Priapism in a 15-Year-Old Boy With Major Beta-Thalassemia

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INTRODUCTION

Priapism (painful erection longer than 6 hours) is an uncommon disease that is almost always due to thromboembolic problems. It may occur at any age. The predisposing factors for priapism include thromboembolic disorders such as sickle cell anemia (SCA, the most common cause), sickle-beta-thalassemia, diseases of the central nervous system and the spinal cord, metastatic cancers, trauma, iatrogenic factors, and infectious diseases (malaria and rabies).[1] We present an adolescent patient with major beta-thalassemia who suffered from an episode of priapism. To our knowledge, no other cases of priapism have been reported in association with major beta-thalassemia, to date.

CASE REPORT

A 15-year-old boy presented with priapism lasted for 7 hours. The patient was admitted and conservative treatment including oxygenation, hydration, analgesics, and epinephrine injection (3 times with 20- to 25-minute intervals) was initiated. After 10 hours, priapism was alleviated. At the time of reference, vital signs were as follows: body temperature, 37.2°C; blood pressure, 100/70 mm Hg; and pulse rate, 84/min. The patient was a known case of major beta-thalassemia, but he did not have any other special conditions including cardiac, pulmonary, or infectious diseases. No history of trauma was mentioned by the patient. Splenectomy had been performed 8 years earlier and blood transfusion had been performed for several years until 5 years earlier.

Results of the laboratory tests were as follows: hemoglobin (Hb), 10 g/dL; hematocrit, 31.5%; white blood cell count, 15.4×10^9/L; platelet count, 156.7×10^9/L; nucleated red blood cell count, 28 per 100 leukocytes; erythrocyte sedimentation rate, 14 mm/h; and serum ferritin, 696 ng/mL (reference range, 38 ng/mL to 457 ng/mL). On the blood smear, Howell-Jolly bodies, target cells, giant platelets, and hypochromia were present. Direct Coombs' test and viral hepatitis tests including hepatitis B antigen and hepatitis C antibody were negative in the serum sample; however, serum was positive for hepatitis B surface antigen. On electrophoresis, the HbA was 34.8% (reference range, 96.5% to 98.5%); HbA_2, 2.6% (reference range, 1.5% to 3.2%); HbF, 62.6% (reference range $< 2\%$); and HbS, 0%. Also, Sickle cells were not detected in the smear. Blood gas analysis of a sample taken from the corpus cavernosum demonstrated that priapism was

low flow (venous type). Other laboratory tests including serum electrolytes, coagulation tests, and liver, kidney, and thyroid function tests had normal results. After 24 hours, the patient was discharged in a good general condition. During a 2-year follow-up, no recurrence was reported.

DISCUSSION

We presented a case of priapism in an adolescent with major beta-thalassemia. Regarding the lack of any other underlying disease or a history of trauma, it seems that in this particular patient, there was a causal link between priapism and major beta-thalassemia. To our knowledge, this is the first case of priapism with major beta-thalassemia as the only risk factor. In a study in Italy, priapism was reported in a patient with intermediate thalassemia.[2] Also, in 52 patients with priapism, Fowler and colleagues showed that 20, 10, 19, and 3 patients had SCA, sickle-C anemia, sickle trait, and sickle-beta-thalassemia, respectively. But, none of them had a major beta-thalassemia.[3]

In children, the etiology of priapism is SCA in 63% of cases and 6.4% of patients with SCA experience priapism.[1] In SCA-induced priapism, the disease is recurrent and gradually results in ischemia of the corpus cavernosum, venous obstruction, and impotence which is more common in adults.[1,4] Therefore, although the disease is mostly prevalent in adolescents, the possibility of impotency is less than the adults.[4] Most of the priapism episodes occur between 2 AM to 6 AM during the day. It usually occurs following dehydration and metabolic acidosis during the sleep.[1] The mechanism of priapism in SCA is an increment in the blood concentration and changes in the form of hemoglobin. In beta-thalassemia, the mechanism might be related to hyperviscosity like other hemoglobinopathies.[4] The 2 types of the disease are venous, which is more common, and arterial.[4]

We successfully treated our patient conservatively with oxygenation, hydration, analgesics, and epinephrine injection. The first therapeutic step is conservative treatment that has been efficient in many patients with hematologic diseases, and the vast majority of children will improve by medical treatment without the need for surgery. If resistant to the routine treatment modalities, priapism should be treated by surgical operation and spongicavernous shunt insertion.[1,5] Conservative treatment in the first stage includes hydration, oxygenation, and metabolic alkalization (for reduction of sickling in SCA). In the second stage, the treatment includes supertransfusion, erythrophoresis, irrigation, and injection of alpha-agonist drugs into the corpus cavernosum every 5 minutes after aspiration of 10 mL to 20 mL of blood.[6]

CONFLICT OF INTEREST

None declared.

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