CASE REPORT

Priapism as the initial presentation of chronic myeloid leukemia in a patient with cerebral palsy: A case report

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ABSTRACT

Priapism is defined as a penile erection lasting more than four hours without stimulation. The majority of cases involve ischemic priapism, in which there is partial or complete absence of cavernosal arterial flow. Priapism is uncommon, with an estimated annual incidence of approximately 1.5 cases per 100,000 men. In chronic myeloid leukemia (CML) patients, leukostasis may occur, which can lead to priapism in 1-2% of cases. We present the case of a 21-year-old patient with a history of cerebral palsy who was brought to the emergency department by his family with priapism persisting for 13 hours. It was noted that the patient had experienced one prior episode of priapism a month earlier, which had resolved spontaneously. He had no history of trauma. On physical examination, extremity contractures related to cerebral palsy were noted, and the penis was observed to be erect. A venous catheter was inserted into the corpus cavernosum, dark-colored blood was aspirated, and blood gas analysis confirmed the diagnosis of ischemic priapism. The corpus cavernosum was irrigated with normal saline, and epinephrine was administered into both corpora cavernosa; however, priapism did not resolve. Laboratory tests revealed a white blood cell count of 581,760 /mm³. The patient was referred to the hematology department. Peripheral blood smear analysis confirmed the diagnosis of CML, and leukapheresis was performed three times within two days. Following normalization of the peripheral white blood cell count, penile detumescence was achieved, and no further episodes of priapism occurred. In conclusion, early diagnosis and a multidisciplinary approach improve the success of priapism treatment and reduce the risk of complications.

Keywords: Priapism; chronic myeloid leukemia; leukostasis.

INTRODUCTION

Priapism is defined as a penile erection lasting more than four hours without stimulation. The majority of cases involve ischemic priapism, in which there is partial or complete absence of cavernosal arterial flow. In ischemic priapism, metabolic changes occur due to hypoxia, hypercapnia, and acidosis, with patients typically experiencing pain after 6-8 hours.[1]

Priapism is an uncommon condition, with an estimated annual incidence of approximately 1.5 cases per 100,000 men. Hematologic disorders account for 20% of priapism cases in males, with sickle cell anemia being the most common cause. Additionally, priapism due to venous occlusion has been reported in 1-5% of male leukemia patients, with chronic myeloid leukemia (CML) representing 50% of these cases.[2,3]

This case report presents a 21-year-old patient with cerebral palsy who was diagnosed with CML following the presentation of priapism.

CASE REPORT

A 21-year-old male patient with a history of cerebral palsy-associated intellectual disability and epilepsy, controlled with antiepileptic medications, was brought to the emergency department by his family with priapism persisting for 13 hours. The family reported that the patient had experienced

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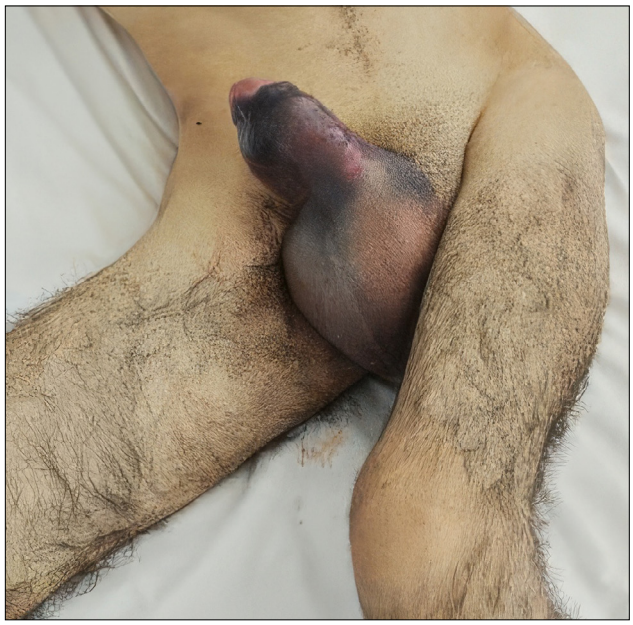

Figure 1. Erect penis and contracted extremities due to cerebral palsy.

