

## Priapism -- A Rare Presentation in Chronic Myeloid Leukemia: Case Report and Review of The Literature

Meng-Wei Chang, MD; Chung-Chih Tang<sup>1</sup>, MD; Shy-Shin Chang, MD

Priapism is a complication rarely seen in leukemia. We report a 21-year-old man presented with persistent painful erection of penis for 19 hours at home. The patient had undergone immediate irrigation and decompression of priapism by urologist at emergency department. This approach resulted in a flaccid penis later. During hospitalization, peripheral blood smear and bone marrow aspiration was confirmatory of chronic myeloid leukemia. No impotency nor other sequela was noted after his discharge. This case illustrates the importance of all physicians in the diagnosis and management of patients with priapism. (*Chang Gung Med J* 2003;26:288-92)

**Key words:** priapism, chronic myeloid leukemia, emergency department, impotency.

Priapism is persistent abnormal erection of penis without accompanied sexual desire. It is traditional to consider priapism as idiopathic and 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 anemia, trauma, leukemia, cancerous invasion of the penis, drugs, alcoholic ingestion, various thromboembolic disease, and intravenous fat for parenteral nutrition. About 20 percent cases of all priapism are related to hematological disorders.<sup>(1)</sup> The incidence of priapism in adult leukemic patients is about 1-5 percent and leukemia is frequently associated with painful priapism.<sup>(2-3)</sup> The following case illustrates priapism as an unusual presenting symptom of chronic myeloid leukemia.

### CASE REPORT

A previously healthy 21-year-old man was referred from local hospital for treatment of priapism and hyperleukocytosis. His penis remained erect,

painful, and swelling when he arrived at emergency department (ED). There was no history of trauma, malaise, night sweats, joint pain, and cough. However, body weight loss about 9 Kg and bleeding tendencies during recent 3 months were noted himself. The vital signs revealed a body temperature 38.2°C, pulse 109 beats/minute, and blood pressure 137/64 mmHg, respirations 21 beats/minute. He was alert 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 no jaundice. The penis was erect, firm, and tender with superficial venous engorgement. Laboratory data showed hemoglobin (Hb) 8.3 g/dl, hematocrit 25.7%, white blood count (WBC) 216,800/mm<sup>3</sup>, and Platelet 1746,000/mm<sup>3</sup>. Serum chemistries were unremarkable for uric acid 452 µmol/L, creatinine 70.7 µmol/L. Treatment of the priapism was initiated performed by cavernosa aspiration and epinephrine irrigation at ED under the impression of low flow type priapism because of his history and physical examination. The erection was relieved later by

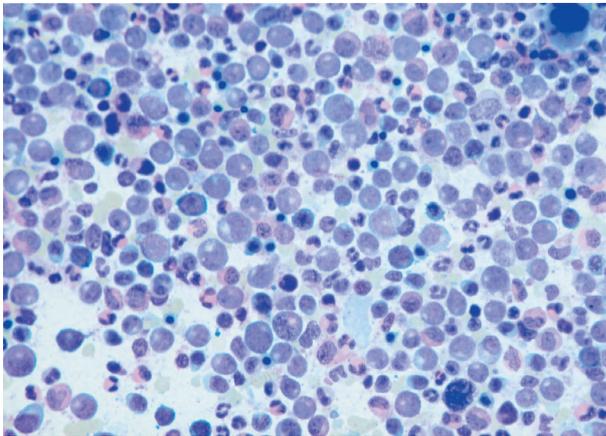
---

From the Department of Emergency Medicine, <sup>1</sup>Division of Hematology-Oncology, Chang Gung Memorial Hospital, Taipei.

Received: Mar. 20, 2002; Accepted: Aug. 20, 2002

Address for reprints: Dr. Shy-Shin Chang, Department of Emergency Medicine, Chang Gung Memorial Hospital, 5, Fushing Street, Gueishan Shiang, Taoyuan, Taiwan 333, R.O.C. Tel.: 886-3-3281200 ext. 2505; Fax: 886-3-3287715; E-mail: sschang@cgmh.org.tw

these procedures. For hyperleukocytosis, he was admitted to hematological ward and was diagnosed as a case of chronic myeloid leukemia on the basis of peripheral blood smear and bone marrow examination (Fig.1). The Philadelphia chromosome was illustrated in the patient. He was started on hydroxyurea tablets at 1.5 grams per day and one vial of interferon alfa-2a (6 MIU/vial) subcutaneously daily. Allopurinol 300mg daily with adequate hydration was also started for potential tumor lysis syndrome. Before discharge, his WBC dropped to 82900/ mm<sup>3</sup> and Hb raised to 9.7 g/dl. Recurrent priapism was not happened to him during his admission period. The patient continues to report to us without any erectile dysfunction till date.



