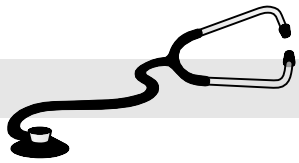

Post-Splenectomy Veno-Occlusive Priapism in a Child with Idiopathic Thrombocytopenic Purpura

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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 mm^3. 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/m^2/24 hours and alkalinization. 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 clotted blood was aspirated from each. The cavenosal blood had O_2 saturation of 18%, and pH 7.14. Despite repeat irrigation with normal saline, tumescence persisted. Partial detumescence was finally achieved with intracavenosal injection of 450 μg (18 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 alkalinization 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 mm^3, then gradually decreased to 580,000 mm^3 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.

ITP = idiopathic thrombocytopenia purpura

Initial management of veno-occlusive priapism includes vigorous hydration and alkalinization. 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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