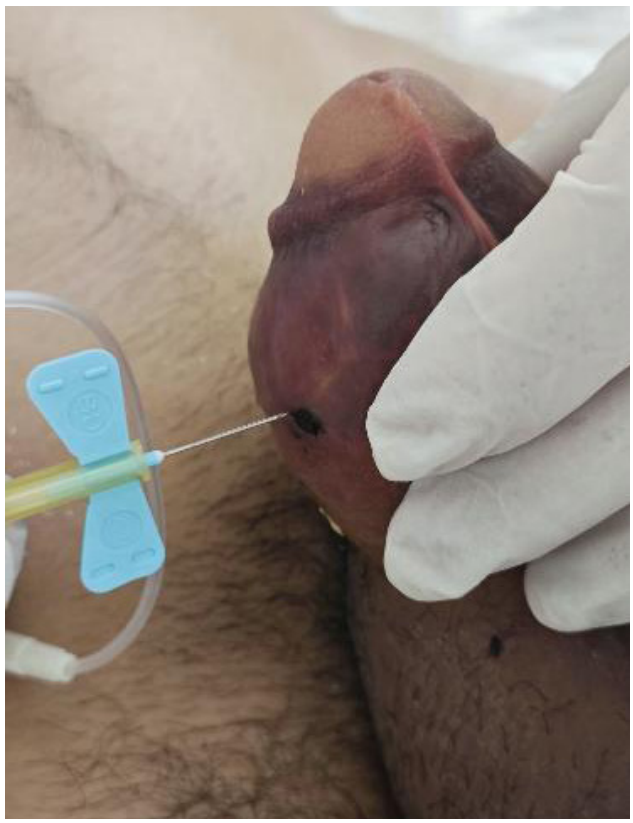

Figure 2. Aspiration of blood from the corpus cavernosum using a 16-gauge venous catheter.

a single prior episode of priapism one month earlier, which had resolved spontaneously. He had no history of trauma. On physical examination, extremity contractures related to cerebral palsy were noted. Due to lack of cooperation, an abdominal examination could not be performed. The penis was observed to be in an erect state (Fig. 1).

For penile pain blockade, 10 mL of 1% lidocaine was administered to the penile radix. Subsequently, for the diagnosis and treatment of priapism, a 16-gauge venous catheter was inserted laterally into the corpus cavernosum. Dark-colored blood was aspirated, and blood gas analysis was performed, revealing a pH of 7.23, PO_2 of 34.5 mmHg, and PCO_2 of 39.2 mmHg, confirming the diagnosis of ischemic priapism. Since priapism persisted after aspiration, bilateral catheterization was performed, and the corpus cavernosum was irrigated with normal saline. Additionally, epinephrine (1:100,000; 5 cc) was administered twice at 20-minute intervals into both corpora cavernosa; however, priapism did not resolve (Fig. 2).

Laboratory tests revealed a white blood cell (WBC) count of 581,760 /mm³, hemoglobin of 8.6 g/dL, hematocrit of 23.5%, and a platelet count of 141,000 /mm³. Blood biochemistry values were within normal limits.

The patient, who exhibited leukocytosis, was referred to the hematology department. Peripheral blood smear analysis led to the diagnosis of CML, and emergency leukapheresis was performed. Oral allopurinol (300 mg twice daily) and hydroxyurea (1 g four times daily) were initiated and continued for 10 days. Leukapheresis was performed three times within two days, and by the end of the second day, detumescence was achieved. The patient continued treatment and follow-up under the hematology department. At the third- and sixth-month check-ups, the WBC count was within normal limits. According to family reports, the patient intermittently had normal erections, and no further episodes of priapism occurred. Written informed consent was obtained from the patient's parents for publication of clinical details and imaging findings.

DISCUSSION

Ischemic priapism is a condition that requires urgent urological intervention and, if not treated early, can lead to permanent sequelae such as erectile dysfunction. It can occur at any age but is most commonly seen in individuals aged 5-10 years and 20-50 years.[2,3] Approximately 70% of cases are idiopathic, while 20% of cases in adults are associated with hematological disorders. In CML patients, significant leukocytosis may be present at the time of diagnosis, which can lead to leukostasis or hyperviscosity syndrome. In hyperviscosity syndrome, visual disturbances such as retinopathy and retinal hemorrhage, pulmonary insufficiency, and priapism (in 1-2% of cases) can occur.[4] In the case presented here, the diagnosis of CML was made after investigations prompted by the development of priapism.

