



## OLGU SUNUMU / CASE REPORT

# Thrombosis of corpus cavernosum in a sickle cell anemia patient with priapism

Priapizmle başvuran bir orak hücreli anemili hastada korpus kavernozum trombozu

Gül İlhan

Mustafa Kemal University Faculty of Medicine, Department of Hematology, Antakya, Hatay, Turkey

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### Abstract

Priapism is prolonged painful erection and can be seen as a sickle cell anemia complication. Thrombosis of the corpus cavernosum is a rare condition. A thirty years old male with of sickle cell anemia was admitted to our clinic with priapism. Corpus cavernosum aspiration and exchange transfusion were performed. Painful erection declined but the pain didn't improved. Corpus cavernosum thrombosis was detected and his pain relieved after systemic anticoagulation treatment. Thrombosis of corpus cavernosum must be in the mind especially for patients who have complaints despite conventional treatment methods.

**Key words:** Sickle cell disease, priapism, thrombosis

### Öz

Priapizm orak hücreli aneminin bir komplikasyonu olarak görülebilen uzamış ağrılı ereksiyondur. Korpus kavernozum trombozu nadir görülen bir durumdur. Otuz yaşında bir orak hücreli anemi hastası priapizm şikayeti ile kliniğimize başvurdu. Hastaya korpus kavernozum aspirasyonu ve exchange transfüzyon uygulandı. Hastanın ağrılı ereksiyonu geriledi ancak ağrısı düzelmedi. Hastada korpus kavernozum trombozu saptandı ve sistemik antikoagülasyon tedavisi sonrası hastanın ağrısı düzeldi. Özellikle konvansiyonel tedavi yöntemleri ile şikayetleri devam eden hastalarda korpus kavernozum trombozu akılda tutulmalıdır.

**Anahtar kelimeler:** Orak hücre hastalığı, priapizm, tromboz

## INTRODUCTION

Sickle cell anemia (SCA) is the most common hereditary hemoglobinopathy. In beta-globin chains, valine-glutamic acid substitution results in formation of an abnormal globin chain. Abnormal hemoglobin named hemoglobin S leads aggregation and polymerization in the case of hypoxia in erythrocytes. In the course of the disease, anemia, painful vasoocclusive crisis, multiple organ damage and premature death may occur.

Priapism is painful erection situation over 4 hours and occurs without any sexual stimulation. Ischemic causes of priapism are SCA, multiple myeloma, fat embolism, glukoz 6 phosphate dehydrogenase deficiency and hemoglobin variants of Olmsted. Ischemic priapism should be treated immediately. Not taking into glans penis, hypoxia and disruption

of blood flow in the corpus cavernosum causes hypercapnia. Finally this situation results painful compartment syndrome and if left untreated can lead to muscle necrosis and fibrosis. Treatment is aspiration of the corpus cavernosum with phenylephrine diluted with normal saline every 3-5 minutes. Simultaneously, hydration, oxygen and exchange transfusion treatments should be done. Non-ischemic priapism is seen with injection or perineal trauma and does not lead to smooth muscle damage<sup>1,2,3</sup>. Thrombosis of the corpus cavernosum is a rare condition. In literature it is known also as a partial priapism. It is characterized by thrombosis of the proximal segment of one corpus cavernosum. We report a patient suffering from this disease that was successfully treated conservatively. SCA causes hereditary tendency to increase thrombosis. Corpus cavernosum thrombosis was rarely reported in the literature in SCA.

Yazışma Adresi/Address for Correspondence: Dr. Gül İlhan, Mustafa Kemal University Faculty of Medicine, Department of Hematology. E-mail: gullhan2002@yahoo.com  
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## CASE

Thirty years old male with SCA admitted to emergency department because of painful erection. He had rare painful crisis before and he had not used hydroxyurea until then. His painful erection prolonged over 4 hours. Laboratory tests revealed mild anemia, leukocytosis and thrombocytosis (hemoglobin: 9 g/dl, leukocyte: 13 000/ $\mu$ L, platelet: 650 000/ $\mu$ L). And serum bilirubin: 4 mg/dl, indirect bilirubin: 3 mg/dl, AST: 55 U/L, ALT: 45 U/L, blood urea nitrogen: 20 mg/dl, creatinin: 1mg/dl, lactic dehydrogenase: 355 U/L, C-reactive protein: 25 mg/L. Intravenous fluid and analgesic treatment were started.

We performed corpus cavernosum aspiration and exchange transfusion therapy. His painful erection declined. But he had still pain spreading to groin. And then venous Doppler ultrasonography showed penile corpus cavernosum thrombosis. He had no thrombosis history. We tested for hereditary thrombophilia tests. Factor V Leiden, prothrombin G20210A and methylenetetrahydrofolate reductase mutations were negative. Bemiparin sodium 7500 units /day dose and warfarin were started. In addition, we gave him hydroxyurea 1500 mg per day. Patient pain decreased within 1 week. After 3 months venous Doppler ultrasonography showed no thrombosis and warfarin was stopped.

## DISCUSSION

First partial cases of priapism have been described by Hillis<sup>4</sup>. All of the cases described in the literature had unilateral application of perineal mass. The exact cause of the disease is not certain. According to Hillis, there is a fibrous septum in the flaccid part of corpus cavernosum and it creates predisposition to thrombosis. According to other authors, the fibrous septum occurs after trauma and keeps blood in itself<sup>5</sup>. In most cases, there is no history of trauma. Our case had no trauma either.

In the literature, angiography, cavernosography, biopsy of the corpus cavernosum, computerized tomography, magnetic resonance imaging and color-coded duplex sonography was used<sup>6,7</sup>. There are some treatment options like surgical corporotomy, cavernosum-spongiosum shunt and intracavernous injection of etilefrin<sup>7-9</sup>. Non invasive systemic anticoagulation therapy can provide erectile function. Invasive procedures are recommended for

selected cases<sup>6,10-12</sup>.

SCA is a hemoglobinopathy which increases the tendency to priapism and thrombosis. Both intrinsic and extrinsic coagulation ways are activated in SCA. Not only coagulation but also vascular inflammation, endothelial activation, asplenia and platelet activation lead to thrombosis<sup>13,14</sup>. Particularly in the cerebrovascular, and pulmonary systems thrombosis have been reported<sup>15</sup>. Thrombosis has rarely been seen in SCA patients presented with priapism<sup>16,17</sup>. Anticoagulation should be done carefully because of risk of intracranial hemorrhage and intravitreal hemorrhage.

In conclusion, priapism in SCA patients should be carefully evaluated. Thrombosis of corpus cavernosum must be in the mind especially for patients who have complaints despite conventional treatment methods.

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