**Fig. 1** The bone marrow smear demonstrates hypercellularity with myeloid hyperplasia, no increase in blasts.

## DISCUSSION

Priapism is an involuntary prolonged erection unrelated to sexual activity and cannot be relieved by ejaculation. Most priapism is painful but not all cases. Priapism is defined as either low-flow (ischemic) or high-flow (non-ischemic).<sup>(4)</sup> Low-flow or ischemic priapism results from pathologically decreased penile venous outflow that eventuates in stasis. Intracavernosal blood sampling reveals acidosis and a decrease in oxygen tension.<sup>(5)</sup> Clinically, low-flow priapism manifests as a painful, rigid erection. This type is more common and represents an actual emergency because irreversible cellular damage and fibrosis occur if treatment is not adminis-

tered within 24 to 48 hours.<sup>(6)</sup> It will result in long-term sequela of erectile dysfunction or predisposition to frequent, prolonged episodes of priapism.<sup>(7)</sup> The cause of low-flow priapism including idiopathic, hematologic disorders,<sup>(8-9)</sup> tumor infiltrate,<sup>(10)</sup> or drugs induced.<sup>(11-12)</sup> 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 was painless. In contrast to low-flow priapism, intracavernosal blood sampling from patients with high-flow priapism reveals bright red oxygenated blood,<sup>(5)</sup> and thus irreversible cellular damage and fibrosis are rare.<sup>(6)</sup> The type of priapism is usually due to penis or perineum trauma that results in injury to the internal pudendal artery.<sup>(13-15)</sup> This establishes a fistula between the cavernosal artery and the corpus cavernosum that unregulated inflow occur. It is not an actual emergency in patients with high-flow priapism, and treatment can be on an elective basis.

Priapism can occur at any age and two peaks in age distribution is described.<sup>(16)</sup> A pediatric peak, 5-10 years old, is noted owing to sickle cell disease in black patients. The secondary peak occurs in patients with active sexual activity age of 20-50 years old. Idiopathic priapism is the most common (64%) while approximately 20% are related to hematologic disorders.<sup>(1,16)</sup> In CML, priapism is an unusual presentation and seldom to encounter. Hyperleukocytosis is thought to be the cause of priapism in patients with leukemia.<sup>(4)</sup> Four different mechanism is described: (1) venous congestion of the corpora cavernosa resulting from mechanical pressure on the abdominal veins by the splenomegaly (2) Sludging of leukemic cells in the corpora cavernosa and the dorsal veins of penis (3) infiltration of the sacral nerves with leukemic cells (4) infiltration of the central nerve system. In our case, significant leukocytosis with hepatosplenomegaly supports the first mechanism in the pathogenesis. An important aspect of priapism is that most physicians will never encounter. The poor experience will result in delay of treatment and irreversible sequel. So all physicians should understand that long-term sequela can be avoided with prompt diagnosis and treatment.

To diagnose the underlying pathophysiology of priapism, the distinction between low and high flow priapism is important because their associated treatment and prognosis differ. Differentiating between

low-flow and high-flow priapism can be achieved with a detailed history, physical examination, gas analysis of the blood within the corpora cavernosa,<sup>(15)</sup> and penile Doppler ultrasound study. Detailed history includes medications, malignant diseases, trauma, sickle cell disease, and the use of intracavernosal agents, all of which provide clues about the type of priapism. On physical examination, patients with low-flow priapism usually have a rigid, painful penile shaft with a soft glans, whereas in those with high-flow priapism, the entire penis is partially rigid and painless. Intracavernosal blood gas analysis is helpful in differentiating between high and low flow priapism. Low-flow priapism have an intracavernosal blood pH < 7.00,<sup>(11)</sup> a PCO<sub>2</sub> > 60 mmHg, and a PO<sub>2</sub> < 30 mmHg.<sup>(17)</sup> These values vary depending on the duration of the low-flow state. High-flow priapism patients have normal blood gas analysis values. Doppler ultrasonography can be used to detect the presence of an arterial-to-cavernosum fistula in the patients with high-flow priapism. Besides, the initial work-up should include complete blood count, hemoglobin electrophoresis, serum chemistries, coagulopathy state, and drugs screen if indicated.

About the management of priapism, there have been many methods described in the literature in the world. Spinal anesthesia, ice water enema, ice packs, fibrinolytic agents and anticoagulants have been tried but no significant success rate obtained. To relieve the painful erection of the patient, immediate aspiration and irrigation of the corpora cavernosa as well as injection of  $\alpha$ -adrenergic agents were performed. The penis should be cleansed and 2% xylocaine is used to anesthetize the glans penis and septum between glans and corpora; general or spinal anesthesia is an alternative. To reduce the anesthesia risk, we did these procedures under sedation and local anesthesia. So this intervention can be done by any physician at anywhere with appropriate preparation. After preparation, repeated intracavernous aspirations through an 18-22 gauge needle into the corpora cavernosa and gentle normal saline irrigation were performed with a 10 ml syringe. The sluggish bloods were removed and the intracavernous pressure was reduced. Erect penis may relieve by these procedures along. However, it is more effective with an additional injection of  $\alpha$ -adrenergic agents such as phenylephrine or epinephrine. If the erection persists for 24-48 hours, the patient should have a surgi-

cal shunt performed to reduce the priapism. The purpose of surgical procedures is to establish a new venous outflow and restore normal arterial flow to the corpora cavernosa. The Urologists should also involve in the early intervention because of their familiar with the management and complication of the priapism.

