

## CASE REPORT

# Priapism and glucose-6-phosphate dehydrogenase deficiency: An underestimated correlation?

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**Summary** *Priapism is a rare clinical condition characterized by a persistent erection unrelated to sexual excitement. Often the etiology is idiopathic. Three cases of priapism in glucose-6-phosphate dehydrogenase (G6PD) deficiency patients have been described in literature. We present the case of a 39-year-old man with glucose-6-phosphate dehydrogenase deficiency, who reached out to our department for the arising of a non-ischemic priapism without arteriolacunar fistula. We suggest that the glucose-6-phosphate dehydrogenase deficiency could be an underestimated risk factor for priapism.*

**KEY WORDS:** Non-ischemic; Glucose-6-phosphate dehydrogenase (G6PD) deficiency.

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## INTRODUCTION

There are three types of priapism with different pathophysiological causes: the ischemic priapism (or “low flow”), which is a dolorous and prolonged erection with a tissue ischemia, the non-ischemic (or “high flow”) priapism, which is usually due to a perineal injury with the formation of an arteriolacunar fistula, and the stuttering (intermittent) priapism, which is a recurrent form of ischemic priapism in which unwanted painful erections occur repeatedly with intervening periods of detumescence. All types of priapisms can lead to an erectile dysfunction (1), not only the ischemic one. For that reason, it is important to treat it in time and in correct way, but also to identify the possible risk factors. Three cases of priapism in patients with G6PD deficiency have been described in literature (2). We report an unique case of “high flow” priapism without fistula in a patient with G6PD deficiency.

## CASE PRESENTATION

In October 2015, a 39-year-old man reached to our observation for an intermittent, painless erection, everyday, for about one month. He reported erections lasting about 4-6 hours during the first days of the month. These erections stopped in the following days, then he had another erection lasting more than 17 hours, which

obliged him to go to the emergency department. He reported a normal and spontaneous erectile function before the arising of the disease. He denied having any gender of trauma and he admitted to have a G6PD deficiency, documented by laboratory tests. He denied having taken any drugs, phosphodiesterase type 5 (PDE5) inhibitors, intracavernosal agents or any other pharmacological agents. He denied any other risk factor for priapism. At clinical examination he presented a painless erection. Systemic hemoglobin was 13.6 g/dL.

Intracorporal blood resulted as arterial at gas analysis ( $PO_2 = 76$  mmHg;  $PCO_2 = 44.5$  mmHg;  $pH = 7.35$ ). The Color Doppler ultrasonography didn't point out any arterocavernosa fistula. After a period of observation in which there wasn't the resolution of the priapism, we decided to submit him to a pelvic arteriography (Figure 1).

Panoramic arteriography of the aorto-iliac and selective of internal pudendal artery showed a bilateral increase of the arterial level of the corpora cavernosa, higher in the left one, without a fistula. We performed a temporary embolization with *Spongostan*<sup>TM</sup> of the distal left internal pudendal artery. A complete detumescence was obtained and the next arteriography confirmed the procedure was a success. The patient was released.

After 2 weeks of a state of good health, he presented a new episode of prolonged erection, after which he returned to the emergency department of our Institute.

## Figure 1.

*Pelvic arteriography showing a bilateral increase of the arterial level of the corpora cavernosa, higher in the left one, without a fistula.*



This time the erection was higher and mildly painful. He still denied having taken drugs or medications. We performed a gas analysis ( $pO_2 = 21.3$  mmHg;  $pCO_2 = 56.4$ ; mmHg;  $pH = 7.34$ ). We carried out an evacuation of blood from the cavernosa corpora through transglanular cannula and injection of Etilefrine which lead to a detumescence. After an observation time of 24 hours without erections, we decided to discharge the patient from our department. Three months later he affirmed to have preserved a sufficient erectile function (IIEF-5: 18).

## CONCLUSIONS

The number of priapisms, classified as idiopathic, both ischemic and not-ischemic, continues to be high. Among the clinically significant enzymatic defects, G6PD deficiency is the most common (3), with hundreds and millions of affected people. With the proliferation of the G6PD deficiency, especially in some countries, we suggest it could be a casual factor or a co-factor, responsible for some cases of priapism classified as idiopathic. Therefore, now we consider the G6PD deficiency as a underestimated risk factor for the emergence of priapism.

## DISCUSSION

Priapism can be associated with some hematologic, coagulative and enzymatic disorders (4-5) such as leukemia and sickle cell anemia. Hematologic diseases are usually associated with a "low flow" priapism, but some associations have also been reported between "high flow" priapism and sickle cell anemia (6). Both type of priapism are also associated with Fabry disease (7, 8). Nowadays, few cases of priapism, related to a G6PD deficiency, are present in literature, even if the correlation between the two disorders has been discovered (2, 9, 10). The cases, which have emerged until now, showed a link between the G6PD deficiency and the "low flow" priapism. In our case, the patient initially showed a "high flow" priapism, without the presence of other risk factors for the disorder.

The interesting thing was the absence of fistula at the arteriography, but the presence of an increase of the blood flow in the corpora cavernosa. Although the arrangements are unknown, the G6PD deficiency could be responsible for that increase of blood flow and it could lead to a not-ischemic priapism other than an ischemic one, as it's already documented and assumed for the other enzymatic and hematologic defects (6-8).

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