:  

---

## Version 2

Reviewer Report 14 January 2022

<https://doi.org/10.5256/f1000research.80090.r119664>

© 2022 Gupta R. This is an open access peer review report distributed under the terms of the [Creative Commons Attribution License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.



**Ritu Gupta** 

Laboratory Oncology Unit, Dr B.R. Ambedkar IRCH, All India Institute of Medical Sciences (AIIMS), New Delhi, New Delhi, Delhi, India

The authors have summarized the subject adequately and I have no further comments.

**Competing Interests:** No competing interests were disclosed.

**Reviewer Expertise:** Hemato-Oncology, Genomics, Single-cell sequencing, Flow cytometry

**I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.**

---

## Version 1

Reviewer Report 02 December 2021

<https://doi.org/10.5256/f1000research.56738.r100511>

© 2021 Gupta R. This is an open access peer review report distributed under the terms of the [Creative Commons Attribution License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.



**Ritu Gupta** 

Laboratory Oncology Unit, Dr B.R. Ambedkar IRCH, All India Institute of Medical Sciences (AIIMS), New Delhi, New Delhi, Delhi, India

Priapism is an unusual complication of hematological malignancy with hyperleukocytosis and may be the presenting feature, especially in chronic leukemia as observed in this case.

The authors have described the clinical features, investigations, and management of the index case and reviewed the literature on the association of priapism with CML.

I have a few comments/suggestions on this manuscript as detailed below:

1. The title is too long and can be abbreviated.
2. Review of hematological malignancies presenting as priapism i.e. including CLL and Acute leukemia would benefit the readers in developing insight on the conditions in which priapism could be the presenting feature. A few references include the following: Johnson *et al.* (2020<sup>1</sup>), Ali *et al.* (2021<sup>2</sup>) and Gogia *et al.* (2012<sup>3</sup>).
3. The meaning of some of the sentences is not clear in several places. The authors may focus on improving the language of the paper.

### References

1. James Johnson M, Hallerstrom M, Alnajjar HM, Frederick Johnson T, et al.: Which patients with ischaemic priapism require further investigation for malignancy?. *Int J Impot Res.* 2020; **32** (2): 195-200 [PubMed Abstract](#) | [Publisher Full Text](#)
2. Ali EA, Sardar S, Yassin MA: Priapism in lymphoproliferative disorders: A systematic review. *Hematol Oncol Stem Cell Ther.* 2021. [PubMed Abstract](#) | [Publisher Full Text](#)
3. Gogia A, Sharma A, Raina V, Gupta R: Priapism as an initial presentation of chronic lymphocytic leukemia. *Leuk Lymphoma.* 2012; **53** (8): 1638-9 [PubMed Abstract](#) | [Publisher Full Text](#)

**Is the background of the case's history and progression described in sufficient detail?**

Yes

**Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?**

Yes

**Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?**

Partly

**Is the case presented with sufficient detail to be useful for other practitioners?**

Yes

**Competing Interests:** No competing interests were disclosed.

**Reviewer Expertise:** Hemato-Oncology, Genomics, Single-cell sequencing, Flow cytometry

**I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.**

Author Response 03 Dec 2021

**Pradana Zaky Romadhon**, Airlangga University, Faculty of Medicine, Surabaya, Indonesia

Firstly, thank you for the detailed review and advice. We have simplified the title and removed unclear sentences then replaced them with more understandable ones.

**Competing Interests:** No competing interests were disclosed.

Author Response 04 Dec 2021

**Pradana Zaky Romadhon**, Airlangga University, Faculty of Medicine, Surabaya, Indonesia

Dear Ritu Gupta,

We already just submitted our new version of the manuscript. We also have included one of the references recommended by you in our discussion. Hope it will upgrade our manuscript quality. Thanks again.

**Competing Interests:** No competing interests were disclosed.

Reviewer Report 18 October 2021

<https://doi.org/10.5256/f1000research.56738.r89693>

© 2021 Rajabto W. This is an open access peer review report distributed under the terms of the [Creative Commons Attribution License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.



**Wulyo Rajabto**

Division of Hematology-Medical Oncology, Department of Internal Medicine, Dr. Cipto Mangunkusumo General Hospital, Faculty of Medicine, University of Indonesia, Jakarta, Indonesia

This case report emphasizes the importance of priapism as the rare clinical presentation of chronic myeloid leukemia so that as a clinician we should think if there is patient with priapism the secondary causal is chronic myeloid leukemia. The treatment of priapism consists of: 1) Local factor by urologist who performs intra cavernous aspiration 2) The systemic factor by hematologist who administers leucapheresis (mechanical and drug eg. Hydroxyurea) and TKIs such as Imatinib.

I find the title of this manuscript indeed captivating. Besides describing the priapismus phenomenon in CML, the author also showed to us a comparison study among several previously published cases known worldwide, that I think it is a very interesting plus point.

- Title: I believe it is very interesting, straightforwardly describes the case.
- Introduction: I believe it contains concise reasoning why the author brought up this case,

emphasizes the rare of similar cases, and interestingly presents one of CML emergencies.

- Case presentation: The author successfully managed to present the case elaborately along with valid data.
- Discussion: The author describes the case comprehensively, referred to similar case studies before, from the clinical course to the outcome, as mention on table 1.

**Is the background of the case's history and progression described in sufficient detail?**

Yes

**Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?**

Yes

**Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?**

Yes

**Is the case presented with sufficient detail to be useful for other practitioners?**

Yes

**Competing Interests:** No competing interests were disclosed.

**Reviewer Expertise:** CML, lymphoma, anemia

**I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.**

---

The benefits of publishing with F1000Research:

- Your article is published within days, with no editorial bias
- You can publish traditional articles, null/negative results, case reports, data notes and more
- The peer review process is transparent and collaborative
- Your article is indexed in PubMed after passing peer review
- Dedicated customer support at every stage

For pre-submission enquiries, contact [research@f1000.com](mailto:research@f1000.com)

**F1000Research**