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Case Report

Priapism secondary to chronic myeloid leukemia treated by a surgical cavernosa-corpus spongiosum shunt: Case report

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Abstract Priapism secondary to chronic myeloid leukemia (CML) is rarely observed in the clinic. Here, we present an 18-year-old patient with priapism for over 72 h due to hyperleukocytosis. Emergent interventions such as therapeutic aspiration and intracorporal injection of phenylephrine failed before a surgical corpora cavernosa-corpus spongiosum shunt was inserted to relieve symptoms. During hospitalization, bone marrow aspiration confirmed the diagnosis of CML.

1. Introduction

Priapism is a condition of an abnormal erectile penis in the absence of physical and psychological stimulation for several hours. Generally, there are two types of priapism: Low-flow (ischemic), which accounts for approximately 80%–90% of cases, and high-flow (nonischemic) [1]. Most ischemic priapisms are idiopathic, whereas other cases might be caused by the injection of vaso-active substances, such as papaverine and phentolamine, or by hematological diseases, such as chronic myeloid leukemia (CML), sickle cell disease, polycythemia vera, and multiple myeloma, resulting in hyperviscosity of the circulating blood [2]. Priapism secondary to CML is extremely rare in clinics. Here, we report a young adult with CML who presented with priapism as the first clinical manifestation.

2. Case report

A previously healthy and sexually active 18-year-old man was admitted to the emergency department of our center. He presented with consistent erectile penis for nearly 1 week and aggravation with pain for over 72 h (Fig. 1A). The

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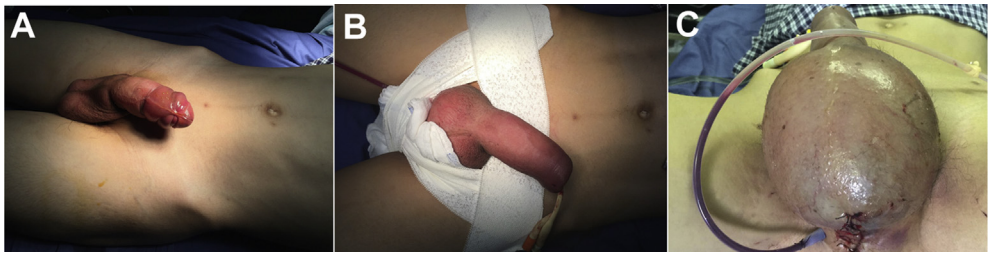

Figure 1 The clinical situation of the patient in the hospital. (A) Pathological erectile penis of the patient; (B) Flaccid penis after surgery; (C) Swelling scrotum due to compression of blood clots.

patient had no history of sexual stimulation, medications, trauma, malaise, night sweats, joint pain or cough. The physical examination revealed that the spleen and liver were 2—3 cm palpable under the arcus costa. The penis was erect and firm with superficial venous engorgement. Laboratory tests showed white blood cell (WBC) and platelets (PLT) counts of 257×10^9/L and 5450×10^9/L, respectively. The patient's hemoglobin level was 71 g/L. Abdominal ultrasonography showed enlargement of the liver and spleen. An ultrasonography of the penis showed low blood flow. Based on the clinical manifestations and laboratory tests, we speculated that the priapism was caused by hematology disease. Thus, bone marrow aspiration was performed before the therapeutic aspiration and intra-cavernous injection of phenylephrine. Unfortunately, the symptoms were not effectively controlled. Given that a long course of the disease would eventually cause irreversible complications, such as erectile dysfunction, we decided to insert a surgical proximal corpora cavernosa-corpus spongiosum shunt to relieve the symptoms instead of a distal shunt due to the severe edema of the glans.

The surgery was successfully performed by shunting bilateral proximal blood from the rigid corpora cavernosa into the corpus spongiosum. The penis was apparently flaccid (Fig. 1B). However, the scrotum became significantly enlarged, which might have been caused by the many blood clots from a coagulation disorder the day after the surgery (Fig. 1C). A secondary surgery was arranged, and the blood clots were evacuated from the subcutaneous layer before the incision was tightly sutured.

The results of the bone marrow analysis showed hypercellularity with myeloid hyperplasia. The BCR-ABL translocation gene test was positive according to fluorescence *in situ* hybridization (FISH), which confirmed the suspicion of CML as the underlying etiology of priapism. Imatinib mesylate (400 mg) was administered once daily with intravenous fluids hydration after wound healing. The 3-month follow-up showed that the patient had no severe complications such as erectile dysfunction. Moreover, he had normal WBC and PLT counts, as shown in repeated laboratory tests.