The importance of prompt diagnosis and treatment of priapism cannot be overemphasized, as there is definite incidence of impotence following this condition. One study cited 35% and 60% impotence rates for patients priapistic for 5 days and 10 days, respectively.<sup>(18)</sup> So decompression of the penis should be done as frequently as possible during the first 24 hours.<sup>(2)</sup>

Besides the initial relief of priapism, the further workup and management of the underlying disease are more important. In our case, with use of a combined urological therapy and oncological treatment to priapism, the patient was recovered and had restored long-term potency. In conclusion, priapism is an uncommon presentation in CML that all physicians should be aware of the disorders and the need for early intervention and management.

## REFERENCES

1. Becker LE, Mitchell AD. Priapism. *Surg Clin North Am* 1965;45:1522.
2. Sheribman SM, Gee TS, Grabstodt H. Management of Priapism in patients with chronic granulocytic leukemia. *J Urol* 1974;111:786-8.
3. Nelson JH, Winter CC. Priapism: evolution of management in 48 patients in a 22-year series. *J Urol* 1977;117:445-8.
4. Winter CC, McDowell G. Experience with 105 patients with priapism: Update review of all aspects. *J Urol* 1988;140:980-3.
5. Lue TF, Hellstrom WJ, McAninch JW, Tanagho EA. Priapism: a refined approach to diagnosis and treatment. *J Urol* 1986;136:104-8.
6. Broderick GA, Gordon D, Hypolite J, Levin RM. Anoxia and corporal smooth muscle dysfunction: a model for ischemic priapism. *J Urol* 1994;151:259-62.
7. Levine FJ, Saenz de Tejada I, Payton TR, Goldstein I. Recurrent prolonged erections and priapism as a sequela of priapism: pathophysiology and management. *J Urol* 1991;145:764-7.
8. Fowler JE Jr, Koshy M, Strub M, Chinn SK. Priapism associated with the sickle cell hemoglobinopathies: prevalence, natural history and sequela. *J Urol* 1991;145:65-8.
9. Hamre MR, Harmon EP, Kirkpatrick DV, Stern MJ,

- Humbert JR. Priapism as a complication of sickle cell disease. *J Urol* 1991;145:1-5.
10. Powell BL, Craig JB, Muss HB. Secondary malignancies of the penis and epididymis: a case report and review of the literature. *J Clin Oncol* 1985;3:110-6.
  11. Saenz de Tejada I, Ware JC, Blanco R, Pittard JT, Nadig PW, Azadzi KM, Krane RJ, Goldstein I. Patho-physiology of prolonged penile erection associated with trazodone use. *J Urol* 1991;145:60-4.
  12. Lomas GM, Jarow JP. Risk factors for papaverine-induced priapism. *J Urol* 1992;147:1280-1.
  13. Ilkay AK, Leverine LA. Conservative management of high-flow priapism. *Urology* 1995;46:419-24.
  14. Ji MX, He NS, Wang P, Chen G. Use of selective embolization of the bilateral cavernous arteries for post-traumatic arterial priapism. *J Urol* 1994;151:1641-2.
  15. Bastuba MD, Saenz de Tejada I, Dinlenc CZ, Sarazen A, Krane RJ, Goldstein I. Arterial priapism: diagnosis, treatment and long-term follow up. *J Urol* 1994;151:1231-7.
  16. Singh N, Bhatnagar DP. Priapism: A rare presentation of Chronic Myeloid Leukemia. *JAPI* 1985;33:741-2.
  17. Stackl W, Mee SL. Priapism. In: Krane RJ, Siroky MB, Fitzpatrick JM. *Clinical Urology*. Philadelphia: J.B.Lippincott, 1994:1245-8.
  18. Pohl J, Pott B, Kleinhaus G. Priapism: a three-phase concept of management according to etiology and prognosis. *Br J Urol* 1986;58:113-6.

# 陰莖持久勃起 -- 慢性骨髓性白血病之罕見初期表現： 病例報告與文獻回顧

張孟維 湯崇志<sup>1</sup> 張詩鑫

陰莖持久勃起是白血病少見併發症。一位21歲男性，陰莖持續勃起19小時而就醫。在急診，泌尿科醫師立即施以陰莖灌洗與抽吸，此病患的症狀才得以緩解。在住院過程中，週邊血液抹片檢查及骨髓檢驗皆證實其為慢性骨髓性白血病，出院後並無陽萎或其他併發症出現。希望藉此病例說明診斷與治療陰莖持久勃起上的重要性。(長庚醫誌 2003;26:288-92)

**關鍵字：**陰莖持久勃起，慢性骨髓性白血病，急診，陽萎。

---

長庚紀念醫院 台北院區 急診醫學科；<sup>1</sup>血液腫瘤科

受文日期：民國91年3月20日；接受刊載：民國91年8月20日。

索取抽印本處：張詩鑫醫師，長庚紀念醫院 急診醫學科。桃園縣333龜山鄉復興街5號。Tel.: (03)3281200轉2505; Fax: (03) 3287715; E-mail: sschang@cgmh.org.tw