Post-Splenectomy Veno-Occlusive Priapism in a Child with Idiopathic Thromboctopenic Purpura

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Veno-occlusive priapism in the pediatric population occurs mainly in children with sickle cell disease. Less common causes for low flow priapism are leukemia, Fabry disease, retroperitoneal sarcoma, parotitis, and congenital syphilis. We report the first documented case of veno-occlusive priapism after splenectomy in a child with idiopathic thrombocytopenia purpura.

Patient Description
A 7 year old boy with ITP underwent an elective laparotomy and splenectomy because of persistent thrombocytopenia refractory to steroids. His postoperative course was uneventful until the fourth night after surgery, when he developed a painful erection. Hematologic evaluation revealed hemoglobin of 12.5 g/dl, hematocrit 36%, platelet count 780,000, and white blood cells 10,400 mm³. Coagulation studies were normal. Eighteen hours after onset of erection, the pediatric urology service was consulted. Examination at that time revealed an erect penis with painful tumescence of the corpora cavernosa and complete detumescence of the glans penis. He was immediately started on intravenous hydration of Ringer lactate, 2,500 ml/m²/24 hours and alkalization. Because of the long-duration priapism we decided to proceed immediately with corporal irrigation.

Under combined general and epidural anesthesia, 23 gauge needles were inserted into each corpus cavernosum and approximately 15 ml of dopped blood was aspirated from each. The cavernosal blood had O₂ saturation of 18%, and pH 7.14. Despite repeat irrigation with normal saline, tumescence persisted. Partial detumescence was finally achieved with intracavernosal injection of 450 µg (1.8 µg/kg) phenylephrine and 200 µg (8 µg/kg) epinephrine in multiple divided doses with close monitoring of pulse and blood pressure. The child required repeat irrigation and installation of intracavernosal epinephrine 10 hours later. Minimal degree of tumescence persisted despite the treatment. Oral aspirin was initiated, while hydration and alkalization were continued for an additional 24 hours.

The patient was discharged 4 days after irrigation with complete detumescence. His platelet count transiently increased to 1,000,000 mm³, then gradually decreased to 580,000 mm³ during 1 month. Three months following discharge his mother noticed normal erections.

Comment
Several etiologies have been described for veno-occlusive priapism. Sickle cell anemia [1], leukemia [2], Mycoplasma pneumoniae, thalassemia, and Rocky Mountain spotted fever [3] are the most common. Various mechanisms of intracavernosal clotting have been suggested [1-3].

Sickle cell disease is the most common cause of low-flow priapism in children, and occurs in 5% of these patients [1]. Priapism may be initiated by dehydration, acidosis secondary to hypoventilation during sleep, or normal nocturnal erections [1,2]. Decreased corporal blood flow during tumescence leads to increased oxygen extraction and crystallization of hemoglobin S, which limits venous outflow and results in priapism. Several mechanisms have been suggested for the etiology of priapism in children with leukemia, including sludging of leukemic cells within the corpora, leukemic infiltration of the sacral nerves, and mechanical obstruction of abdominal veins by splenomegaly [2]. A hypercoagulable state was the cause of priapism in a child with an upper respiratory infection secondary to Mycoplasma pneumoniae, and an inflammatory reaction initiated priapism in patients with Rocky Mountain spotted fever or thalassemia [3].

Post-splenectomy priapism has been reported previously, mainly in adults [3]. To our best knowledge there has been only one prior report of priapism following splenectomy in a prepubertal child, aged 12 years, who suffered from unstable hemoglobin [4]. We suspect that the priapism in our patient occurred due to the relative hypercoagulable state caused by thrombocytosis combined with relative dehydration and hypoventilation during sleep. In the present case, priapism occurred 4 days following splenectomy, while in all previously reported cases 4 weeks to 23 years passed between splenectomy and the occurrence of priapism. After detumescence had been obtained, we initiated oral aspirin in an attempt to decrease platelet aggregation and, hence, reduced the likelihood of recurrence.

1ITP = idiopathic thrombocytopenia purpura
Initial management of veno-occlusive priapism includes vigorous hydration and alkalization. Should these modalities fail, invasive treatment with corporal irrigation and injection of intracavernosal alpha agonist agents should be initiated immediately [5]. Prompt and decisive treatment may preclude long-term implications of priapism.

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References

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Hemolytic Anemia in a G6PD-Deficient Man after Inhalation of Amyl Nitrite ("Poppers")

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Inhaled nitrates are illegal to use as aphrodisiacs or as mood-altering substances, usually by young adults. These substances are commonly known by the terms “pop- pers”, “snappers” or “rush”. Their smooth muscle-relaxant property makes them popular especially among homosexual men, and their use was found to be significantly associated with unsafe sexual habits in this population [1].

There are no data on the extent of the phenomenon in Israel, but it seems to be quite prevalent. In the United States, the use of “poppers” has been reported to be as high as 11% among occasional recreational drug users and 22% among heavy drug abusers [2]. Inhaled nitrates (most commonly the amyl derivative) have several adverse effects, some of which are hematologic. A number of cases of methemoglobinemia, Heinz body formation, and hemolytic anemia after “poppers” inhalation has been reported worldwide, and are attributed to the oxidative stress caused by nitrates [3-5]. We describe here a 19 year old man with glucose-6-phosphate dehydrogenase deficiency, who developed acute hemolysis after inhalation of amyl nitrate.

Patient Description
A 19 year old male of Jewish Sephardic origin was hospitalized in an internal medicine department complaining of severe headache and jaundice. The patient’s medical history was unremarkable except for G6PD deficiency and Kawasaki disease at the age of 10. Upon questioning, the patient admitted having inhaled volatile substance he identified as “poppers” on the previous day. He claimed that it was the first time he had used this substance. After inhaling the drug he felt severe headache, and profound jaundice was noted the following day. The patient denied taking any other medications, or food that could possibly have exacerbated the hemolytic anemia of G6PD deficiency.

On physical examination the patient was hemodynamically stable, blood pressure was 130/60 mmHg and heart rate 82 beats/minute. Jaundice of the skin and sclera was prominent with no splenomegaly or hepatomegaly. The rest of the physical examination was unremarkable. The complete blood cell count on admission revealed a hemoglobin level of 13.0 g/L (normal 13.5-17.3 mg/dl), hematocrit 38.4% (normal 35-49%); and the indexes of red cell distribution width, mean cell volume, mean cell hemoglobin, and mean cell hemoglobin concentration were normal. White blood cell count and platelet count were also normal. The noteworthy finding in blood chemistry results was high total bilirubin, 12.4 mg/dl, of which the direct component was 0.49 mg/dl. Lactate dehydrogenase was 430 U/L and all other chemistry parameters were normal. No traces of blood or bilirubin were detected in a urine sample. The following day the patient was severely icteric, but otherwise felt well. Hemoglobin level dropped to 11.9 mg/dl, hematocrit dropped to 33.9%, reticulocyte count was 5.2% (reticulocyte index = 2.6), haptoglobin level was <3.8 mg/dl.