CASE REPORT

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Ischemic Priapism Associated with Glucose-6-phosphate Dehydrogenase Deficiency: A Case Report

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ABSTRACT

Ischemic priapism is a male urologic emergency. Most cases have been linked to genetic conditions such as sickle cell disease and (much more rarely) glucose-6-phosphate dehydrogenase deficiency, and the use of certain drugs. Here, we report the case of a 34-year-old male who was a known case of the recurrent ischemic type of priapism, which was relieved by penile aspiration. Genetic investigation revealed that the patient had glucose-6-phosphate dehydrogenase.

Priapism is a painful penile erection that lasts at least four hours. If left untreated for longer periods, risks of long-term damage increase significantly, including long-term erectile dysfunction.[1] Early presentation and prompt detumescence may enable nearly 95% of the patients to maintain satisfactory sexual performance.[2]

Stuttering or recurrent priapism, commonly an ischemic type, is less common and is associated with sickle cell disease (SCD) and cannabis usage.[3,4] Repeated attacks of painful erections may occur, especially late at night, often beginning with partial penile hardness and slowly progressing to last a long time.

The physiological changes in ischemic priapism are not fully understood, but the involvement of an intra-cavernosal-controlling mechanism associated with phosphodiesterase-5 type has been hypothesized.[5,6] With high consanguinity levels, the Omani population has a high incidence of various genetic disorders, including SCD and associated conditions such as priapism.[4]

Cases associating glucose-6-phosphate dehydrogenase (G6PD) deficiency with ischemic priapism are extremely rare.[7–9] Herein, we are reporting a new case of ischemic priapism linked to G6PD deficiency, which, to our knowledge, will be the first in Oman.

CASE REPORT

A 34-year-old man with no past medical background presented to our emergency department on an early morning with the complaint of persistent and painful erection for three hours. He had some episodes of painful erections during the past three months, which were managed conservatively with penile aspiration and intra-cavernosal phenylephrine injections. Thereafter, he had remained asymptomatic till the current presentation. He denied that his present condition was in response to any sexual excitement or activity, penile trauma, or drug taking. He and his family had no history of blood disorders such as SCD, thalassemia, or leukemia. There was no past surgical history. On examination, the penis was hard and rigid, with severe pain on palpation of the tense corpora. The glans penis and corpora spongiosum were soft on palpation. There was no sign of infection. Approximately 100 mL of corporeal blood (dark red in color) was aspirated with a 22-G butterfly cannula. Blood gas analysis indicated low oxygen and severe acidosis (associated with ischemic flow type of priapism). As there was minimal response to aspiration, he was further treated with an alpha agonist. A total quantity of 400 mcg of phenylephrine dissolved in saline solution was injected into the cavernosal bilaterally. The patient's blood pressure was continuously watched during the

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procedure; vitals were checked post procedure and found stable. After 30 minutes, detumescence was achieved, and the patient felt free of pain, rendering the surgical procedure of distal shunting unnecessary.

At baseline, all relevant investigations were conducted to determine the possible cause of his ischemic type of priapism [Table 1].

Autoimmune-antiphospholipid syndrome tests were included (Anti-B2 Glycoprotein 1, immunoglobulin (Ig) G). The results of other tests such as IgM, anti-beta 2 glycoprotein, and anti-cardiolipin antibodies (IgG and IGM) were within a normal range. Extractable nuclear antigens were all negative, except for the anti-DFS70 value which was weakly positive. The hematology team advised no further investigations.

The patient was discharged on antiandrogen medication cyproterone acetate (50 mg once daily) for three months. On discharge, he was made aware that there was no specific medication to prevent recurrent priapism secondary to G6PD deficiency. He was advised to apply an ice pack to the genital area to manage early symptoms of priapism. Regular physical activity was also recommended. On later follow-ups, he continued to be asymptomatic. On his last visit, he revealed that he was taking tadalafil 5 mg (a phosphodiesterase 5 inhibitor) to treat weak erections. He was advised to continue taking cyproterone acetate (50 mg once daily) for three more months. A prescription for medication to treat erectile dysfunction was also given, to be used when needed, and its potential side effects were explained to him.

Informed consent was obtained from the patient.

DISCUSSION

G6PD deficiency is a genetic disorder leading to hemolytic type of anemia due to early breaking down of red blood cells (RBCs).[10] G6PD enzyme is crucial for keeping the blood cells in proper formation and preventing premature hemolysis. G6PD acts in the pentose phosphate pathway by catalyzing the conversion of nicotinamide-adenine dinucleotide phosphate (NADP) in the RBC to its hydrogenated form (NADPH), which is a reversible reaction. In normal conditions, RBCs contain sufficient amounts of NADPH. The ratio of NADPH to NAD indicates the extent of G6PD deficiency. When the RBCs are unprotected from the oxidative stress process, it

Table 1: Laboratory investigation results.

Test	Result	Reference range
G6PD	deficient (30% activity)	Normal (> 80% activity)
Antithrombin function 11a inhibition 20 seconds, u/mL	1.253	0.820–1.10
Antithrombin function Xa inhibition, u/mL	1.237	0830–1.100
Protein C functional chromogenic, u/mL	1.048	0.720–1.540
Protein C functional clotting, u/mL	1.078	0.800–1.810
Protein S functional, u/mL	1.271	720–1.450
Protein S antigenic, u/mL	1.013	0.65–1.390
Pro C global/factor V, AH	1.12	0.80–1.10
Factor VIII chromogenic assay, u/mL	0.982	0.580–1.880
HPT, g/L	0.79	0.30–2.00
LDH, U/L	290	135–250
Hb electrophoresis, %		
Hb A2	2.5	2.5–3.4
Hb F	0.3	0.1–1.1
HS	0.0	0.0–0.1
CBC	Within normal range	
Liver functions	Within normal range	
CRP, mg/L	2	0–5
Coagulation profile	Within normal range	
Bone profile	Within normal range	

G6PD: glucose-6-phosphate dehydrogenase; HPT: haptoglobin; LDH: lactate dehydrogenase; CBC: complete blood count; Hb: hemoglobin HS: high-sensitivity; CRP: C-reactive protein.

will cause a decrease in the level of NADPH, thus lowering the NADPH-NAD ratio.

G6PD deficiency is observed to be more prevalent in populations originating from the Eastern Mediterranean and Africa. The published case reports of G6PD-related ischemic priapism were on male patients of African-American and Afro-Caribbean ethnicities.[8,9] One case was of a 29-year-old youth with G6PD deficiency and recurrent ischemic priapism.[8] Another case was that of a 32-year-old Afro-Caribbean man, similarly affected.[9] G6PD deficiency is a common etiology of neonatal jaundice and admissions in urban hospitals in Jamaica.[11] There may be more cases of idiopathic priapism worldwide, whose potential link to G6PD deficiency may not have been evaluated. Therefore, non-SCD patients presenting with recurrent painful priapism should be investigated for G6PD deficiency, especially in countries with high levels of consanguinity such as Oman.

CONCLUSION

We have reported a rare case of ischemic penile priapism attributable to G6PD deficiency, which, to our knowledge is the first to be reported in Oman. Clinicians should consider G6PD deficiency in the differential diagnosis of idiopathic priapism.

Disclosure
The authors declared no conflicts of interest.

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