Systemic treatments frequently used in CML patients include high-dose hydroxyurea, tyrosine kinase inhibitors, and leukapheresis to reduce hyperviscosity.[4] Leukapheresis is a proce-

dure in which circulating blood is mechanically separated into liquid and cellular components, with excess white blood cells removed and the remaining components returned to the patient. Recent reviews of case reports from the literature have shown that, in addition to acute penile interventions, emergency leukapheresis is routinely performed—where available—for the treatment of ischemic priapism secondary to CML in both pediatric and adult patients. This approach has demonstrated favorable outcomes in managing leukostasis.[5,6]

The American Urological Association (AUA) emphasizes that systemic treatment of underlying conditions such as CML should not be considered the only treatment for ischemic priapism. Ischemic priapism should be regarded as a compartment syndrome, and penile-specific interventions are required as the initial approach.[7]

CONCLUSION

In cases of ischemic priapism, blood tests in addition to physical examination findings are crucial to avoid missing underlying hematological pathologies. When priapism is related to CML, primary penile interventions should be performed alongside systemic treatments, and emergency leukapheresis therapy should be considered in patients with hyperviscosity syndrome. Early diagnosis and a multidisciplinary approach can improve priapism treatment success and reduce the risk of complications.

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REFERENCES

1. 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 2015;4:8-10. [CrossRef]
2. Broderick GA. Priapism. In: Wein AJ, Kavoussi LR, Partin AW, Peters CA, editors. Campbell-Walsh-Wein Urology. 12th ed. Philadelphia: Elsevier; 2020. p.1539-63.e4.
3. 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;26:288-92.
4. Rodgers R, Latif Z, Copland M. How I manage priapism in chronic myeloid leukaemia patients. Br J Haematol 2012;158:155-64. [CrossRef]
5. Suttorp M, Sembill S, Kalwak K, Metzler M, Millot F. Priapism at diagnosis of pediatric chronic myeloid leukemia: Data derived from a large cohort of children and teenagers and a narrative review on priapism management. J Clin Med 2023;12:4776. [CrossRef]
6. Van der Velde MGAM, Tiellemans SMB, de Lil H, Nieuwenhuizen L. The value of leukapheresis for treatment of priapism as presenting feature of chronic myeloid leukemia-Case report and review of literature. EJHaem 2022;3:1100-5. [CrossRef]
7. Montague DK, Jarow J, Broderick GA, Dmochowski RR, Heaton JP, Lue TF, et al. American Urological Association guideline on the management of priapism. J Urol 2003;170:1318-24. [CrossRef]

OLGU SUNUMU - *ÖZ*

Serebral palsi hastasında kronik Myeloid Lösemi'nin ilk bulgusu olarak priapizm: Vaka sunumu

Priapizm, uyarı olmaksızın 4 saatten uzun süren penil ereksiyon olarak tanımlanır. Olguların çoğu, kısmen veya tamamen kavernöz arter akımının olmadığı iskemik priapizmdir. Priapizm nadir görülen bir durum olup yıllık insidansı yaklaşık 100.000 kişide 1.5 vakadır. Kronik miyeloid lösemi (KML) hastalarında lökostaza bağlı olarak %1–2 oranında priapizm gelişebilmektedir. Sunulan olguda; serebral palsi öyküsü bulunan 21 yaşında erkek hasta, 13 saattir devam eden priapizm nedeniyle ailesi tarafından acil servise getirildi. Travma öyküsü olmayan hastanın, bir ay önce kendiliğinden düzelen tek bir priapizm atağı öyküsü olduğu öğrenildi. Hastanın fizik muayenesinde serebral palsiye bağlı ekstremite kontraktürleri tespit edildi ve penisin rijid olduğu gözlemlendi Korpus kavernozuma venöz kateter yerleştirilerek koyu renkli kan aspire edildi ve kan gazı analizi sonucunda iskemik priapizm tanısı doğrulandı. Korpus kavernozum fizyolojik serum ile irrige edildi ve her iki korpus kavernozuma epinefrin uygulandı; ancak priapizm gerilemedi. Laboratuvar testlerinde kan lökosit sayısı 581.760/mm³ olarak saptanması üzerine hasta Hematoloji bölümü tarafından değerlendirildi. Yapılan periferik yayma incelemesi sonucunda KML tanısı kondu ve hastaya iki gün içerisinde üç defa lökoferez uygulandı. Kan lökosit sayısının normal sınırlara gerilemesinin ardından penil detümesans sağlandı ve tekrar priapizm atağı görülmedi. Sonuç olarak, erken tanı ve multidisipliner yaklaşım, priapizm tedavisinde başarıyı artırmakta ve komplikasyon riskini azaltmaktadır.

Anahtar sözcükler: Kronik miyeloid lösemi; lökostaz; priapizm.

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