3. Discussion

An erection is caused by the parasympathetic stimulation of the cavernosum smooth muscle and hyperemia of the corpus cavernosum. Under normal circumstances, there is microscopic arterial vascularization of the corpora cavernosa. The corpora cavernosa was found to be vascularized by both the cavernous and dorsal arteries of the penis [3]. Conversely, priapismis is pathological hyperemia of the corpus cavernosum. Priapism can occur at any age, particularly in 5- to 10- and 20- to 50-year-olds. Approximately 70% of cases are idiopathic whereas 20% are related to hematological disorders in adults [4]. Based on the blood flow, there are two types of priapisms: Nonischemic and ischemic, which occur via different mechanisms. Nonischemic priapisms are observed in clinics much less frequently than ischemic priapisms. The etiology of nonischemic priapism remains unclear, although pharmacological, traumatic and neurological diseases that can cause unregulated, continuous arterial inflow into the lacunar spaces have been proposed. Usually, ischemic priapism is more commonly observed in clinics.

The incidence of priapism is only 1%—2% in patients with CML [5], which is quite rare (Table 1). Most patients present with painful erectile penis without any sexual stimulation for several hours. The arteria dorsalis penis is usually unreached, which can be further confirmed by ultrasonography. The laboratory tests generally show that WBC and PLT counts are significantly elevated. Severe long-term complications, such as erectile disorder, resulting from fibrosis and necrosis of the abnormal erectile penis can easily occur if treatment is insufficient. Several emergent treatments, including therapeutic aspiration, intracavernosal injections of phenylephrine, and rapid detumescence by leukapheresis, can be applied in the emergency room. As a final approach, a surgical shunt should be considered to lower the incidence of inevitable complications. A systemic evaluation and therapy for the underlying etiology, such as using imatinib mesylate to treat CML, is necessary after providing first aid.

Obviously, therapeutic aspiration is the first treatment option. A penile block using 10—20 mL of 1% lignocaine should be applied before the procedure. Lateral injection and trans-glans injection are both available. The aspiration of 20—30 mL of blood is followed by the injection of heparinized saline, which must be repeated a few times. A success rate of less than 30% has been reported [6].

Intracavernosal injections of phenylephrine is a secondary treatment attempt; however, it has several risks and side effects, including malignant hypertension, tachycardia and cardiac arrhythmia, particularly in patients with cardiovascular disease. Generally, phenylephrine is diluted in normal saline to a concentration of 100—200 μg/mL and is injected at a rate of 1 mL/5 min under blood pressure monitoring.

Table1 Case reports of priapism secondary to chronic myeloid leukemiain in recent 10 years.

Study	Number of cases	Age (year)	White blood cell count ($\times 10^9$/L)	Treatment	Result
Kurosawa et al. [7], 2016	4	NA	NA	3 for therapeutic aspiration, 1 for leukapheresis	2 with erectile dysfunction
Nerli et al. [8], 2016	1	19	296.8	Corporal aspiration and phenylephrine irrigation	No follow-up
Ergenc et al. [4], 2015	1	18	100	Leukapheresis	No erectile dysfunction
Shaeer et al. [9], 2015	1	21	410	Initiated by cavernosal aspiration/irrigation and injection of epinephrine, followed by penileprosthesis surgery	Erectile dysfunction followed by penile prosthesis implantatio
Hazra et al. [10], 2013	1	14	226.9	Cavernosal aspiration, irrigation and phenylepinephrine injection	Maintaining erection
Veljkovic et al. [11], 2012	1	16	320	Leukapheresis	No follow-up
Tazl [12], 2009	1	33	400	Cavernosa aspiration and epinephrine irrigation	No erectile dysfunction
Gupta et al. [13], 2009	1	12	346	Administration of terbutaline	No follow-up

NA, Not available.

Leukapheresis is a method in which circulating blood is mechanically separated into liquid and blood cells, followed by the removal of WBCs and returning the remaining components to the patients. The side effects include electrolyte disturbance and hypothermia, which can be prevented by electrolyte infusion and rigorous body temperature monitoring.

A surgical shunt is inserted to reverse the priapic state by shunting blood from the rigid corpora cavernosa into the corpus spongiosum. A distal shunt is the primary choice unless the distal penile is inappropriate because of the condition of edema or tissue damage. A proximal shunt is applied less frequently because it may lead to more complications, such as erectile dysfunction, urethral fistula, purulent cavernositis, and pulmonary embolism [6]. Early penile prosthesis implantation is recommended under the circumstances of a shortened, indurated and non-erectile penis. Apart from the early resumption of sexual activity, early implantation can avoid the formation of dense fibrosis and hence a shortened penis.

As a matter of course, the underling etiology of the disease must be treated, such as via folic acid and penicillin or blood transfusion for sickle-cell disease, imatinib mesylate for CML, deferoxamine or bone marrow transplantation for thalassemia, *etc*.

4. Conclusion

The case we present here is a rare clinical manifestation of priapism secondary to CML. A routine blood test is the most valuable evidence in the emergency room to support the diagnosis of CML in addition to physical examinations and ultrasonography. A bone marrow analysis and gene test can confirm the diagnosis but require more time. We hope to remind all doctors and increase their awareness of the importance of an accurate diagnosis and rapid intervention so that irreversible complications are avoided. A surgical shunt is not a primary recommendation for treatment in the emergency room due to its related complications. However, when all other procedures fail, it is the most effective way to relieve pain.

Author contributions

Study design: Min Qu, Xin Lu.
Surgery performing: Lei Wang, Zhiyong Liu.
Drafting of manuscript: Min Qu.
Critical revision of the manuscript: Yinghao Sun, Xu Gao.

Conflicts of interest

The authors declare no conflict of interest.

References

[1] Van der Horst C, Stuebinger H, Seif C, Melchior D, Martinez-Portillo FJ, Juenemann KP. Priapism—etiology, pathophysiology and management. Int Braz J Urol 2003;29:391—400.
[2] Burnett AL, Bivalacqua TJ. Priapism: new concepts in medical and surgical management. Urol Clin North Am 2011;38:185—94.
[3] Diallo D, Zaitouna M, Alsaid B, Quillard J, Droupy S, Benoit G. What is the origin of the arterial vascularization of the corpora cavernosa? A computer-assisted anatomic dissection study. J Anat 2013;223:489—94.
[4] Ergenc H, Varim C, Karacaer C, Cekdemir D. Chronic myeloid leukemia presented with priapism: effective management with prompt leukapheresis. Niger J Clin Pract 2015;18:828—30.

[5] Rodgers R, Latif Z, Copland M. How I manage priapism in chronic myeloid leukaemia patients. Br J Haematol 2012;158:155—64.
[6] Kandel GL, Bender LI, Grove JS. Pulmonary embolism: a complication of corpus-saphenous shunt forpriapism. J Urol 1968;99:196—7.
[7] Kurosawa H, Tanizawa A, Tono C, Watanabe A, Shima H, Ito M, et al. Leukostasis in children and adolescents with chronic myeloid leukemia: Japanese Pediatric Leukemia/Lymphoma Study Group. Pediatr Blood Cancer 2016;63:406—11.
[8] Nerli RB, Magdum PV, Hiremath SC, Patil AY, Pai SV, Handigund RS, et al. Priapism — a rare presentation in chronic myeloid leukemia: Case report. Urol Case Rep 2016;4:8—10.
[9] Shaeer OK, Shaeer KZ, AbdelRahman IF, El-Haddad MS, Selim OM. Priapism as a result of chronic myeloid leukemia: case report, pathology, and review of the literature. J Sex Med 2015;12:827—34.
[10] Hazra SP, Priyadarshi V, Gogoi D, Sharma PK, Pal DK, Chakraborty SC. Pediatric priapism: a rare first manifestation of leukemia. APSP J Case Rep 2013;4:39.
[11] Veljkovic D, Kuzmanovic M, Micic D, Šerbić-Nonković O. Leukapheresis in management hyperleucocytosis induced complications in two pediatric patients with chronic myelogenous leukemia. Transfus Apher Sci 2012;46:263—7. https://doi.org/10.1016/j.transci.2012.03.012.
[12] Tazi I. Priapism as the first manifestation of chronic myeloid leukemia. Ann Saudi Med 2009;29:412.
[13] Gupta A, Seth T, Gupta A. Successful use of terbutaline in persistent priapism in a12-year-old boy with chronic myeloid leukemia. Pediatr Hematol Oncol 2009;26:70—3.